Sinus Histiocytosis Presenting as a Mediastinal Mass*

Mark Jay Friedman, M.D.; Len J. Rossoff, M.D.; Belinde Aftalion, M.D.; Arfa Khan, M.D.; Robert Decker, M.D.; and Harry Steinberg, M.D.

A 32-year-old man with a ten-year history of bilateral cervical adenopathy, parotid gland swelling, and a posterior mediastinal mass, presented with headaches. A CAT scan of the head revealed a suprasellar mass. Craniotomy and biopsy of this lesion led to a diagnosis of sinus histiocytosis.

This male patient had, for ten years, clinical findings suggestive of sarcoidosis—lacrimal and parotid gland swelling, cervical lymphadenopathy, posterior mediastinal adenopathy, and a suprasellar mass. Histopathologic findings of a brain biopsy in conjunction with a review of previous biopsy material were consistent with a diagnosis of sinus histiocytosis with massive lymphadenopathy.

REFERENCES


*From the Long Island Jewish-Hillside Medical Center, Division of Pulmonary Medicine and Critical Care, New Hyde Park, NY. Reprint requests: Dr. Steinberg, Long Island Jewish Hospital, New Hyde Park, New York 11042

FIGURE 1. Recent chest roentgenogram reveals a posterior mediastinal mass.

CASE REPORT

A 32-year-old black man presented to the neurosurgical service with a one-year history of headaches. Computed tomography of the head revealed a suprasellar mass. Significant past medical history included right nephrectomy in 1964 for "xanthogranuloma" believed to be a complication of chronic pyelonephritis. In 1971, biopsies were performed of bilateral painless cervical adenopathy and parotid gland swelling. Simultaneously, the patient underwent mediastinoscopy for a posterior mediastinal "mass." He was informed that he had an "inflammatory process" not consistent with sarcoid and received no treatment.

Physical examination on this admission revealed cervical adenopathy, parotid gland swelling, as well as lacrimal gland swelling. Chest and neurologic examinations were unremarkable. The admission roentgenogram is presented in Figure 1. The roentgenographic findings were those of a posterior mediastinal mass best seen superiorly on the right and as a double density behind the heart on the left. Surgical clips of the prior mediastinoscopy were also seen. An aortogram and bilateral oblique hilar tomography were negative. The patient underwent a craniotomy and biopsy of yellow plaque-

FIGURE 2. Meninges: Histiocytic and fibroblastic proliferation (hematoxylin-eosin, original magnification x 112). Inset: Erythrophagocytosis within histioocytes (hematoxylin-eosin, original magnification x 900).
like lesions at the base of the brain.

**DISCUSSION**

A case of sinus histiocytosis with massive lymphadenopathy is reported with a clinical history spanning ten years. The characteristic histologic findings of this disease were seen in the brain biopsy (Fig 2) and on review of previous renal and scane node biopsy specimens. Mediastinal specimens were unavailable. There was an infiltration of large bland histiocytes with phagocyted inflammatory cells (lymphocytes and plasma cells). Spaces, resembling the sinusoids of lymph nodes, were distended with these histiocytes, hence “sinus histiocytosis.”

This condition was first described in 1969 by Rosai and Dorfman1 who confirmed the histologic findings in this case. The majority of the patients present before age 20 characteristically with bilateral painless cervical adenopathy.6 Other lymph node regions frequently involved are axillary and mediastinum. More than a quarter of the patients have been reported to have extranodal disease most commonly involving the orbits and eyelids; skin;7,8 salivary glands;9 upper respiratory tract; and bones.10 This patient is believed to be only the ninth case with CNS involvement.8 Occasionally, only extranodal disease is manifest.

This is typically a chronic benign disease with occasional spontaneous remission. The etiology is unknown, and no organism has been isolated to date. There is no known treatment but benefits from steroids or chemotherapeutic agents have been reported.3

ACKNOWLEDGMENTS: The authors wish to thank Dr. Rahsa Mir for her assistance in reviewing the pathologic findings, and Mrs. Lois Hirsch for her secretarial assistance.

**REFERENCES**

1 Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: a newly recognized benign clinicopathological entity. Arch Pathol 1969; 87:63-70
2 Sanchez R, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: an analysis of 113 cases with special emphasis on its extranodal manifestations (abstract). Lab Invest 1977; 36:349-50
4 Foucar E, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: ear, nose, and throat manifestations. Arch Otolaryngol 1978; 104:887-93
6 Foucar E, Rosai J, Dorfman RF, Russell B. The neurologic manifestations of sinus histiocytosis with massive lymphadenopathy. Neurology 1982; 32:365-71

**Respiratory Augmentation of Left Ventricular Function during Spontaneous Ventilation in Severe Left Ventricular Failure by Grunting**

**An Auto-EPAP Effect**

**Michael R. Pinsky, M.D.,† George M. Matuschak, M.D.; and Jerome M. Itzkoff, M.D.**

A patient with severe congestive cardiomyopathy demonstrated involuntary forceful expiratory grunting that was associated with an elevated intrathoracic pressure and stable hemodynamics. Face-mask administration of 20 cm H2O as continuous positive-airway pressure (CPAP) abolished the grunting without hemodynamic compromise. However, loss of CPAP by mask leak resulted in rapid hemodynamic deterioration and resumption of grunting. Endotracheal intubation with 20 cm H2O of positive end-expiratory pressure restored hemodynamic stability. This suggests that spontaneous ventilatory efforts can augment the failing myocardium.

In patients with depressed left ventricular (LV) function and volume overload, cardiac output does not decline with the application of positive end-expiratory pressure (PEEP) and, in some cases, may increase compared with intermittent positive-pressure breathing (IPPB) alone. We have recently shown that this improvement in cardiac performance during positive-pressure breathing is secondary to a decrease in LV afterload resulting from the increase in intrathoracic pressure (ITP).5 Although cardiac augmentation during mechanical ventilation has been reported by many groups, the hemodynamic effects of increases in ITP during spontaneous ventilation have not.

We recently cared for a patient with severe congestive heart failure in whom increases in ITP during spontaneous ventilation generated by expiratory grunting were associated with improved cardiac performance.

**CASE REPORT**

A 28-year-old man was transferred to Presbyterian–University Hospital for possible cardiac transplantation because of severe progressive congestive cardiomyopathy of six weeks’ duration.

On admission, the patient was an obese man with moderate respiratory distress; he had a dusky coloration and prominent audible expiratory grunting. His blood pressure while receiving 7 μg/kg/min of dobutamine was 110/70 mm Hg. The pulse rate was 120/min, the temperature was 37.2°C, and the respiratory rate was 40/min. There was dullness to percussion at the lung bases bilaterally and diffuse end-inspiratory crackles. The jugular veins were distended to the angle of the jaw with the patient at 90°. The cardiac...