A 20-year-old white man sought medical attention after noting the development of nontender masses in the left supraclavicular and left flank regions. There was no history of trauma, recent illness, prior similar masses, fever, chills, weight loss, or other symptoms.

Past medical history was notable only for the excision of a "rib cyst" at age 12. He did not smoke, use alcohol or take any medications.

On physical examination, two ill-defined subcutaneous soft mass lesions, one in the left supraclavicular fossa (4 × 5 cm) and a second in the left flank (6 × 6 cm) were identified. The lesions were nontender and no overlying skin change or evidence of trauma were evident. Chest examination was notable for percussive dullness and diminished breath sounds over the left base posteriorly, without signs of consolidation. The remainder of the physical examination was unremarkable.

The complete blood count, urinalysis, serum electrolytes, blood urea nitrogen and serum creatinine were all normal. A chest roentgenogram showed elevation of the left hemidiaphragmatic contour consistent with a subpulmonic effusion (Fig 1). In addition, two cystic lesions involving the left seventh and ninth ribs were identified (Fig 2). A lateral decubitus chest roentgenogram confirmed the presence of a left pleural effusion.

A subsequent thoracentesis yielded chylous pleural fluid with a triglyceride content of 1,050 mg/dl and cholesterol content of 300 mg/dl.

A diagnostic procedure was performed.
Diagnosis: Lymphangiomatosis

The patient underwent biopsy of the left supraclavicular mass (Fig 3). The tissue was consistent with a diagnosis of lymphangiomatosis, an infrequently encountered disorder characterized by abnormal proliferation of lymphatic vessels. A subsequent lymphangiogram demonstrated multiple sites of tortuous lymphatics in the retroperitoneum and the mediastinum. In addition, a discrete thoracic duct could not be identified. A radiographic bone survey revealed multiple radiolucent cystic areas involving the skull, scapulae, ribs, humeri, and tibiae. Computerized axial tomographic scanning of the head, chest, and abdomen did not demonstrate adenopathy or abnormal masses.

The combination of chylothorax and bone lesions in association with lymphangiomatosis was first described in 1967. A number of additional cases and reviews have since been presented.

Although pathologically a benign disorder, the potential morbidity and mortality from this condition stems chiefly from the nutritional and immunologic complications of draining chylothorax. The control of chyle leak in the presence of thoracic lymphangiomata can be quite troublesome. Chest tube drainage in combination with restriction of enteral fat to slow the rate of chyle flow, usually in conjunction with parenteral alimentation, has been generally accepted as an initial approach to management. Usually however, a surgical procedure has been needed to effectively halt the leak of chyle into the thorax. Various approaches, usually in combination, have included ligation of the thoracic duct, ligation and excision of mediastinal lymphangiomata, and parietal pleurectomy. In most cases, outcome following surgery has been good.

Two reports suggest a role for radiation therapy where attempts at surgical management have been unsuccessful.

In our patient, chest tube drainage in conjunction with parenteral alimentation did not succeed in slowing the pleural chyle leak. With clinical evidence of progressive nutritional deterioration, the patient underwent left thoracotomy with decortication of a thick peel from the left lung, and a parietal pleural abrasion. A cystic rib lesion was also excised at the time of surgery. Histologic section of the decalcified specimen confirmed the presence of lymphangioma involving bone.

Postoperatively, the patient's effusion did not recur and he continues to do well after one year of followup.


REFERENCES


Roentgenogram of the Month (Duckett, Lazarus, White)