JUNE 1991
VOL. 50, No. 6
ISSN: 0017-8594

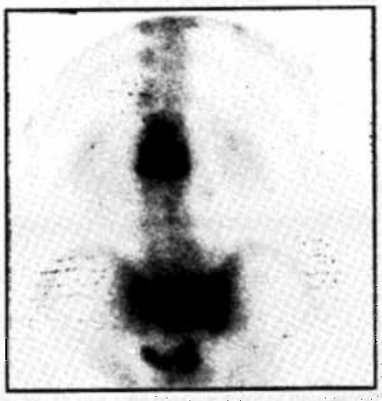
Hawaii Medical Journal

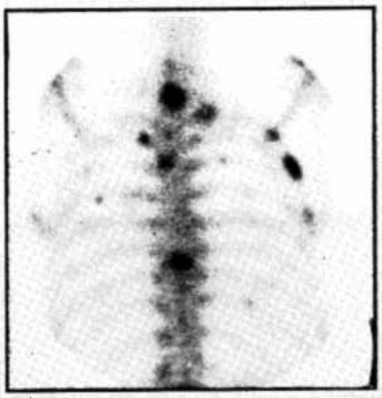
CONTENTS

NUCLEAR MEDICINE TEACHING FILE

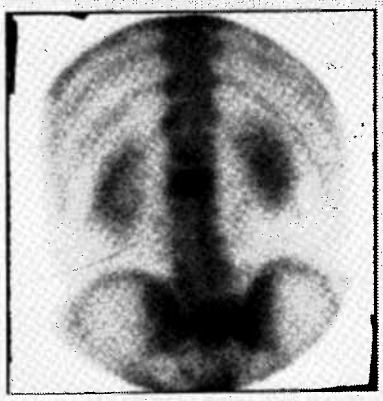
BONE SCANS IN MONITORING METASTASES

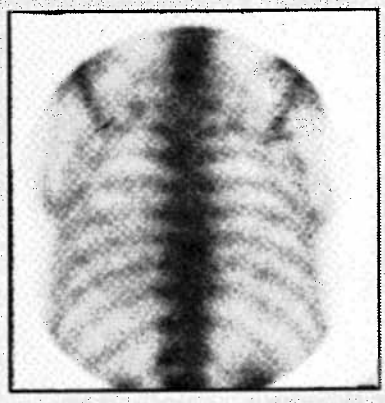
65 year old male with prostate CA followed with bone scans after therapy with diethylstilbesterol (DES).





1 A & B
Initial scan demonstrates multiple spinal and rib metastases.





2 A & B
Six months later there is marked reduction in the number and size of the bony metastases indicating an excellent response to therapy.

Hawaii Medical Journal

(USPS 237-640)

Published Monthly for the
Hawaii Medical Association
(Incorporated in 1856 under the Monarchy)
1360 South Beretania, Honolulu, Hawaii 96814
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Look carefully at sashimi

Anisakiasis should be one of the more common afflictions in Hawaii, considering the amount of raw seafood consumed by our people.

The *Journal* had an extensive review article on the subject in its January 1991 issue; however, we cannot resist publishing a follow-up.

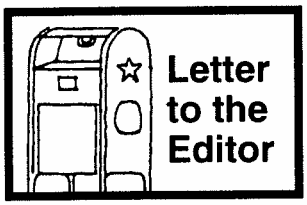
JABSOM student Joy Hiramoto, who is about to receive her MD and graduate from the school, produced the article in the current issue under the aegis of Jinichi Tokeshi.

Not only was the diagnosis remarkable in that the patient made it (he was a physician, after all!), but we would like to challenge any of our readers to find that worm on the photocopy of the film.

Sorry, there is no monetary reward for doing so.

And, what did happen to the worm?

J. I. Frederick Reppun MD
Editor



A response

To the Editor:

On behalf of Papa Ola Lokahi (POL), I would like to express our appreciation for the editorial in your November 1990 (Vol. 49, No. 11) issue of the *Hawaii Medical Journal,* bringing the attention of your members to our efforts to improve the health of Hawaiians through the Native Hawaiian Health Care Act (Public Law 100-579) (hereinafter referred to as "the Act").

We also appreciate the editorial's closing statement that "the Hawaii Medical Association (HMA) is willing and able to help *Papa Ola Lokahi* whenever we are asked to do so." I have in fact met several times with your Native Hawaiian Committee over the past year and a half and will identify what other actions the HMA might take. First, however, I would like to respond to some of the specific assertions contained in your editorial.

First, on your editorial's statements that POL has "only 5 members," and that "(n)otable in an organization that is to plan health care is the absence of a physician (not even a *Kahuna Lapaau!*/Ed)." POL is an organization created by the Act, and is statutorily determined as being comprised of the 5 organizations listed in your editorial: Alu Like, Inc., E Ola Mau, the Office of Hawaiian Affairs, the Office of Hawaiian Health of the State Department of Health and the University of Hawaii. We have 2 health professionals on the Board. The representative from the Office of Hawaiian Health of the State Department of Health is Fern Clark, the Office's Director and a nurse; and E Ola Mau's representative is Nanette Judd, its president and also a nurse.

As for "the absence of a physician," I am POL's Executive Director and am a physician as well as a lawyer. As for "an organization that is to plan health care," I also chair the Long-Range Planning Committee of the John A. Burns School of Medicine.

More importantly, 3 of the 5 Native Hawaiian planning committees that have been established are chaired by allopathic physicians, and all are Hawaiians and graduates of the John A. Burns School of Medicine. They are R. Wayne Fukino MD, of Kaua'i, Noa Emmett Aluli MD, of Moloka'i, and Joseph Kamaka III MD, of Maui. The co-chair for O'ahu is Mitchell Eli DC.

Second, your editorial states that "the total appropriation is $2.6 million the first year, $6 million the second year and $11 million the third year," and also states that "Federal funds became available in FY 90, ie in October 1989, initially for *Papa Ola Lokahi's* organizing and planning process to the tune of $100,000 for its administrative effort and $700,000 for the development of the Master Plan." One might infer from these statements that POL has chosen to spend only $800,000 of $2.6 million in the first year in which funds became available.

In the legislative process, when a law is enacted, this *authorizing* legislation specifies the *maximum* amount of funds that can be made available in each year. As each new fiscal year approaches, the legislature specifies the amount to be actually *appropriated.* In the first year, $2.6 million was *authorized,* not appropriated; similarly, in year two, the authorized amount was $6 million, and in year three, $11 million. Appropriations seldom, if ever, equal the authorized amounts. In the first year, $800,000 was the amount actually *appropriated* by the U.S. Congress, compared to the authorized amount of $2.6 million. Our fiscal year 1991 appropriations are $2.7 million, out of an authorized level of $6 million. Of the $2.7 million appropriated, $2.3 million is for initiation of services, to be applied for and granted to each island's Native Hawaiian service delivery system. The remaining $400,000 is

(Continued on page 198) ➤

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for POL's various activities.

Additionally, the $2.6 million in authorizations for the first year were specified for the following purposes: a) $700,000 for development of a Master Plan, b) $900,000 to plan for up to 9 health systems throughout the state, and c) $1,000,000 for all other POL responsibilities. As your editorial states, our actual appropriation was $800,000; but in addition to POL's organizing and planning, and development of a Master Plan, these funds were also to cover the costs of planning the health delivery systems. We have allocated $525,000 of the $800,000 to planning of these systems.

The $800,000 did not become available in October 1989. That was the *beginning* of the 1990 federal fiscal year. We received instructions from the federal government for writing our grant proposal in April 1990; we responded by the end of that month and were notified of acceptance of the proposal and received our first payment of the $800,000 in July 1990. We were fortunate in being able to start our initial activities in July 1989 with $200,000 that the Hawaii State Legislature provided. (For a more detailed explanation of the funding history, please see the December 1990 issue of the Office of Hawaiian Affairs' newsletter, *Ka Wai Ola O Oha*).

Third, we are acutely aware that "the (master) plan (should) meld into the existing health care system, especially with the introduction of the State Health Insurance Program (SHIP) enacted into law by the Hawai'i State Legislature earlier this year."

