Eosinophilic granuloma of the cervicothoracic junction

Case report

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A case is reported of eosinophilic granuloma at the cervicothoracic junction presenting with profound quadriparesis preoperatively. The patient underwent excision via an anterior approach, with splitting of the sternum to gain access to the T-1 vertebra. Postoperatively, he has had an excellent return of function.

Key Words • eosinophilic granuloma • histiocytosis X • thoracic spine

Eosinophilic granuloma is part of a spectrum of diseases known as histiocytosis X. Granuloma of the bone and, in particular, of the vertebra has been reported, but rarely causes neurological deficit. We recently treated a patient with an eosinophilic granuloma involving the T-1 vertebra with a corresponding level of neurological deficit. This case is presented along with a review of the literature.

Case Report

This 10-year-old boy was admitted after the onset of back pain. He was hit in the back at school approximately 2 months prior to admission. Initially, he suffered back and shoulder pain and for a time was treated for a possible shoulder dislocation. The symptoms gradually progressed to pain radiating into both arms and to the mid-scapular region with respiration. Weakness of both legs developed and later he was unable to walk or stand. Computerized tomography (CT) scans and cervical laminograms showed destruction of the T-1 vertebral body. A course of corticosteroids was started, and there was marked improvement in his symptoms after several doses. His medical history was noncontributory.

Examination. The admission examination showed the patient's cranial nerves to be intact. Muscle strength was 4/5 throughout the left side, including the deltoids, biceps, and triceps. On the right side, strength was 5-/5, again including the deltoids, biceps, and triceps. His reflexes were 0 at the biceps and brachioradialis and 1+ at the ankles. Toe reflexes were upgoing bilaterally. He had ankle clonus on the left. The sensory examination was normal, except for hyperesthesia in the C6-7 distribution bilaterally. He had normal sacral sensation.

Plain spine x-ray films showed decreased height of the T-1 vertebral body. A CT scan showed destruction of the vertebral body, with bone and soft tissue pressing against the anterior aspect of the cord and impinging on the neural foramina bilaterally, more significantly on the left (Fig. 1 left). A bone scan showed increased uptake at the T-1 vertebral body, but no other lesions were noted. A magnetic resonance study showed destruction of the T-1 vertebral body with soft tissue impinging on the spinal cord (Fig. 2). Admission laboratory studies showed a normal white blood cell count and normal sedimentation rate.

Operation. The patient underwent surgery via an anterior approach to the cervicothoracic spine with a sternal splitting procedure combined with a left transverse cervical exposure. The T-1 vertebral body was exposed and a small amount of soft tissue was found and removed. A groove was drilled into the inferior surface of C-7 and into the superior aspect of T-2. A wedge-shaped bone graft was taken from the iliac crest and was placed between C-7 and T-2.

Postoperative Course. Initially, the patient was managed in a skull-occiput mandibular immobilization (SOMI) brace; however, he developed left C-8 radicular pain. Follow-up films showed the bone graft to be in good position. The patient was placed in a halo body cast and his radicular symptoms resolved within 3 weeks. He remained in the halo vest for 3 months, at which time CT scanning showed spinal fusion and
Eosinophilic granuloma of the spine

FIG. 1. Computerized tomography scans through the T-1 vertebral body. Left: Preoperative scan showing almost complete destruction of the vertebral body. There is extension of soft tissue into the canal and into the neural foramina. Right: Scan obtained 6 months after the operation showing the bone graft and a normally sized spinal canal.

FIG. 2. Magnetic resonance image of the cervicothoracic spine showing decreased height of the T-1 vertebra with a projection of soft tissue posteriorly impinging on the spinal cord.

revealed that the spinal canal was of normal size (Fig. 1 right). His symptoms have almost resolved with only slight spasticity of gait. Prior to discharge from the hospital, he underwent a radiographic bone survey which showed no other eosinophilic granulomas.

