CASE REPORTS

RARE MANIFESTATIONS OF SARCOIDOSIS

James G. McPherson, III, MD, MPH, and Chin Bor Yeoh, MD
New York, New York

This case report describes a patient with sarcoidosis who developed superior vena cava syndrome, breast granuloma, and tracheobronchial stenosis, all as a direct result of the primary underlying disease. While each of these entities has been described individually as being associated with sarcoid, this is the first case in which all have occurred in the same patient. (*J Natl Med Assoc.* 1993;85:869-872.)

Key words • sarcoidosis • superior vena cava syndrome • breast granuloma • tracheobronchial stenosis

Sarcoidosis is 10 times more prevalent among American blacks than whites and most commonly affects the pulmonary system. The disease is primarily responsible for an interstitial lung disease in which the inflammatory process involves the alveoli, small bronchi, and blood vessels.[1] However, sarcoidosis can affect all of the body's organ systems, including the skin, eyes, bone marrow, liver, spleen, and heart. This article reports a patient who developed several rare sequelae of sarcoidosis. This patient was diagnosed with sarcoidosis and subsequently developed superior vena cava syndrome, breast granuloma, and tracheobronchial stenosis, all due to the disease. The superior vena cava syndrome responded favorably to daily steroid therapy, but she succumbed to pulmonary complications of her disease 13 years after her initial presentation. Although there have been numerous reports of individual sequelae of sarcoidosis, there have been no reported cases of superior vena cava syndrome, breast granuloma, and tracheobronchial stenosis all due to sarcoid in the same patient.

From the Department of Surgery, Section of Thoracic Surgery, St Luke's-Roosevelt Hospital Center, New York, New York. Requests for reprints should be addressed to Dr James G. McPherson, III, Dept of Surgery, Section of Thoracic Surgery, St Luke's-Roosevelt Hospital Ctr, 428 W 59th St, New York, NY 10019.

CASE REPORT

In December of 1972, a 64-year-old African-American woman was admitted to St Luke's-Roosevelt Hospital Center following a single acute episode of hemoptysis in which she expectorated approximately 50 mL of bright red blood. She had been plagued for 10 years prior to her admission by a progressively worsening skin condition involving her scalp axillae and both lower extremities. She underwent a complete dermatologic evaluation, including a positive Kveim test, and was diagnosed with Boeck's sarcoidosis. Four months prior to this admission, the patient developed chest wheezes of mild to moderate severity. She was started on oral prednisone, which provided partial relief of these symptoms. One month prior to this admission, she developed marked swelling of the face and neck, as well as distention of the veins of the face, neck, and anterior chest wall. She reported gradual regression of these symptoms after treatment with antihistamines. She denied any history of cardiac disease, hypertension, or diabetes mellitus. Earlier tests for tuberculosis and systemic lupus erythematosis were negative. The patient also denied travel outside of New York City and exposure to animals of known allergies. She had no history of tobacco use.

On physical examination, the patient was a well-developed, nourished female with a blood pressure of 110/70 mm Hg, a heart rate of 110 beats/minute and regular, and a respiratory rate of 24 breaths/minutes. Inspection of the skin revealed multiple, diffuse, erythematous and excoriated lesions over the scalp with patchy alopecia. Several nontender macular lesions were present on the anterior left thigh and shin. In addition, raised, linear scarred areas were present in

both axillae. There was no appreciable lymphadenopathy. Examination of the head and neck was notable for marked distention of the external jugular veins up to the angle of the mandible at 90°. Smaller cervical veins also were dilated. In adddition, generalized facial swelling was present with mild periorbital edema. Examination of the lungs was remarkable for bilateral, diffuse expiratory wheezes. The patient also had decreased breath sounds and dullness to percussion over the entire left hemithorax. The remainder of the physical examination was essentially normal.

Results of the complete blood count on admission revealed a hematocrit of 41.7% and a white blood cell count of 6100 mm^3 of which 85% were polymorphonuclear leukocytes, 14% were lymphocytes, and 1% were eosinophils. Electrolyte panels were within normal limits. Serologic tests for rheumatoid latex factor were positive. An electrocardiogram demonstrated nonspecific ST segment depression in the lateral precordial leads. Roentgenograms of the chest showed homogeneous opacification over the left lower lung field. There was no widening of the mediastinum. The heart appeared to be of normal size.

The patient was admitted with a diagnosis of superior vena cava syndrome resulting from a presumed intrathoracic neoplasm, possibly of pulmonary origin. Cytological and bacteriological studies of the sputum were performed but were negative for aerobic, anaerobic, fungal and acid-fast organisms. Bronchoscopy was performed on the eighth hospital day and revealed erosion and inflammation of the distal trachea as well as distortion and widening of the carina. Both mainstem bronchi were narrowed, and bright red blood emanated from the right mainstem bronchus. The bronchoscope could not be advanced beyond the right upper lobe bronchus. No discrete endobronchial lesions were identified.

A pleural biopsy and aspiration of pleural fluid were performed on the ninth hospital day. Approximately 125 mL of pale yellow fluid was extracted from the left chest along with several fragments of pleura using the Cope biopsy needle. The fluid was negative for bacterial, fungal, or acid-fast organisms. No malignant cells were identified. Examination of the pleura revealed only chronic inflammation.