To quote from the summary of the Master Plan document which we distributed in November 1989 and which your editorial states that Dr. Mills has studied in-depth:

> "In addition to the health services provided under the Act, there exists and will continue to be a need for other State, Federal, and private efforts directed at improving the health of Native Hawaiians. Therefore, *a comprehensive health-care master plan must take these other resources and services into consideration,* and the following objectives must be met in order to address the health needs and improve the health status of Native Hawaiians: 1) assessing the health status and health-care needs of Native Hawaiians living in Hawai'i; 2) planning and establishing the resources and services that are called for under the Native Hawaiian Health Care Act; 3) *identifying and evaluating all other current and planned health resources and services for Native Hawaiians;* 4) *coordinating these various resources and services;* and 5) developing monitoring and evaluation systems in order to improve and create health resources and services and to measure progress toward improving the health of Native Hawaiians (emphasis added)."

As for SHIP, you can be assured that we are well aware of the interrelationships between the Act, SHIP and our Master Plan. I represented both POL and the School of Medicine on the SHIP committee advisory to the Department of Health throughout the Department's planning and implementation of SHIP.

Fourth, your editorial states that "*Papa Ola Lokahi* would do well to avoid discrimination in reverse to raise its ugly head." This is accompanied by a warning to POL that "the matter of establishing primary health-care centers ... smacks somewhat of establishing another tier of health care ... ie of a different quality. This is not desirable. However, if this mandate in the Act can be stretched to allow Health-Care Centers or Organizations to farm out services to existing primary care facilities, then it might not be so difficult to implement the Act."

Services under the Act will be under the management and control of Hawaiians, and planning is being conducted under the direction of Hawaiians, who are striving to create health services that are special and uniquely Hawaiian. There is no need to lecture POL and the island planning committees in such patronizing language.

Furthermore, the mandate of the Act need not "be stretched" to achieve this purpose. In fact, our plans for providing primary care (the 3 broad service areas under the Act are primary care, health promotion and disease prevention) are quite similar to the editorial's hopes, although I would not characterize these primary care services as being "farmed out." We want the beneficiaries of the future Native Hawaiian systems to have a choice, and we want providers of services to be sensitive to the culture and style of Hawaiians.

This brings me to the identification of actions that the HMA could take to assist in realizing the objectives of the Act.

As stated earlier, I have had several meetings with the Native Hawaiian committee of the HMA, which was first chaired by Dr George Mills, and now by Dr Ernest Bade, who is also president of your Hawai'i County Medical Society. Dr. Bade attended the most recent meeting of the Hawai'i Island Planning Committee on November 26, offering your Association's assistance. Previously, at an August 8 meeting between your committee and myself, we discussed ways of directly involving more physicians in the island planning committees. I subsequently wrote a short article for your newsletter, describing our efforts and asking physicians to contact POL if they were interested in participating in the planning activities on their respective islands. We received only a single inquiry, from a physician on Maui. So we need to explore other ways of getting more of your members interested in direct participation.

Finally, we do not want to limit our beneficiaries' access to "existing primary care facilities" to those providers who serve primarily the economically disadvantaged and medically underserved. We want to avoid placing our beneficiaries not only in "another tier of health care," but also in the lower tier of current services.

One of the requirements of the Act is that our systems and those providers to whom services are "farmed out" must be participants in Medicaid. Thus, *the most important action the HMA could take* in helping "to avoid discrimination in reverse to raise its ugly head" *is to encourage your members to increase their participation in the Medicaid program.* Let us not be satisfied in this undertaking until Native Hawaiians have access to the services of all physicians in Hawai'i.

Lawrence Miike MD JD
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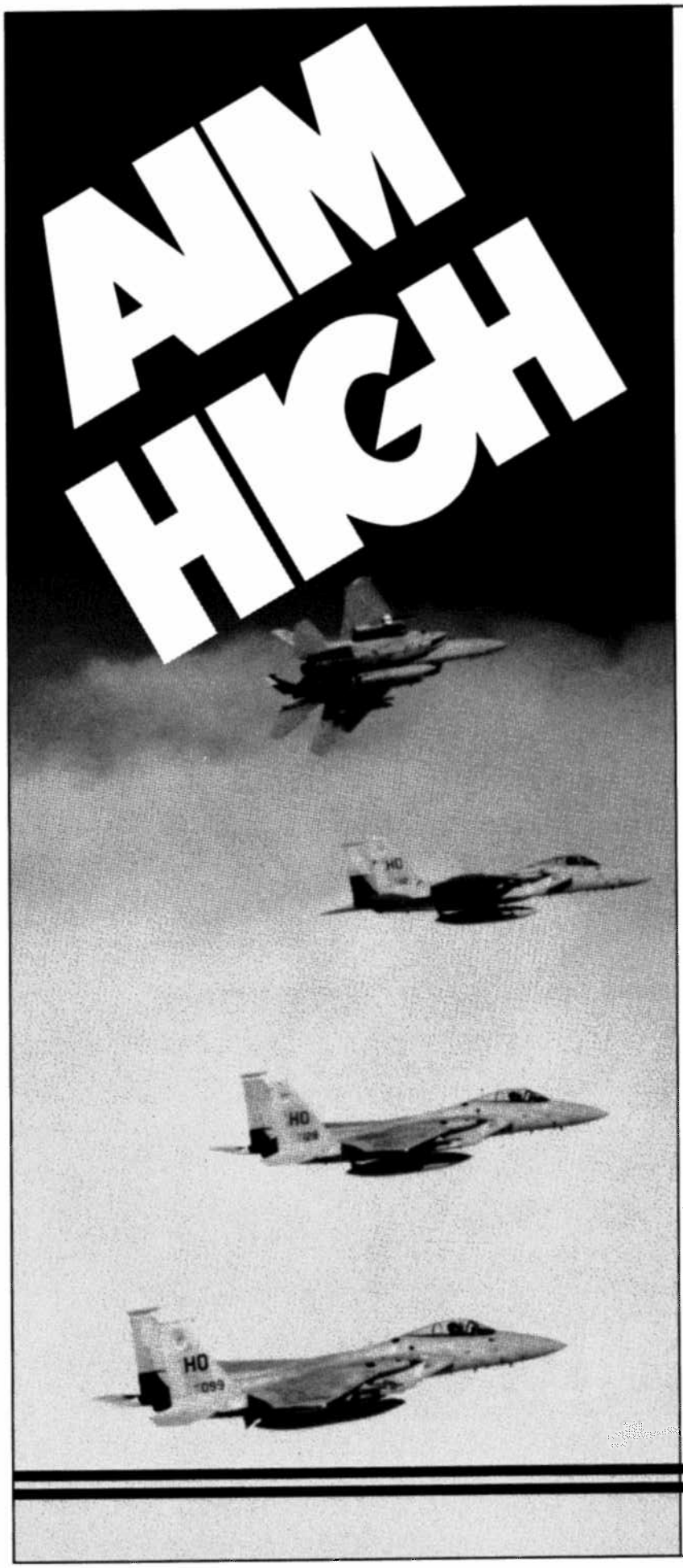
Honolulu, Hawai'i 96813

The editor responds:

We bid Dr Miike's pardon in failing to recognize his status as a physician and a lawyer. We are also appreciative of his clarifications.

J. I. Frederick Reppun MD
Editor



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AIR FORCE

Highlights of the HMA Council Meeting — May 3, 1991

Present: J. McDonnell, S. Wallach, A. Don, R. Stodd, C. Kam, A. Kunimoto, R. Ando, B. Shitamoto, C. Wong, R. Laird, P. DeMare, H.K.W. Chinn, M. Dung, E. Morgan, M. Shirasu, J. Spangler, H. Fong, W. Young, E. Bade, R. Goodale, D. Fu, H. Percy, T. Smith, G. Goto, J. Lumeng, W. Chang, W. Dang, and N. Winn. Also present were: Legal Counsel - V. Woo, HMJ Editor - F. Reppun MD, medical students - N. Kaneshiro and J. Taitague; M. Cooper MD, and guest Lawrence Hart MD - DOH. HMA staff present: J. Won, N. Jones, B. Kendro, J. Asato, J. Estioko, L. Tong Esq, P. Kawamoto, and M. Lindsey, recording secretary.

