

# Treatment of chylothorax in Gorham's disease: case report and literature review

Grant G. Miller, MD

A spontaneous chylothorax is rare in children outside the neonatal period. In the absence of trauma this condition may be associated with osteolytic bone lesions (Gorham's disease), but it does not occur often enough to be readily recognized. A chylothorax occurs in less than 1 in 5 cases of Gorham's disease and carries a high mortality. The management of an infant with chylothorax complicating Gorham's disease is reported.

## Case report

A 2-year-old boy presented with 3-day history of shortness of breath. A chest radiograph showed a large right-sided pleural effusion (Fig. 1). A large volume of "milky" fluid (protein 3.3 g/L, triglyceride 3.3 g/L and leukocyte count  $5.9 \times 10^9$  cells/L, with 90% lymphocytes) was aspirated. Over the next 48 hours, the chest tube drained approximately 3 L/d of chyle. Eliminating oral intake reduced the output to 1 L/d. Computed tomography of the chest did not reveal any specific disorder. Thoracoscopically, an area of inflammation that was weeping fluid was seen in the right costophrenic angle posteriorly. Biopsies of this area demonstrated an increase in the size, number and complexity of small lymphatic channels and large endothelial cells. After 10 days of unabated chyle loss, he underwent thoracic duct ligation and parietal pleurectomy of the involved area. In addition, the fibrin tissue adhesive Tisseel (Baxter Healthcare, Mississauga, Ont.) was applied to the affected area. The effusion resolved, and the chest

tube was removed 48 hours later. The following day a fluctuant swelling developed in the right lumbar subcutaneous tissues. Computed tomography (Fig. 2) revealed an isodense soft-tissue mass involving the right paralumbar musculature adjacent to the posterior elements of the vertebral column, extending from the level of the iliac crest to the T11 vertebral body. The vertebral bodies from L1-L5 demonstrated patchy osteopenia and lucent lesions with sclerotic rims.

Based on the clinical and radiologic findings the infant was considered to have Gorham's disease. His pleural effusion did not recur. He functioned well with normal growth and development and without obvious progression of his



FIG. 1. Chest radiograph from a 2-day-old neonate shows a large right pleural effusion.

disease. Two years after diagnosis he died suddenly of a massive retroperitoneal hemorrhage.

## Discussion

Gorham's disease<sup>1</sup> is a combined clinical, radiologic and histologic entity characterized by a nonfamilial, histologically benign endothelial proliferation in bone. It produces lysis of bone with or without involvement of the adjacent soft tissues. Approximately 17% of patients will have an associated chylothorax. The mechanism of the osteolysis is unknown and the biologic behaviour of the lesion cannot be predicted.

This condition may occur at any age but most commonly presents in childhood. There is no sex predilection or inheritance pattern. The cause is unknown. The disease has a propensity to involve the maxilla, shoulder girdle and pelvis. Involvement of the lumbar spine has only been reported in 3 other cases.<sup>2</sup> The bone lesions may be monocentric or polyostotic but are usually without skip areas. Signs and symptoms are usually related to the area of involvement. Biopsies of affected bones may show irregular wide spaces in cortical bone, enlarged marrow spaces with thin-walled capillary-like vessels and variable fibrous stroma surrounding the vascular spaces. Both lymphangiomatosis and hemangiomatosis have been described. This likely represents a developmental endothelial defect.

The clinical findings usually rule out other causes of osteolytic lesions such as metastatic disease and osteomyelitis.

From the Department of Surgery, University of Saskatchewan, Saskatoon, Sask.

Accepted for publication Aug. 14, 2001.

Correspondence to: Dr. Grant G. Miller, Department of Surgery, University of Saskatchewan, Royal University Hospital, 103 Hospital Dr., Saskatoon SK S7N 0W8; fax 306 966-7988; miller@duke.usask.ca



FIG. 2. Computed tomography scan showing a soft-tissue mass (large arrow) involving the right paralumbar musculature adjacent to the posterior elements of the vertebral column and osteolysis of the vertebral body (small arrow).