A superior vena cavagram demonstrated complete obstruction of the superior vena cava. A scalene node biopsy revealed a single noncaseating granuloma with giant cells consistent with sarcoidosis.

A liver biopsy was negative for malignancy. With no apparent malignancy, sarcoidosis emerged as the underlying cause of the patient's caval obstruction. She was placed on oral prednisone, to which she responded favorably. After complete resolution of her symptoms, she was discharged on 60 mg of prednisone daily.

The patient remained in good health until July of 1977 when she was admitted with a chief complaint of right posterior chest pain of 24 hours duration. Physical examination revealed bilateral and equal breath sounds, and her electrocardiogram showed no evidence of acute changes. An arterial blood gas drawn in the emergency room revealed a PO_2 of 60 mm Hg. Pulmonary arteriograms showed complete cutoff of the left main pulmonary artery. This finding was confirmed by a lung scan, which demonstrated failure of perfusion of the left lung.

She was diagnosed with acute pulmonary embolism. She was placed on heparin therapy, but after 10 days she developed tarry guiac positive stools. Her hematocrit during this period dropped from 32% to 23%. Heparin was then discontinued, and she was started on oral iron therapy. Her hematocrit rose gradually over the next several days without transfusion therapy. She was subsequently discharged in good condition.

In February of 1980, the patient presented for elective excisional biopsy of a firm mass in her left breast, which had been present for approximately 1 year. On examination, a 3×3 cm firm mass was palpated in the upper outer quadrant of her left breast. There was no peau d'orange, nipple discharge or inversion, or axillary lymphadenopathy. She was taken to the operating room where a left subtotal mastectomy was performed. Pathologic examination of the biopsy specimen revealed noncaseating granulomatous inflammation of the breast consistent with sarcoidosis. She was discharged after an unremarkable postoperative course.

The fourth hospital admission for this patient came after she developed an ulcer on her lower left leg. Although she had no history of venous stasis disease or lower extremity edema, this ulcer grew progressively larger. She was evaluated as an outpatient and treated with daily dressing changes without result. A biopsy of the edge of the ulcer again revealed noncaseating granulomatous changes consistent with sarcoidosis. She was treated with enzymatic debridement and eventually received a split thickness skin graft. Following management of her ulcer, she was discharged from the hospital.

Finally, in December of 1985, the patient presented to the emergency room in acute respiratory distress with diffuse, bilateral wheezes and dyspnea. She was admitted to the intensive care unit where she required

endotracheal intubation, mechanical ventilation, and subsequent tracheostomy. Bronchoscopy revealed obstruction of the trachea and mainstem bronchi by a large granulomatous mass. Biopsy of this mass showed a granuloma consistent with sarcoidosis. She died suddenly from massive pulmonary hemorrhage, probably secondary to granulomatous erosion into a major pulmonary vessel. Postmortem evaluation was denied by the patient's family.

DISCUSSION

Sarcoidosis is a rare cause of superior vena cava syndrome. In the United States, the vast majority of cases of superior vena cava syndrome are due to some form of malignant disease. The most common malignant neoplasms causing superior vena cava syndrome are bronchogenic carcinoma and lymphoma. Superior vena cava syndrome resulting from benign neoplasm or granulomatous disease is generally associated with a good prognosis.[2] Five cases of superior vena cava obstruction caused by sarcoidosis have been reported in the literature.[2-6] In each of these cases, superior vena caval obstruction was the first manifestation of sarcoidosis.

In two cases, the signs and symptoms of caval obstruction responded favorably to steroid therapy.[3,4] The third case received no steroid therapy, but experienced remission of symptoms gradually over a 3-year period.[5] The fourth patient had coexisting mitral stenosis and died from sepsis 4 weeks after commissurotomy.[6] The fifth patient died of cardiogenic shock and was found at autopsy to have superior vena caval obstruction due to enlarged lymph nodes as well as extensive sarcoid involvement of the heart.[2] Although our patient was diagnosed with sarcoidosis prior to the onset of her superior vena cava syndrome, she was effectively managed with steroid therapy.

Mammary sarcoidosis is another infrequent manifestation of sarcoidosis, with 20 cases being reported in the literature.[7-22] Of these 20 cases, the diagnosis was made on the basis of histological examination in only 10.[22] These lesions usually present as a firm, mobile, nontender mass mimicking a fibroadenoma. However, they also have presented as tender masses adherent to the overlying skin, giving the appearance of a mammary carcinoma.[8,17] Accurate diagnosis, as well as definitive therapy, are achieved by excisional biopsy.

Of the three rare sequelae of sarcoidosis mentioned in this report, bronchial stenosis occurs with the greatest frequency. One study using flexible fiberoptic bronchoscopy in 101 patients with sarcoidosis demonstrated that 26% had bronchostenosis.[23] Steroid therapy has proven effective for mild to moderate cases. Fiberoptic balloon dilatation has been performed in patients with segmental stenosis of the mainstem bronchus with excellent results.[24]

Our patient is notable in that she developed three uncommon sequelae of sarcoidosis. This is the first patient in the literature to have this constellation of findings. Although a histologic examination of the mediastinal mass was not performed, the lymph node biopsy showing a noncaseating granuloma strongly suggests that sarcoidosis was responsible for the caval obstruction.

CONCLUSION

Sarcoidosis should be considered among the more common causes of superior vena cava syndrome, breast granuloma, and bronchial stenosis, particularly in patients who are already known to have the disease.

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