• Larry Hart was introduced to Council. He recently joined the DoH to work with the Medical School in coordinating services to the public.

• President John McDonnell, with assistance from President-elect Steve Wallach, told of the 3-day Washington DC trip as part of California Medical Association's annual visit with Congressional representatives. Western states: Hawaii, Alaska, Washington, Idaho and Oregon were invited to participate with California. Meetings were held with members of HCFA and Hawaii members of Congress, including former Hawaii Congress member Pat Saiki, who is the recently appointed chair of the Small Business Administration.

• The Senior Physicians Committee recommended that HMA investigate the pros and cons of a proposed Long-Term Care Financing Authority and, in the event it is authorized, that at least one, but preferably more, practicing physicians be appointed to serve on the Long-Term Care Financing Board and planning committee. The physician who is appointed to the Board should be practicing primary care actively in Hawaii, should be involved in long-term care, should be a member of the HMA and to serve as a liaison with organized medicine. It also recommended that the HMA be used as a consulting base whenever possible.

• An HMA policy statement was approved: Laser surgery or other invasive procedures using lasers for medical purposes should be done only by appropriately trained physicians.

• A new HMA membership benefit was approved. This benefit will allow HMA members to purchase medical office supplies from Galaxy Business and Medical Products, Inc. at approximately an 18% discount from the retail prices.

• Three resolutions to be submitted to the AMA Annual Meeting of the House of Delegates in June were approved. 1) It asked the AMA to work with the American Bar Association to promulgate guidelines for the role of an expert witness; 2) it asked the AMA to take appropriate action to alert the public concerning the Philip Morris Company's use of a nationwide Bill of Rights Tour which focuses on America's youth in advertising tobacco products; and 3) it asked for the AMA Council of Medical Education to reconsider the new mandated Category II credits for the Physicians Recognition Award and to return to previous regulation.

• Steve Moser of Maui, Chair of the HMA Environmental Health Committee, requested HMA's help in alerting all county medical societies and their officers about the likelihood of contamination with lead (P6) in any water catchment systems in their respective counties. The recommendation was approved.

Andrew Don, MD
HMA Secretary

Anisakiasis in Hawaii: A radiological diagnosis

Joy T. Hiramoto MS IV *
Jinichi Tokeshi MD**

Human anisakiasis, an increasing medical problem in Japan, was recently identified in 7 people in Hawaii[1]. We report an 8th case occurring in an individual who had consumed a large amount of raw squid sushi at a restaurant. The diagnosis of anisakiasis in this patient was made after the parasite was identified by X-ray. To our knowledge, radiographic demonstration of the anisakiasis parasite has not been reported in Hawaii previously.

Introduction

Infection by the nematode anisakis in humans was first reported in 1960 by Thiel[2]. Since that time the incidence of anisakiasis has increased, especially in Japan, where numerous new cases are reported each year. In Hawaii, 7 cases have been reported, the diagnosis made by the endoscopy and the radioallergosorbent test (RAST). The lifecycle, diagnosis, treatment, and overall management of infection with anisakis was extensively reviewed by Deardorff, Kayes and Fukumura[1].

In brief, Anisakis is a nematode that commonly infects the gastro-intestinal tract of marine mammals, after these mammals have ingested the infected flesh of fishes or crustaceans[1,3,4]. Humans can also become infected after eating the raw flesh of infected fishes such as salmon, mackerel and rockfish (red snapper). Infections have also occurred after eating raw squid[1,4,5,6].

Typically, a patient may present with severe, intermittent epigastric pain 1 to 12 hours after eating such food. Nausea, vomiting, diarrhea and urticaria may or may not ensue. Fever, leukocytosis with or without eosinophilia, and occult blood in the stool may also be present in some cases[1]. Symptoms may mimic acute gastroenteritis, food poisoning[5], appendicitis, cancer, ileitis, TB peritonitis, cholecystitis[8] and Crohn's Disease[9].

Diagnosis can be made radiographically by actually seeing the parasite on film. The presence of broad gastric folds, bowel edema[10,11], or widening of the gastric angle, in symptomatic patients with a history of consuming seafood can also suggest anisakiasis[4,5,6,11]. However, the diagnosis is usually made by endoscopy and/or RAST[1,12,13].

Treatment consists of endoscopic removal of the worm, in addition to symptomatic management[1,4,5,6].

Complications of severe anisakiasis include intussuception with intestinal obstruction[7], severe ascites, penetration of the bowel wall by the juvenile form[9] or ileus[8]. Anisakiasis has also been linked to the formation of an eosinophilic, granulomatous, gastric tumor[14]; the worm has also been reported to have migrated into the mesentery, the pancreas[8] and the liver[9].

Infections by anisakis can be prevented if raw items are cooked or blast-frozen prior to consumption[1,3].

Case report

A 33-year-old physician was seen in the office with a chief complaint of intermittent, sharp, non-radiating epigastric pain 3 hours following consumption of a large amount of raw squid at a sushi bar. The pain was accompanied by diaphoresis and nausea, but no fever or vomiting.

Past medical history and family history were not remarkable, and the patient denied being on any medications. Social history was significant only in that alcohol was imbibed infrequently. Review of symptoms was non-contributory.

On physical examination, the vital signs were: BP 130/88 P 84/min, T 99°F. Examination of the abdomen revealed mild tenderness to deep palpation in the epigastric region. Guarding, rigidity, and rebound were not present; bowel sounds were normal. Rectal examination was unremarkable; the stool was brown and guiac negative. The rest of the examination was not remarkable.

Laboratory data revealed a WBC of 11.1 with a normal differential.Eosinophilia was not present. The Hgb was 16.1, Hct 46.6, platelets 189,000, BUN 15, Creat 1.0, SGOT 25, SGPT 41, GGT 54 and total Bilirubin 0.8. An upper GI series was obtained. The initial reading reported no abnormalities. The patient was treated symptomatically and left Hawaii the same day. He later reported spontaneous resolution of the symptoms within 24 hours. No further followup seemed necessary, therefore.

The patient, being a physician, reviewed the upper GI films and found a 0.5 mm x 2 cm thread-like, radiolucent, filling-defect partially surrounded by a pool of barium in the body of

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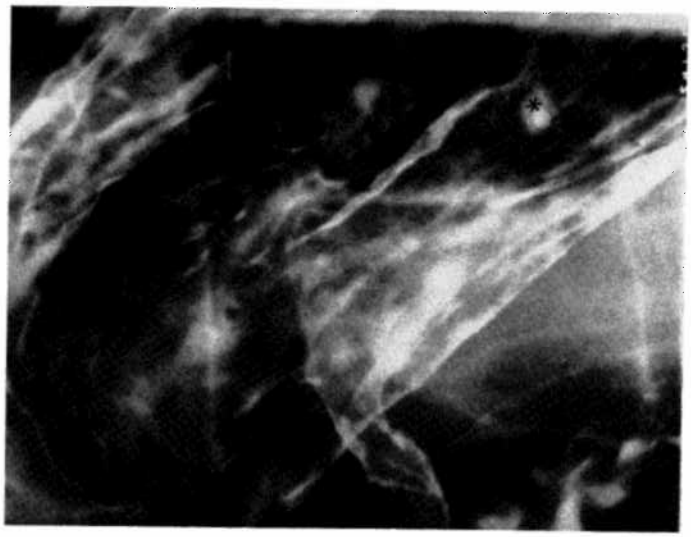

FIG. 1 — X-ray photograph of the nematode Anisakis in the wall of the stomach. The parasite appears beside the asterisk as a serpiginous 0.5 mm x 2 cm filling-defect. Note the small pool of barium, representing the site of attachment, partially surrounding the worm.

the stomach (Fig. 1). A surgeon by profession, the patient immediately made a diagnosis of anisakiasis based on his clinical experience with similar infections in his own patients. He returned to Hawaii approximately 2 months later and brought up his case for discussion by his peers. Consequently, a corrected diagnosis of anisakiasis was made.

Discussion

This case, in which the patient made the diagnosis, emphasizes the need for greater awareness by physicians concerning anisakiasis. No further workup, such as endoscopy or RAST, was necessary on this patient because his symptoms resolved within 24 hours; the upper GI X-ray was read as "normal" and anisakiasis was simply not suspected.

