While our patient’s tragic complication may have resulted in part from the lack of skill of his attendants, we believe that his chronic debility, his inability to communicate clearly, and his marked muscular weakness which precluded an adequate gag or cough made him particularly vulnerable. In patients of this type, we recommend that a chest x-ray film be taken following insertion of a nasogastric tube in order to clearly document the tube’s position. This would be especially important for cases in which intubation seemed to be unusually difficult.

This case illustrates the need to maintain soundness of technique in even the most “benign” procedures.

REFERENCES

Gorham’s Disease of the Clavicle with Bilateral Pleural Effusions*

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A patient with Gorham’s disease (massive osteolysis, disappearing bone disease) of the right clavicle had bilateral sanguinous pleural effusions. Complete cure was achieved by removal of the bony remnants with the hemangiomatosus mass which caused bone destruction, and by obliteration of the pleural spaces using repeated talcum insertion. In six out of seven previously described cases of Gorham’s disease with pleural effusions, the patients died, while the only survivor had a unilateral chylous effusion.

Massive osteolysis, disappearing bone disease, or Gorham’s disease, is a rare condition, first described by Jackson in 18381 in the humerus of a 13-year-old boy. About 70 additional cases have been described since then. The disease is manifested by lysis of one or more adjacent bones. The process has been described in various bones, including ribs, vertebrae, and scapula.2,5,7

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Figure 1. Initial roentgenogram taken following contusion of the right shoulder (5/3/77). Destruction of lateral aspect of right clavicle is evident.

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and phosphorus were all within normal limits. On the chest roentgenogram, large bilateral pleural effusions were seen (Fig 2A). The right clavicle had almost completely disappeared, with a few spicules seen on the medial third of the bone site (Fig 2A).

A right-sided thoracentesis yielded 1,500 ml of sanguinous fluid, with a protein content of 4.95 g/100 ml. The electrolyte and lipid content was similar to that of the patient's serum. Repeated right and left sided thoracenteses were needed because of rapid reaccumulation of sanguinous fluid (with a hemoglobin of 5.7 g/100 ml). No malignant cells were seen on cytologic examination. There was no growth of acid-fast or other bacteria.

Surgical exploration of the right clavicular region revealed an almost complete loss of bone structure. Only a few bone spicules were seen, especially in the medial third. The periosteum was partially preserved. An ill-defined, highly vascular mass replaced the missing bone, measuring approximately 8 x 3 x 3 cm. It was removed as far as possible. Extension of the mass toward the right pleura was identified.

Histologic Findings: Most of the bone was replaced by a tumor-like tissue, composed of new vessel formations lined by mature endothelial cells. Nonneoplastic proliferation of small vascular channels, predominantly capillaries, some of them containing blood elements, was evident. In other areas, endothelial proliferation formed clusters of budding cell masses (Fig 3A). Vessels with thicker walls, composed of overlapping layers of endothelial cells were seen. A few osteoclasts could be identified along the remaining bone spicules (Fig 3B).

The rate of accumulation of the pleural effusions did not change after the operation (Fig 2B and C), and repeated thoracenteses were required. Bilateral thoracoscopy was performed, and pleural biopsy specimens were taken on each side. Talcum was instilled into each cavity twice. The biopsy specimen revealed nonspecific inflammatory changes.

After the second instillation of talcum, there was no further accumulation of fluid.

The patient was followed-up closely. She was last seen in October 1979 (2½ years after the initial diagnosis of the clavicular lesion). She had no complaints. No signs of any additional bone lesions were evident. Repeated chest roentgenograms have been normal (Fig 2D).

Discussion

The clinical course and pathologic findings of the presented patient are compatible with the diagnosis of Gorham's disease. A hemangioendothelioma with bilateral pleural metastases could not account for the completely benign course 2½ years after its first presentation. Nor did the thoracoscopy show any evidence of a malignant process. Most previous reports emphasize the highly vascular nature of the tissue which replaces the bone. Definitions such as "skeletal angiomatosis" or "hemangiomatosis" are commonly used.2-4,7-10

The tumor may extend and destroy adjacent bones.2-4,7 On the other hand, spontaneous arrest was noted in some cases.5,7 This renders the evaluation of any given treatment unsure. Both surgical removal and radiotherapy were tried, with varying degrees of success.4 As the clavicle is easily accessible, we preferred to remove it surgically. No further bone lesions were evident in the following two years.

Pleural effusions in the reported cases were either bloody or chyloous, caused by hemangiomma or lymphangiomma respectively.2,4,8-11 A penetrating hemangiomma into the pleural cavity seems to be the cause of the bloody effusion in our patient. The fact that it was bilateral can be explained by the extension of the
infiltration mass bone cavity) 20. Ficunz 3 1. FEIGL, pleural effusions. Repeated instillations of talcum into the pleural spaces, without any further surgical intervention, was followed by complete recovery.

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mass into the contralateral pleura. A similar case, with infiltration of both pleuras (producing a common pleural cavity) has been described.10

Recurrent pleural effusions were the cause of death in six of the seven reported cases, as a consequence of circulatory failure, cachexia, and infection.2,4,9-11 The only survivor had a unilateral chylous effusion. He was cured by surgical obliteration of a lymphangioma and pleurodesis followed by the insertion of talc into the pleural cavity.3

The operation in our patient did not stop the reaccumulation of the sanguinous pleural effusions. Repeated instillations of talcum into the pleural spaces, without any further surgical intervention, was followed by complete recovery.

Figure 3 A (upper). Biopsy specimen of right clavicle. Endothelial proliferation forming clusters of budding cell masses. B (lower). Vessels composed of overlapping layers of endothelial cells. One osteoclast is seen on upper right angle of bone spicule (hematoxylin eosin, original magnification X 50).