

# Mediastinal Lymphangioma and Chylothorax: Thoracic Involvement of Gorham's Disease

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We report a case of mediastinal lymphangioma associated with Gorham's disease in a 38-year-old man who had suffered recurrent clavicular fractures during a seven-year period. Mediastinal widening associated with osteolysis of the clavicles and the sternal manubrium was revealed by chest radiography, while computed tomography demonstrated a cystic anterior mediastinal mass infiltrating mediastinal fat and associated with osseous destruction of the clavicles and manubrium. Chylothorax recurred during the course of the disease.

**G**orham's disease is a rare condition characterized by a non-malignant proliferation of vascular or lymphatic bone structures resulting in progressive bony destruction and often extending into surrounding soft tissues. Associated mediastinal lymphangioma has been very rarely reported. We present a case in which thoracic involvement of Gorham's disease was complicated by chylothorax in a patient with mediastinal lymphangioma.

## Index terms:

Computed tomography (CT)  
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## CASE REPORT

A 38-year-old man was admitted due to the sudden onset of anterior chest and neck pain. His medical history disclosed that during a seven-year period, repeated clavicular fractures had occurred after minor injuries.

Posteroanterior chest radiographs obtained at admission showed bilateral widening of the mediastinum and resorption of the proximal clavicles and the sternal manubrium (Fig. 1A). Postcontrast CT scans demonstrated a well-defined mediastinal mass of low attenuation, infiltrating mediastinal fat and enveloping mediastinal structures. Osteolysis of the clavicles and manubrium was noted (Fig. 1B). The lesion extended superiorly to the internal jugular chain, anteriorly to the pectoralis major and posteriorly to the paratracheal area, without displacing or compressing mediastinal structures. Lymph node enlargement was indefinite. Percutaneous fine needle aspiration of the lesion yielded 340 cc of wine-colored serous fluid; analysis of the aspirate revealed the presence of many lymphocytes and macrophages. There was no evidence of malignancy, and neither special staining nor subsequent culture indicated that microorganisms, including acid-fast bacilli and fungi, were present.

Six days after aspiration, incisional biopsy of the mediastinal mass was performed. It was found to be cystic in nature, and 150cc of turbid pinkish fluid was aspirated. Pathologic examination revealed cavernous lymphangioma (Fig. 1D), and on the basis of the radiologic and clinical features, Gorham's disease was diagnosed. Follow-up examination indicated that in spite of radiation therapy (total dose 3060 cGy), the extent

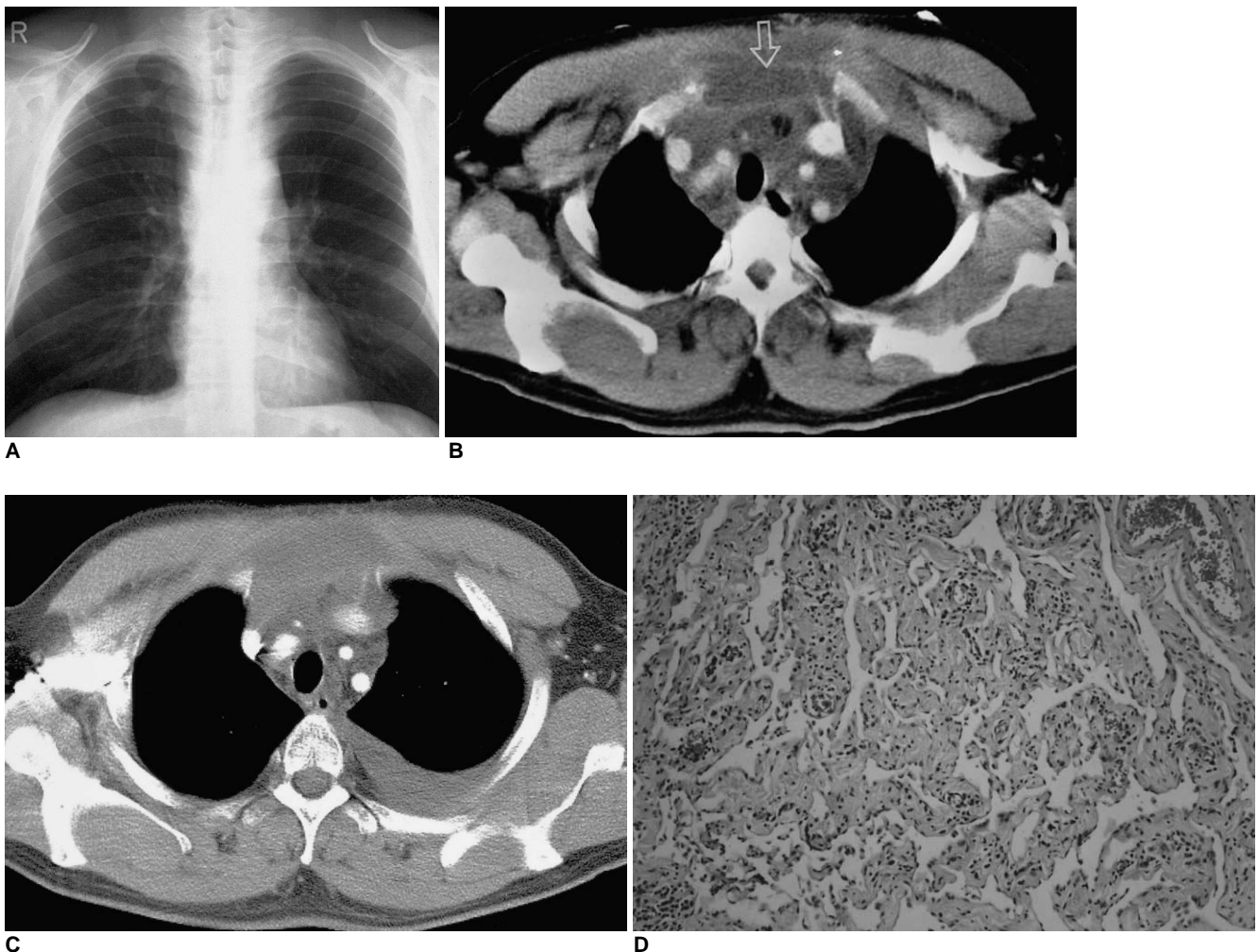
of the mass increased as bony destruction and pleural effusion progressed (Fig. 1C).