Since then, we have discussed this case with several local radiologists. They also read the X-rays as normal and were surprised that the nematode could be seen on film. However, the thread-like filling-defect, surrounded by a small pool of barium (the site of attachment by the worm), that can be seen in this patient's films is what is frequently demonstrated in Japan. In one Japanese study of 130 patients, thread-like filling-defects in circular or ring-like shapes were seen in 47.7% of patients with gastric anisakiasis[6]. Sugimachi et al[6] also interpreted the small, round collection of barium associated with the thread-like filling-defect as the parasite's site of attachment to the mucosa. Nakata et al[5] reported the same kind of serpiginous, thread-like filling-defect in 31 of 41 patients (76%) studied. Other workers have also been able to demonstrate this in the small bowel[11] and in the large bowel[10].

Residents living in Hawaii have many opportunities to sample new seafood dishes in the home or at local restaurants. Although relatively few cases have been reported locally, it is quite likely that such infestations have gone undiagnosed[1]. The radiographic evidence of the presence of anisakis, for example, is subtle and easily overlooked, and the edema in the stomach or bowel which may occur could also be caused by other gastrointestinal diseases. As a result, endoscopy with or without RAST would be more useful than X-ray to confirm the presence of the anisakis. However, if the parasite is apparent radiographically, this finding may obviate invasive procedures (eg exploratory laparotomy for a diagnosis of acute appendicitis) and aid in the overall management of the patient's illness.

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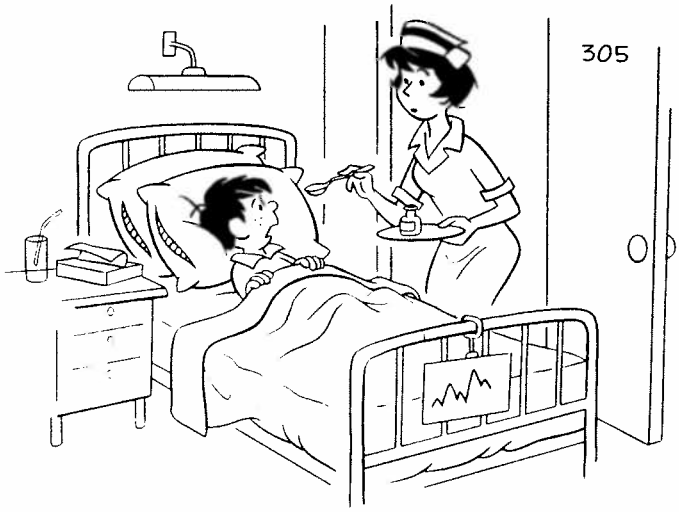

Sarcoidosis: A Hawaii rarity

Jonathan Charles MD*
David J. Elpern MD**

Sarcoidosis is so rarely seen in residents of Hawaii that it may not be considered as a diagnostic possibility. The differential diagnosis is more complex in Hawaii due to the presence of granulomatous diseases such as tuberculosis and leprosy. We present the first known case of sarcoidosis in a Hawaii resident together with an overview of the disorder.

Introduction

Sarcoidosis is a noncaseating, granulomatous disease which can affect virtually any organ system. A high percentage of patients undergo spontaneous resolution.

Sarcoidosis is common in certain areas of the U.S. and its clinical presentation is well known to most physicians there. The disease is rare in Hawaii and, therefore, may not be easy to diagnose at first glance. Complicating the diagnosis is the fact that other entities in the differential diagnosis are far more prevalent in Hawaii than on the Mainland. For instance, more cases of leprosy and Mycobacterium tuberculosis are seen in Hawaii, yet on the Mainland these diseases are rarely seen. In addition, granulomatous reactions to plants (particularly cactus and bromeliads), insects and sea urchins are commonly encountered in Hawaii and may simulate sarcoidosis.

We recently diagnosed the first case of sarcoidosis occurring in a long-standing resident of Kauai. The clinical findings were subtle and the diagnosis was not immediately obvious.

Case report

A 40-year-old woman presented to one of us (DJE) with small, somewhat painful nodules on the extensor aspects of both forearms that had been present for 4 months.

The patient was born in Portugal, where she had resided for the first 16 years of life; thereafter she lived in Mozambique. Later on, she emigrated to Kauai, Hawaii, where she has resided for the past 13 years.

The patient's past medical history was unremarkable. She was a para III, the last pregnancy having been in 1982, at which time she had undergone an elective bilateral tubal ligation. She had had a similar skin eruption on both forearms in 1988 which had resolved in 4 months. At that time, a chest X-ray was normal (Fig. 1).

The patient and her husband are farmers. Two months prior to her presenting herself to us, several red, tender nodules appeared on the anterior aspects of both lower legs. They disappeared in a few weeks. There were no other symptoms, specifically no respiratory difficulties and no visual complaints.

The physical examination was remarkable for several erythematous, flat-topped papules and plaques measuring 2 to 15 mm in diameter, arranged in an almost symmetric fashion on the extensor surfaces of both forearms. A barely visible circinate lesion measuring 8 cm in diameter was present over the anterior surface of the lowermost portion of the right leg.

A 3 mm punch biopsy of one forearm lesion revealed closely-packed, noncaseating epithelioid granulomas confined primarily to the upper dermis (Figs. 3, 4). The granulomas were bordered by a moderate lymphocytic infiltrate. A few multinucleated giant cells were present, including a rare Langhans cell. Birefringent particles were not identified under the polarizing lens. Fungal elements were not identified with the PAS stain and the AFB stain was negative. These findings were interpreted as consistent with sarcoidosis. However, neither leprosy nor a foreign body reaction (including to cactus

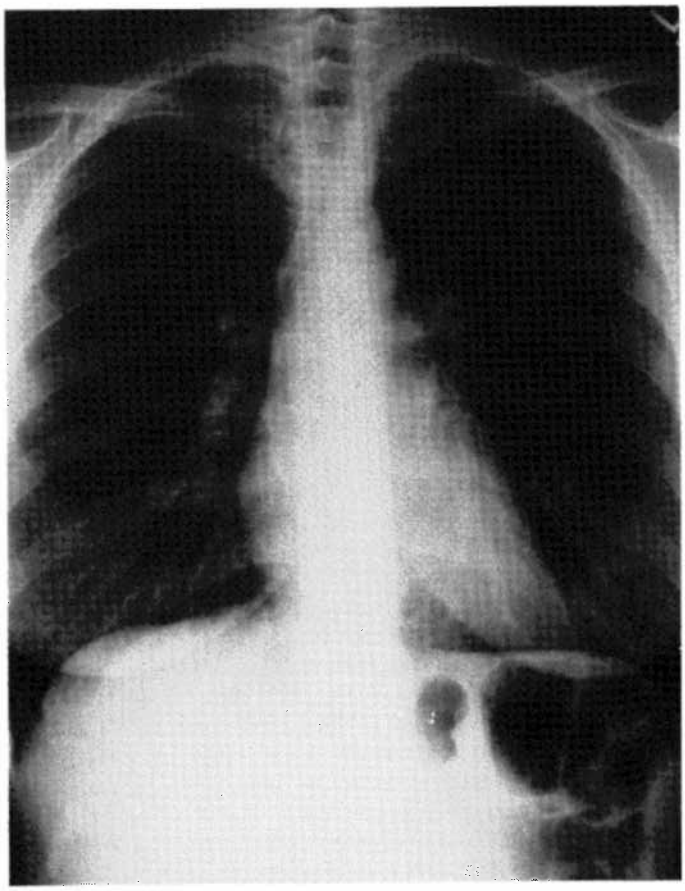

Fig. 1 Normal baseline chest X-ray, 1988.

