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Case report

Solitary eosinophilic granuloma of the sternum

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Abstract We report a rare case of eosinophilic granuloma of the sternum in a 25-year-old woman, who presented with anterior chest pain and a tender mass over the sternum. Total-body bone scintigraphy and computed tomography scanning of the thorax revealed an isolated lytic lesion of the manubrium. An open biopsy showed the typical histologic appearance of an eosinophilic granuloma. Surgical curettage of the solitary lesion was performed, and the sternal defect was filled with a bone replacement material. At the 2-year follow-up, no local recurrence was found, and the patient was in good health.

Key words Eosinophilic granuloma · Sternum

Introduction

Eosinophilic granuloma of bone was first independently described in 1940 by Otani and Ehrlich[9] and Lichtenstein and Jaffe.[5] In 1953 Lichtenstein[6] reviewed cases of eosinophilic granuloma of bone, Hand-Schüller-Christian disease, and Letterer-Siwe disease; because of an essential histologic similarity of these three disorders, he proposed the term "histiocytosis X" and treating the three disorders as a single malady. The pathologic feature common to the three disorders is parenchymal infiltration with large numbers of histiocytes. Langerhans cell histiocytosis (LCH), formerly known as histiocytosis X, is currently considered a disorder of immune regulation manifested by abnormal proliferation of histiocytes and granuloma formation. The presence of Langerhans cells, a unique histiocyte, is the distinctive pathologic component of the disease.[19] The disease may affect any organ, although the reticuloendothelial system (i.e., bones, skin, lymph nodes, liver, spleen) is mostly involved. Depending on the number of affected organs, two clinical types of disease (single system and multisystem) can be defined. Eosinophilic granuloma, the most common and mildest form, usually has a favorable outcome and can be either monostotic or polyostotic. Hand-Schüller-Christian disease is a chronic recurrent polyostotic condition, and Letterer-Siwe disease is known to be a more diffuse form of LCH.[15]

Eosinophilic granuloma usually occurs in children under the age of 10 years. The bones most commonly involved are the flat bones of the skull, pelvis, ribs, and scapula[20] as well as vertebra and diaphyses of long bones.[8] To date, only a few cases of solitary eosinophilic granuloma of the sternum have been reported,[3,4,7,10,14,16] although eosinophilic granulomas in other bones have been reported extensively in the literature. In the present study, an unusual presentation of solitary eosinophilic granuloma of the sternum is discussed.

Case report

A 25-year-old woman presented with anterior chest pain and swelling of 2 months' duration. There was no history of trauma. On physical examination the sternal mass was tender, was bony hard with a smooth surface, and measured 3.0 × 3.0 cm. Extensive laboratory tests (C-reactive protein 0.1 mg/dl, eosinophilic granulocytes 2.0%, alkaline phosphatase 126 IU/l) and plain radiographs of the chest were normal apart from a radiolucent osteolytic lesion localized in the sternal manubrium on the radiograph (Fig. 1). The clinical history revealed only mild bronchial asthma. Computed tomography (CT) confirmed the existence of a partially sclerotic, delineated osteolytic lesion that interrupted both anterior and posterior cortices of the manubrium (Fig. 2). Radiographic data did not indicate any signs of a lung lesion. Magnetic resonance imaging (MRI)

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Fig. 1. Radiograph of the sternum in the lateral view shows an osteolytic lesion in the manubrium (*arrow*)

Fig. 2. Computed tomographic (CT) scan shows the osteolytic lesion with interruption of the cortices in the manubrium of the sternum

showed a hypointensive signal area in the bone marrow on T1-weighted images (T1WI) that was hyperintensive on the T2-weighted image (T2WI). The mass-like lesion was seen as an area of signal intensity higher than that of the surrounding bone marrow on T2WI and as an extensive signal change with hyperintensity on T2WI in soft tissue areas. The MRI findings indicated a tumor mass spreading between the anterior and posterior sternal cortices and expanding to the soft tissue of the anterior chest wall; however, no invasion into the thoracic cavity was found (Fig. 3). Technetium bone scintigraphy revealed pathologically increased uptake of the isotope only in the manubrium (Fig. 4); there were no other lesions in closely located bones (ribs, clavicle, scapula). Thallium-201 scintigraphy showed almost normal accumulation of the tracer in the lesion during both early and late phases (Fig. 5). An open biopsy of the lesion was performed. Histopathologic examination revealed a mixed cellular infiltrate that consisted of eosinophilic granulocytes, lymphocytes, macrophages, and plasma cells. Immunohistochemical staining was markedly positive for S-100 proteins in the histiocyte cytoplasm (Fig. 6). Such a histology is quite characteristic of eosinophilic granuloma.

The patient underwent surgical curettage of the tumor. The excised tumor was soft, friable, and yellowish-white. Tumor invasion into parietal pleurae was not observed. After curettage, the sternal defect was filled with highly purified beta-tricalcium phosphate (β-TCP) (OSferion; Olympus Corporation, Tokyo, Japan) as bone replacement material. Pathologic examination confirmed the diagnosis of eosinophilic granuloma. Postoperatively, no chemotherapy or radiotherapy was carried out. Two years after the surgery, the grafted β-TCP had been bioresorbed and replaced by regenerated bone. No local recurrence has been found, and complete relief of pain has been obtained.

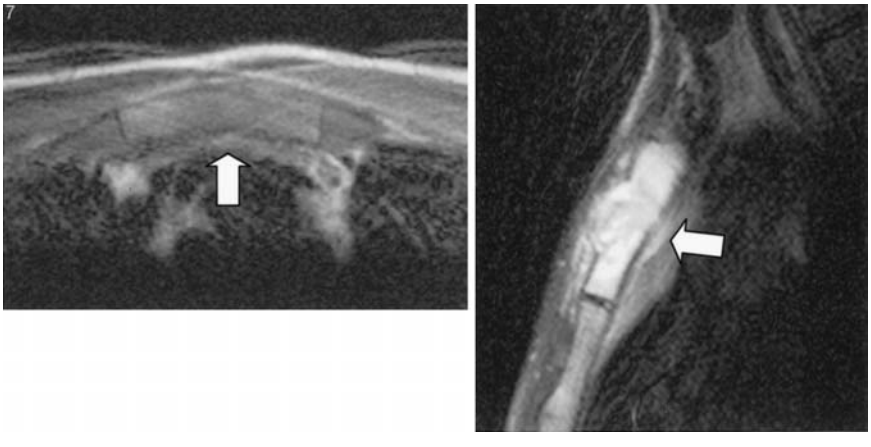

Fig. 3. Magnetic resonance (MR) images. **Left** The signal intensity of the tumor is slightly lower or almost identical (*arrow*) to that of the skeletal muscle on a transverse T1-weighted image. **Right** Sagittal T2-weighted image shows that signal intensity of the tumor is higher (*arrow*) than that of the skeletal muscle