Other types of primary osteolysis are familial and multicentric. The imaging characteristics, when correlated with the clinical findings, usually will permit the diagnosis of Gorham's disease positively rather than by exhaustive exclusion. This is particularly true when it is associated with a chylothorax.

The chylothorax, in most cases, occurs as a direct extension of lymphatic dysplasia into the pleural cavity, and the bony disease is usually readily identified on chest radiography. The results of treatment are difficult to evaluate because of the variable natural history, and there is no consensus on the most efficacious therapy. Radiotherapy and chemotherapy have not been consistently effective.<sup>3,4</sup> Interferon- $\alpha$ , with its antiangiogenic properties, has been successful in treating life-threatening hemangiomas of infancy and 1 case of Gorham's disease associated with a chylothorax.<sup>5,6</sup> Spontaneous regression without treatment has been reported.<sup>7</sup>

The treatment of chylothorax complicating Gorham's disease is of highest priority owing to the high mortality associ-

ated with chronic chyle loss. Hemorrhage as a lethal complication of Gorham's disease, as in this case, has not been reported before. Serial thoracentesis, fat restricted diets or eliminating oral intake may help temporarily, but alone they are not adequate. Thoracic duct ligation alone is usually inadequate treatment. Pleurectomy with or without thoracic duct ligation has been reported as successful in 4 out of 5 cases of chylothorax associated with skeletal lymphangiomatosis in children, including this case.<sup>8-11</sup> There was no recurrence of the chylothorax, with follow-up ranging from 3 months to 3 years.

### Conclusions

Gorham's disease is rare and most often affects children. When a child presents with a chylothorax in the absence of trauma, the physician should consider Gorham's disease and make a search for osteolytic bone lesions. The risk of death is high, but pleurectomy can be effective. Other therapies are unproven although interferon- $\alpha$  shows promise.

### References

1. Gorham LW, Stout AP. Massive osteolysis (acute spontaneous absorption of bone, phantom bone, disappearing bone): its relation to hemangiomas. *J Bone Joint Surg Am* 1955;37:985-1004.
2. Drewery GR, Sutterlin CE 3rd, Martinez CR, Brantley SG. Gorham's disease of the spine. *Spine* 1994;19:2213-22.
3. Heffez L, Doku HC, Carter BL, Feeney JE. Perspectives on massive osteolysis: report of a case and review of the literature. *Oral Surg Oral Med Oral Pathol* 1983;55:331-43.
4. Gutierrez RM, Spjut HJ. Skeletal angiomatosis: report of three cases and review of the literature. *Clin Orthop* 1972;85:82-7.
5. Ezekowitz RA, Mulliken JB, Folkman J. Interferon alpha-2a therapy for life-threatening hemangiomas of infancy. *N Engl J Med* 1992;236:1456-63.
6. Hagberg H, Lamberg K, Astrom G. Alpha-2b interferon and oral clodronate for Gorham's disease. *Lancet* 1997;350:1822-3.
7. Campbell J, Almond HG, Johnson R. Massive osteolysis of the humerus with spontaneous recovery — report of a case. *J Bone Joint Surg Br* 1975;57:238-40.
8. Canil K, Fitzgerald P, Lau G. Massive chylothorax associated with lymphangiomatosis of the bone. *J Pediatr Surg* 1994;29:1186-8.
9. Berberich FR, Bernstein ID, Ochs HD, Schaller RT. Lymphangiomatosis with chylothorax. *J Pediatr* 1987;87:941-2.
10. Dunkelman H, Sharief N, Berman L, Ninnan T. Generalized lymphangiomatosis with chylothorax. *Arch Dis Child* 1989;64:1058-60.
11. Bhatti MA, Ferrante JW, Gielchinsky I, Norman JC. Pleuropulmonary and skeletal lymphangiomatosis with chylothorax and chylopericardium. *Ann Thorac Surg* 1985;40:398-401.