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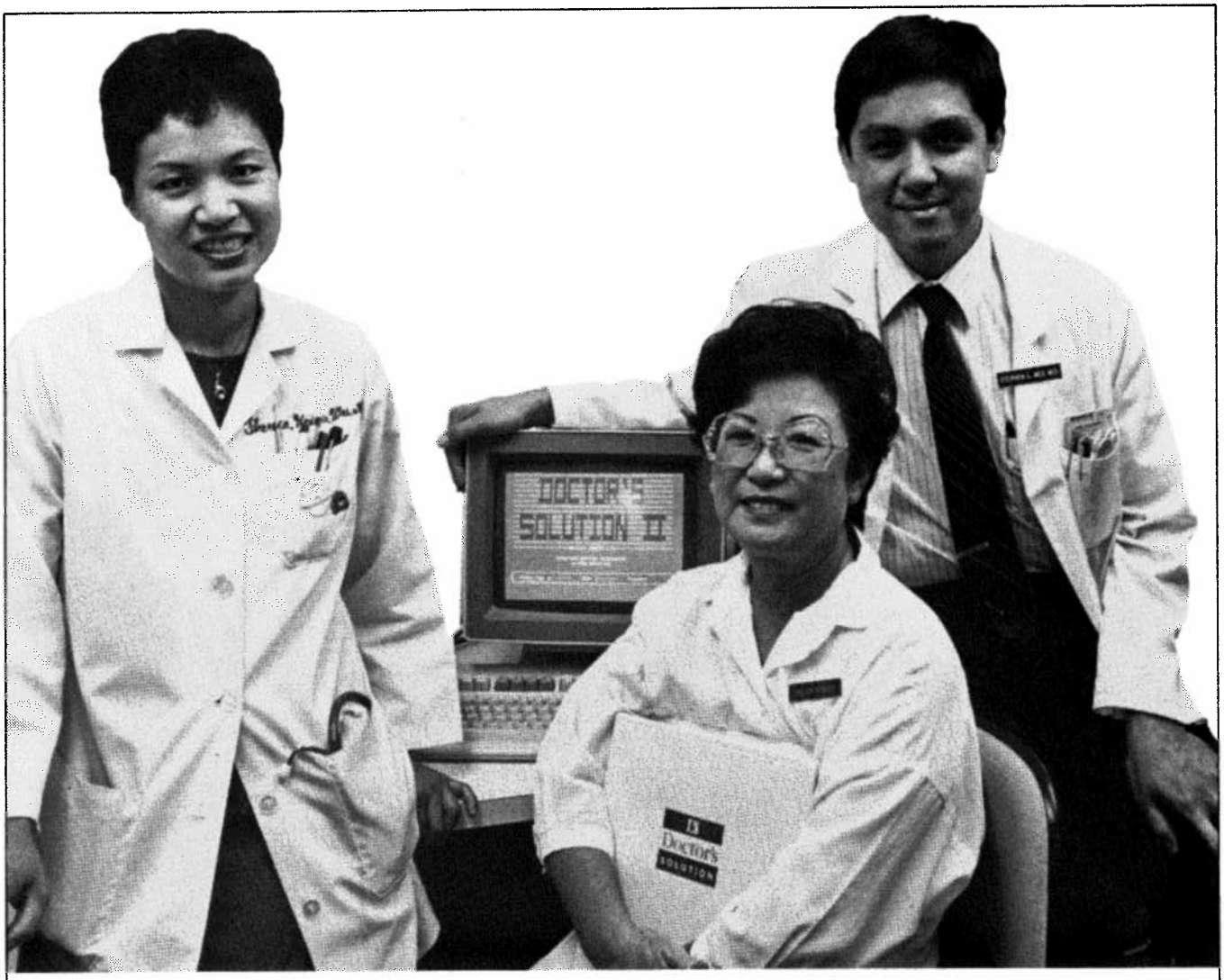
Kauai Medical Group and Wilcox Memorial Hospital
Lihue, Hawaii 96766



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and similar plants, sea urchins, and insect bites) could be ruled out by histologic methods alone.

A chest X-ray revealed prominent, left hilar adenopathy and possible right hilar adenopathy as well (Fig. 2). Pulmonary function tests were normal; sputum culture was negative.

Laboratory studies (including calcium), urinalysis, and CBC were all within normal limits. Angiotensin Converting Enzyme (ACE) was 10.4 activity units (normal 2.0-7.5).

The patient was diagnosed as having sarcoidosis and no specific treatment was offered due to the mild presentation. The lesions on her lower extremities were diagnosed as erythema nodosum. At the time of this report, the skin lesions are resolving.

Discussion: Sarcoidosis

Our patient demonstrated a mild form of sarcoidosis which affects one-quarter to one-third of patients with this disorder; they often have with constitutional symptoms that may include cough, dyspnea, arthralgias, uveitis and a variety of skin lesions. A further one-quarter of patients, especially those on the Mainland, are asymptomatic, the disease having been discovered on routine chest X-rays. In our opinion, this subset may well be even larger, since many so afflicted obviously never visit a physician because they have no constitutional complaints.

The remainder of patients have a more insidious onset that develops over a period of months. This group is more likely to develop chronic disease with subsequent severe pulmonary and other organ damage.

It is quite likely that our patient was exposed to the putative agent (or agents) of sarcoid before moving to Hawaii, and that initiated sarcoidosis after a latent period of at least 13 years. Speculation concerning this activation process could center on poorly understood immunologic and/or genetic mechanisms. A further unknown factor acting as a hapten may have aided in the recognition of the dormant antigen by the patient's reticuloendothelial system. Or, it is possible that this new factor — or even the offending agent itself — was genetically transcribed, its coding system having been unlocked by an unknown environmental stimulus.

Epidemiology

The highest incidence of sarcoidosis is found in northern Europe and in North America. Sweden has the highest incidence of any country, at 64/100,000; however, according to autopsy studies, the true incidence may be 10 times higher[1].The incidence in the U.S. is 11 to 40/100,000, the highest being among Blacks in the Southeast. Parts of Africa and Japan also show a relatively high incidence. Sarcoidosis is rare in southern Europe and in southeast Asia.

In the U.S., Blacks are affected 10 to 17 times more often than Whites. Black women are affected twice as often as Black men, but the sex ratio in Whites is approximately equal. The disease is most common in the 3rd and 4th decades of

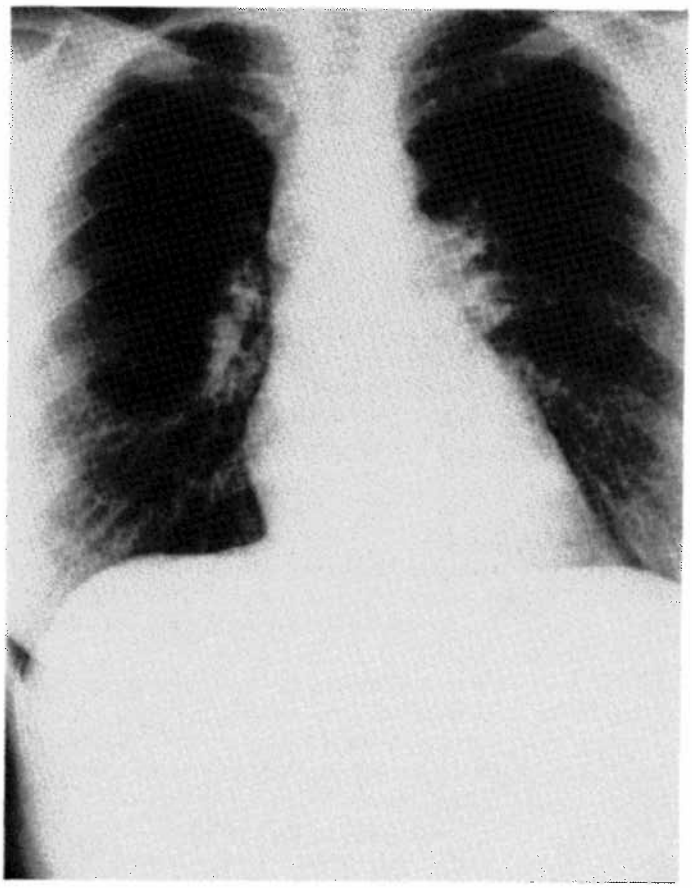

Fig. 2 — Left hilar adenopathy with possible right hilar adenopathy. Chest X-ray, 1990.