Discussion

Eosinophilic granuloma has been recognized as a discrete entity since 1940.¹⁰,¹⁵ This lesion is believed to be part of a spectrum of diseases including Letterer-Siwe disease and Hand-Schüller-Christian disease.⁶ These three entities are included under the broad category of histiocytosis X.⁹ The common histopathological hallmark of these three disease entities is the Langerhan's cell. Letterer-Siwe disease is an acute and fulminating disease affecting bone, viscera, and the reticuloendothelial system. It results in death of the patient in a matter of months. Hand-Schüller-Christian disease tends to be a more chronic, moderately disseminating disease. Eosinophilic granuloma as an entity tends to be a benign, self-limiting disease, localized to one or two lesions in bone without involvement of the viscera or the reticuloendothelial system.¹³ Eosinophilic granuloma represents 1% of all bone tumors and occurs in the vertebral column in 7% of the cases. It represents 60% to 80% of all forms of histiocytosis X, making solitary eosinophilic granuloma the commonest form of histiocytosis X.² It occurs with a male to female ratio of 1.5 to 2.5:1, but has no genetic predisposition. Current etiological theories suggest a defect in the immune system as the cause for disease, but no specific immunological defect has been documented.

The commonest clinical complaint of eosinophilic granuloma is local pain.¹²,¹⁴ The duration of symptoms prior to diagnosis generally ranges from a few weeks to months, often with a history of trauma. The earliest radiographic change is destruction of the center of the vertebra. This is not often seen since the lesion tends to progress to compression of the vertebra ("coin on edge"). The lesion may also be lytic,⁸ or it can undergo a regeneration, demonstrated by an increase in height of the involved vertebral body. On CT, vertebral destruction is evident, but there may also be soft tissue extending into the neural canal. A radionuclide bone scan is almost always positive, with increased uptake in the region of the eosinophilic granuloma. Laboratory evaluation may show elevation in the sedimentation rate or a mild leukocytosis.⁹⁷

Sheets of histiocytes mixed with eosinophils, plasma cells, and lymphocytes are seen microscopically. Langerhan's cell is essential for the diagnosis of histiocytosis X in general and eosinophilic granuloma in particular.
This cell is considered to be derived from the bone marrow and is of the mononuclear phagocyte family. Initially, the lesions consist mainly of histiocytes; however, with time the lesions can become difficult to differentiate from chronic osteomyelitis with its infiltrate of white blood cells. In children, collapse of the vertebral body (vertebra plana) is often considered pathognomonic of eosinophilic granuloma. Support for this conclusion is involvement of only one vertebra, preservation of the disc spaces, and consistent radiographic density of the collapsed vertebral body. Generally, the pedicles are spared and a paravertebral soft tissue mass is usually absent. However, this lesion must be differentiated from bacterial or fungal infections, or tumors such as neuroblastoma, Ewing’s sarcoma, chordoma, and leukemia. Occasionally, an aneurysmal bone cyst or hemangioma may present in a similar fashion. The most common location is in the thoracolumbar region, whereas the cervical region is rarely affected.

The goals of treatment for eosinophilic granuloma include spinal stability and preservation of neurological function. Some authors believe that if the granulomatous lesion progresses to vertebra plana without neurological deficit, then neurological deficit is unlikely to occur. In these instances, they recommend conservative treatment with biopsy and immobilization. There can be expansion of the compressed vertebra with conservative therapy. In the past, radiation therapy was recommended in low dosages (400 to 1500 rads); however, it is now considered unnecessary by most authors.

More controversy involves treatment of patients with neurological deficits. Some authors consider that immobilization with radiation therapy provides adequate treatment for these patients. A major disadvantage of this treatment is the impairment of growth secondary to radiation destruction of vertebral endplates in the pediatric spine. Others believe that the neural elements should be decompressed. If spinal instability occurs in the process of decompression, then fusion should be performed.

With the advent of MR imaging, the amount of neural compression in both symptomatic and asymptomatic patients can be readily seen, which aids management decision-making. Our patient had progressive neurological decline until he was started on a course of steroids. The compression of his neural elements included both bone and soft tissue. For these reasons, an anterior decompression and fusion were performed and the result has been excellent. Radiation therapy was not used.

References


Manuscript received April 19, 1989.
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