Recurrent chylothorax was managed by repeated aspiration and chest tube placement. Fifteen months after diagnosis, opacification of the lung, together with copious amounts of sputum, were observed. Bronchoscopy revealed an abundance of chylous fluid in the superior segmental bronchus of the left lower lobe, suggesting the presence of a bronchopleural fistula.

## DISCUSSION

The clinical course and radiographic findings of the case described are compatible with a diagnosis of Gorham's disease.

The condition, characterized by massive and progressive osteolysis, is rare: since Jackson, in 1838 (1), first described its presence in the humerus of a 13-year-old boy, less than 200 cases, worldwide, have been reported in the literature. The pathologic cause of Gorham's disease is a non-malignant proliferation of vascular or lymphatic tissue; the hemangiomatous or lymphangiomatous lesions which arise are progressive, without regeneration, and may extend to adjacent soft tissue or organs. Complete bony resorption, often affecting contiguous bones regardless of the joint involved, finally occurs. Radiographically, the case we describe clearly demonstrated progressive osteolysis without regard to joint space. The bones most frequently affected are the clavicle, scapula, humerus, ribs, and pelvis (2):



**Fig. 1.** Gorham's disease in a 38-year-old man with mediastinal lymphangioma.

**A.** Posteroanterior chest radiograph shows widening of the mediastinum and osteolysis of the proximal clavicles.

**B, C.** Contrast-enhanced CT scan demonstrates an anterior mediastinal mass of low attenuation, which infiltrates mediastinal fat and envelops adjacent structures. Lysis of the manubrium (arrow in **B**) is apparent and follow-up CT (**C**) after three months shows left pleural effusion.

**D.** Irregular-shaped dilated lymphatic spaces lined by attenuated endothelium are embedded in connective tissue stroma, and small lymphoid aggregates are dispersed in the stroma. These findings are compatible with cavernous lymphangioma (original magnification,  $\times 250$ ; hematoxylin-eosin staining).

widespread involvement of the skeleton, particularly of the spine and thorax, may result in neurological and pulmonary sequelae which are sometimes fatal (3, 4). The prognosis is unpredictable, and depending on the site and extent of bone involvement, ranges from minimal disability to death. Chylothorax, a rare and life threatening complication of Gorham's disease, may occur when the thoracic cage is affected, as in our patient, though less than 30 such cases have been reported in the literature in English (5). Chylothorax results from invasion of the thoracic duct or penetration of the pleural cavity by dysplastic lymphatic vessels (6). Medical management of the condition (drainage, parenteral feeding, cortico- or radiotherapy) is often ineffective, with a 64% mortality rate (5). For surgical treatment, consisting of ligation of the thoracic duct followed by pleurectomy, the reported mortality rate is lower, at 36% (5). In our case, osteolysis progressed in spite of radiation therapy, and the patient suffered recurrent chylothorax and pleural effusion.

Fewer than one percent of all lymphangiomas occur in the mediastinum, and are few reports have described mediastinal lymphangioma associated with Gorham's disease. A review of 14 cases of intrathoracic lymphangioma evaluated by Brown et al. (7) revealed this association in only one case, in which extensive lymphangiomatosis involved the mediastinum and pleural space, with destruction of several ribs.

In our case, the mediastinal lymphangioma appeared as a well-defined, low-attenuated homogenous mass infiltrating mediastinal fat. Despite its large size, it showed no mass ef-

fect on adjacent structures. These CT findings are identical to those described in several previous reports (8, 9).

Gorham's disease is very rare, and if a radiologist is unaware of its existence, may well be missed. A knowledge of this entity, on the other hand, will assist in differentiation of a progressive osteolytic lesion. When the thorax is involved, the possibility of chylothorax, a recurrent and sometimes life-threatening complication, should be borne in mind as a differential diagnosis of any pleural effusion.

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