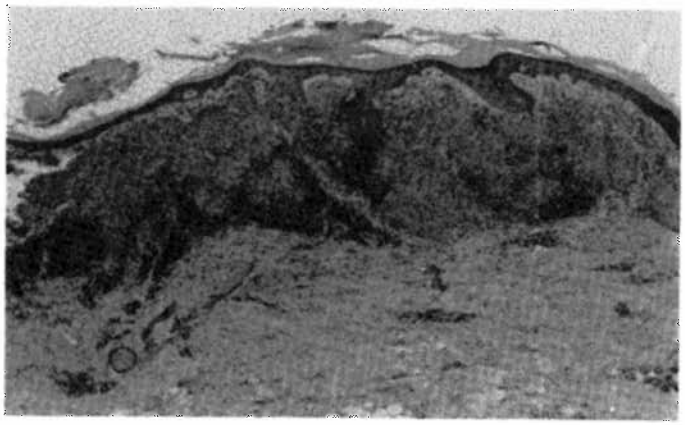

Fig. 3 — Skin biopsy demonstrating granulomas in upper dermis. Low power. H & E stain.

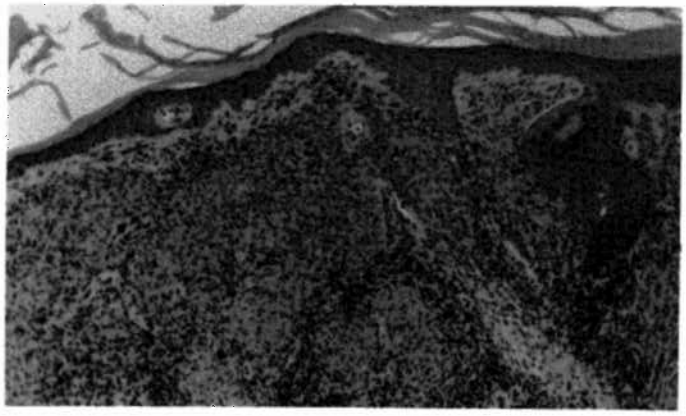

Fig. 4 — Skin biopsy. Mid power. H & E stain.

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life. An increasing number of familial cases are being noted, most frequently brother and sister[1].

Etiology and pathogenesis

Extensive studies so far have not elucidated an etiologic agent for sarcoidosis[1]. Many agents have been postulated as to the cause. These include Mycobacterium tuberculosis, Mycobacterium leprae, atypical Mycobacteria, a variety of viruses, fungi, inorganic agents and chemicals; however, the data are inconclusive.

Whatever the inciting event, the histopathogenesis of sarcoidosis suggests an immunologic basis[2]. It is postulated that an unknown antigen (or antigens) is recognized by a macrophage/histiocyte, which then stimulates the proliferation of helper T4 lymphocytes, that in turn stimulate B-lymphocytes to proliferate. The end-result is a granuloma composed of epithelioid histiocytes bordered by T-4 lymphocytes.

Clinical manifestations

Ninety percent of patients with sarcoidosis have an abnormal chest X-ray but only 5 to 15% develop serious pulmonary disease[2]. Overall, approximately 50% of patients have respiratory symptoms such as a dry cough, chest pain, shortness of breath, rarely hemoptysis. Less than 2% develop a pneumothorax. Intraparenchymal pulmonary granulomas have a predilection for the peribronchial, subpleural and interlobular septal connective tissues. Progressive disease with fibrosis is an interstitial process.

The skin is involved in 20 to 35% of cases[4]. A great variety of skin lesions have been associated with sarcoidosis; the most common are plaques, maculopapular eruptions, subcutaneous nodules, erythema nodosum and lupus pernio. The first 3 categories are ubiquitous in distribution. The papular form is most frequently located in the periorbital region, where it may simulate granulomatous rosacea. Erythema nodosum occurs in up to 25% of patients and may be the initial symptom. It is most commonly located on the anterior aspects of the lower legs and presents as tender, erythematous, or violaceous nodules. Most patients with sarcoid and erythema nodosum have a shortened disease course, with early spontaneous resolution. Those with Lofgren's Syndrome (a complex of erythema nodosum, bilateral hilar adenopathy, together with possible joint symptoms and uveitis) have an excellent prognosis. In one study, cases with erythema nodosum, acute arthritis and hilar adenopathy resulted in complete resolution of sarcoidosis in 85%, 83% and 73% of patients, respectively[5]. Sarcoid skin lesions have been reported in skin scars (including keloids) and in skin damaged by trauma, infection and radiation.

The eye is involved in 25 to 50% of patients with systemic disease[4]. The most common symptoms are blurred vision, tearing, and photophobia. Granulomatous uveitis is the most common eye lesion; approximately 75% of these patients have anterior uveitis and the remainder have posterior uveitis. The clinical course may be acute or chronic. Heerfordt's Syndrome (uveoparotid fever) consists of uveitis, parotid gland enlargement and facial nerve palsy. Conjunctival involvement is the second most common ocular finding and can lead to keratoconjunctivitis sicca. Eye involvement can be serious, leading to adhesions between the iris and lens, cataract formation, glaucoma and blindness.

The peripheral lymph nodes are involved in 50 to 75% of patients; most commonly the cervical, axillary, epitrochlear and inguinal nodes[3]. It is important to remember that sarcoid-like granulomas may occur in lymph nodes that drain a nearby malignant tumor. The spleen can be involved in up to 60% of patients, but symptoms occur only in the 15% of patients who demonstrate splenomegaly[3].

Liver biopsies have indicated involvement in 60 to 90% of patients[2]. The periportal regions are the most often affected by the granulomas. Cholestasis is rare, yet mild elevations of liver enzymes (alkaline phosphatase and serum bilirubin) are not uncommon.

Neurologic findings are present in 5% of patients[3]. Space-occupying granulomatous lesions of the cerebrum may cause confusion, seizure activity, hydrocephalus or hemiparesis. These mass lesions are most common at the base of the cerebrum and can elicit symptoms and signs related to derangement of the hypothalamic-pituitary axis. Chronic aseptic granulomatous meningitis has been recorded. Cranial and peripheral nerves are frequently involved, whereas spinal cord involvement is rare.

Cardiac symptoms are present in 5% of patients[1]. At autopsy, 27% of patients with systemic sarcoidosis have cardiac granulomatous lesions. The most common cardiac symptom is an arrhythmia caused by granulomas in the septum and ventricles. Sudden cardiac death may occur, and at autopsy these patients are found to have massive fibrosis and granulomatous involvement of the myocardium.

Kidney involvement is rare. Renal disease is usually due to an alteration in calcium metabolism, not to granuloma formation.

Sarcoid granulomas of the bone marrow have been reported in 15 to 40% of cases, although their effects on hematopoiesis are minimal[2].

Diagnosis

The diagnosis of sarcoidosis is usually based on the patient's clinical presentation, the primary ancillary test being a chest X-ray which, in at least 25% of patients, may be the only evidence of disease. The classic findings of Lofgren's Syndrome are adequate for a diagnosis of sarcoidosis[4]. Of utmost importance is the demonstration of noncaseating granulomas which must be differentiated from the granulomas of tuberculoid leprosy and foreign body reaction. Biopsies of skin or accessible lymph nodes are the most obvious choice to verify the existence of granulomata. If these sites are not revealing, bronchoscopic biopsies may be considered. Ophthalmologic examination and pulmonary function tests should be performed on all patients. The traditional Kveim test is rarely performed at present because the extract is not available and it lacks specificity.

ACE may be elevated. Produced by the epithelioid histiocytes of the granulomas, it is not a specific marker for sarcoidosis and is elevated in many of the diseases that fall within the differential diagnosis of sarcoidosis[2,6]. Sarcoidosis is a well-known cause of hypercalcemia.

Two specialized tests include broncho-alveolar lavage and the Gallium-67 lung scan. These are used in centers that regularly see patients with sarcoidosis. Lavage is done to ascertain the preponderance of lymphocytes, the majority of which are T-4[1]. This may be of aid in differentiating sarcoidosis from

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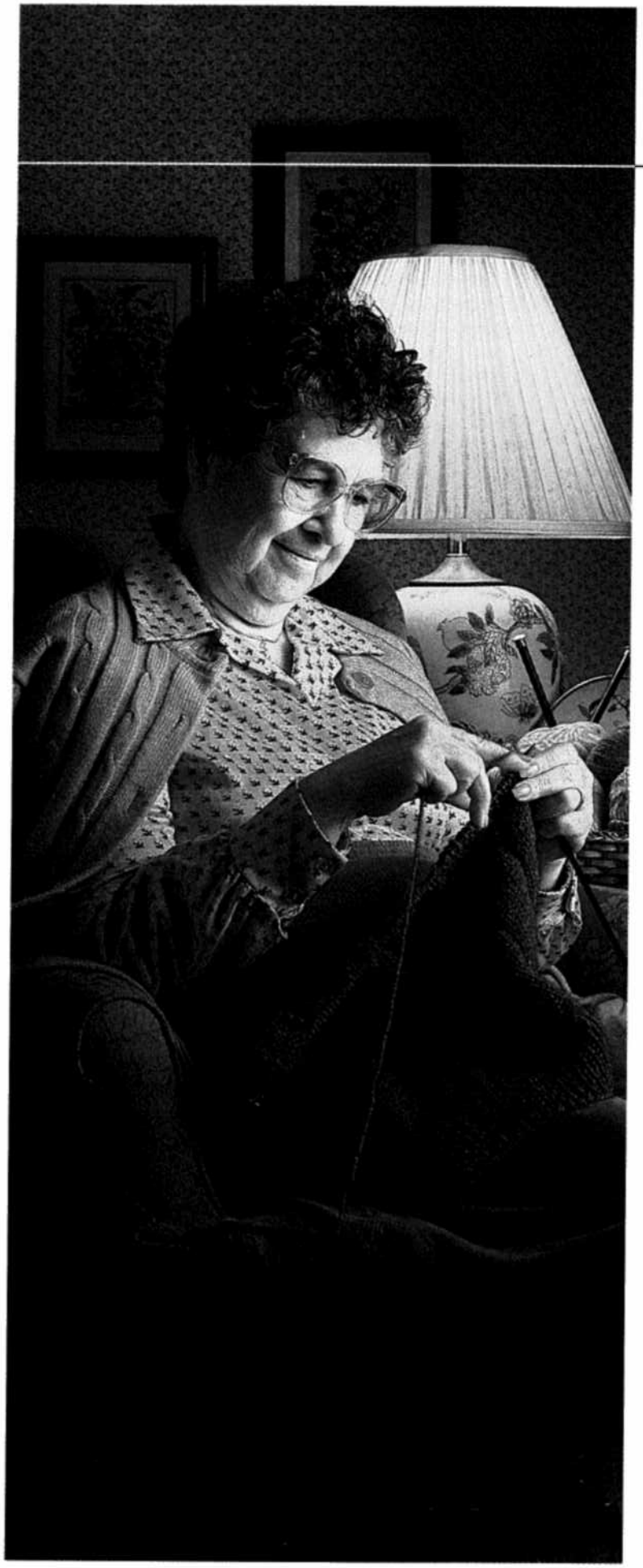

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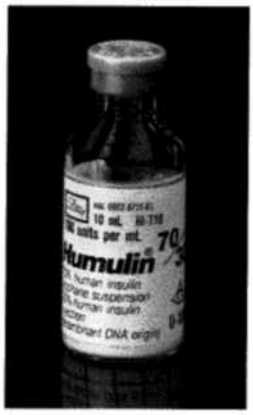

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allergic alveolitis in which there is also a marked increase in the lymphocyte population; however, these are T-8 (suppressor) lymphocytes[3]. The Gallium-67 scan will often display a diffuse uptake consistent with the interstitial disease process of sarcoidosis. However, the scan is not specific; it is expensive and exposes the patient to a significant amount of radiation[3].

Differential diagnosis

The early chest X-ray changes of sarcoid can mimic tuberculosis, lymphoma and metastatic carcinoma, as well as systemic fungal diseases, most notably histoplasmosis and coccidiodomycosis. More advanced lung disease demonstrates changes on X-ray similar to that of extrinsic allergic alveolitis, fibrosing alveolitis, histiocytosis, systemic lupus erythematosus, rheumatoid lung, scleroderma and tuberculosis. On chest X-ray the most advanced disease can be confused with tuberculosis, bullous emphysema, bronchiectasis and melioidosis[6]. Furthermore, berylliosis, asbestosis, the diseases caused by silica, talc, mineral oil, and certain drugs (methotrexate, cromolyn sodium) can produce granulomatous lung disease and evoke a granulomatous response in lymph nodes but rarely in the skin.

Certain infectious diseases, apart from those already enumerated, are likely to provoke a granulomatous response in lymph nodes alone. These include toxoplasmosis, infectious mononucleosis, brucellosis, chlamydia, pneumocystis carinii, tularemia, various fungi, all stages of syphilis, the fairly early stages of cat scratch disease and lymphogranuloma venereum. Granulomas may arise in the bowel and mesenteric lymph nodes in Crohn's Disease, Whipple's Disease, and infection caused by Yersinia enterocolitica. Malignant tumors may evoke a sarcoid-like response in adjacent lymph nodes.

Many of the aforementioned entities can also produce granulomas in the skin. Further causes of skin granulomas include insect bites, delayed hypersensitivity reactions, leprosy, acne rosacea, granuloma annulare, tertiary syphilis, and pricks by sea urchins and by plants with spikes.

Sarcoid arthritis may be confused with gout, acute rheumatic fever, tuberculosis, Yersinia enterocolitica arthritis, and rarely rheumatoid arthritis.

Treatment and prognosis

Treatment is initiated depending on the severity of systemic or organ disease[1,3]. Therapy is not required for patients who present with an incidental bilateral hilar adenopathy on chest X-ray but are otherwise asyptomatic. Likewise, those presenting with an acute onset with some or all of the manifestations of Lofgren's Syndrome rarely require treatment because 80% will experience spontaneous resolution in any case.

Of all organ systems, the lung is the most often involved and the disease activity must be assessed fully. In most cases, clinical examination, pulmonary function tests, and chest X-ray will be sufficient. The chest X-ray findings are classically divided into 5 grades: Grade O - absence of abnormal radiographic findings; Grade I - bilateral hilar adenopathy without involvement of lung parenchyma; Grade II - bilateral hilar adenopathy and parenchymal infiltration; Grade III - widespread parenchymal infiltration without hilar adenopathy; Grade IV - irreversible fibrosis with the formation of bullae[1]. Treatment must be initiated in patients with Grade II disease if they are symptomatic or if more specialized studies (Gallium-67 scan or broncho-alveolar lavage) in asymptomatic patients establish an increase in the activity of the disease process.

Corticosteroids are the drug of choice for most patients with pulmonary and extrapulmonary disease (uveitis, cardiac arrhythmias, neurologic lesions and arthritis). Chloroquine and hydroxychloroquine are occasionally efficacious in the various skin manifestations associated with sarcoidosis, in particular lupus pernio. Interestingly, both corticosteroids and chloroquine tend to lower high serum calcium levels by inhibiting the action of active Vitamin D on the gut and also by inactivating the compound itself. This in turn will reduce the urinary excretion of calcium and prevent stone formation.

The prognosis in sarcoidosis is good. However, in perhaps 15% of patients the disease follows a chronic, persistent or recurrent course. Death directly related to sarcoidosis occurs in up to 6% of patients[4].

Conclusion

Sarcoidosis, a common disease in the U.S. and various parts of Europe, is very rarely encountered in Hawaii. Most physicians here have never seen a case in our state. Diagnosis may be difficult, and in fact may not even be considered. Both tuberculosis and leprosy can present with granulomatous skin lesions remarkably similar to sarcoidosis[2,7,8].

Skin involvement by either tuberculoid leprosy or sarcoidosis often cannot be differentiated, clinically or histologically one from the other. However, lepromatous leprosy usually presents with few or no granulomata, and macrophages tend to be packed with acid fast mycobacteria. Even so, the skin lesions of lepromatous leprosy can be difficult to differentiate clinically from sarcoidosis because they tend to present in the form of symmetric and bilateral plaques, nodules, or papules; erythema nodosum-like lesions may appear during reactional states[8]. Hypaesthesia is usually present in the skin lesions of tuberculoid leprosy yet not in those of sarcoidosis. Additional problems are encountered in distinguishing the 3 intermediate forms of leprosy: Borderline tuberculoid, borderline, and borderline lepromatous leprosy. In these situations, both the clinical and histologic appearance of the involved skin may be an exact replica of sarcoidosis.

In summary, we have described the first case of sarcoidosis to be reported from Hawaii. Undoubtedly, others have been seen but have not been documented in the medical literature. With the ever-increasing influx of residents to Hawaii from endemic areas, more cases of sarcoidosis are bound to be seen by physicians in the state.

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■

Maka O Ka Kauka

Russell T. Stodd, MD

If things are going well, you have overlooked something.

In a recent prospective study conducted at New York Eye and Ear Infirmary, investigators demonstrated that the incidence of culture-growth acute postoperative endopthalmitis was reduced markedly by using a surgery preparation of povidone-iodine 5%. Nearly 8,000 patients were included in an 11-month study in which the povidone-iodine group had one-fourth the infection rate of that seen in a control population. Mark Speaker MD, stated that no significant complications have been seen among the more than 15,000 patients in whom it has been used.

"Academy politics is the most vicious and bitter form of politics, because the stakes are so low."

While the American Academy of Ophthalmology oligarchy has never (to my knowledge) been particularly enamored with the concept of one man-one vote, the parliamentary maneuvers in regard to 1990 elections of the Board of Directors were alarmingly odious. Hard working Oliver Dabezies MD, of New Orleans, intelligent, courteous and respectful, was nominated by petition since the Board and its nominating committee chose not to place his name on the ballot. However, when the time came to organize the ballot, the Board ruled that positions were "slotted," and that Dr Dabezies would have to select a Board nominee to challenge, a maneuver the doctor did not expect and certainly did not desire. He had reason to expect that the election of Directors would be an open one with the various candidates achieving office based on general competition, much as occurs when the AMA House of Delegates selects its officers. He considered withdrawing his name, but felt a duty to those members who had petitioned him to run (myself included). Hence, Dr Dabezies was obliged to make a choice as to which Board nominee he would "challenge." Of course, he lost the election; and furthermore, he was criticized because of the Board nominee that he selected to challenge. Our Academy leadership, in its obligation to be egalitarian, will put off change as long as possible.

(Continued on page 213) ➤

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MAKA O KA KAUKA

(Continued from page 211)

A man should be greater than some of his parts.

Ironic indeed, that the man who directed his home state into a financial disaster, tied medical licensure to Medicare participation, and endorsed retroactive assessment of medical liability premiums in support of insurance carrier malfeasance, should be invited by the University of Hawaii to deliver a series of lectures on a national health policy. Former Governor Dukakis should come here to learn, not to instruct! Hawaii is far ahead of all other states in providing access to health care. Of course, the invitation to the former Presidential candidate was at the behest of our Governor, who seems to remain charmed by the New England loser. What subtle political design is at work here?

Good words are worth much, and cost little.

A recent PBS broadcast carried a 1-hour production of "cataract surgery" performed by Wills Eye Hospital in Philadelphia. The video presented an excellent demonstration of microsurgery, using phakoemulsification and a flexible lens implant. The surgeon, Raymond Adams, and department chairman William Tasman, emphasized various techniques and implants that are available, all yielding very good visual results. Included were careful patient education with a comprehensive anatomic and surgical discussion, a plan of pre- and post-op care, and a description of cataract visual impairment. At Wills, there are 6 on the surgical team with surgeon and ophthalmologist assistant, anesthesiologist and nurse anesthetist, plus scrub and circulating nurses. The sum was an apt model to establish that cataract surgery is a safe, easy, productive experience for the patient, but requires a skilled team and considerable technology.

Friends may come and go, but enemies accumulate.

Once again the HHS Inspector General's office is threatening action in regard to practitioners who routinely waive the copayment. Under recent guidelines, Part B practitioners and suppliers who fail to undertake a "reasonable" copayment collection effort are at

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MAKA O KA KAUKA

(Continued from page 213)

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2. Warn that unjustified waivers could precipitate a government suit under the Federal False Claims Act — and triple-damage and penalty provisions can transform a modest amount of incorrect charges into hefty judgments.
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Addenda —

— Henry William, an American ophthalmologist, was the first to use a corneoscleral suture to close a cataract wound, in 1865.

— Americans spend $1,369,863 on laxatives and $3,673,973 on vitamins *each day!*

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rts

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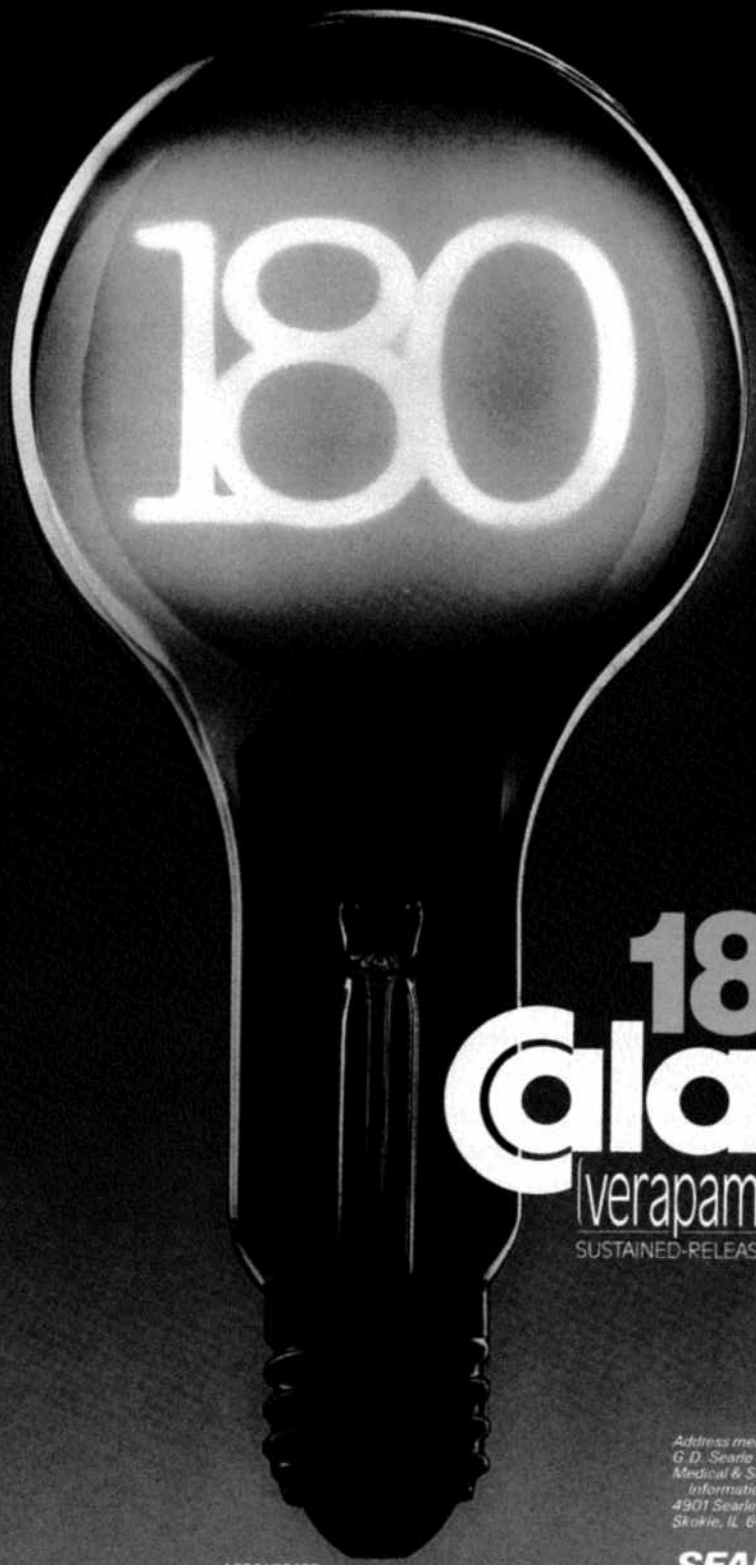

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