37. LYMPHANGIOMA, CYSTIC ANGIOMATOSIS

A solitary bone lymphangioma is exceedingly rare. Anecdotal cases have been reported both in the axial and in the appendicular skeleton. They present with mild pain and, radiographically, with a pure osteolysis having rather well defined borders. Grossly, the lesion, rather than blood, contains a chyloid fluid, clear to cloudy. Histologically, the lymphangioma is composed of vascular spaces, from capillary to cavernous, larger, more elongated and spaced further apart than hemangioma. The cavities are empty or contain a proteinaceous fluid, the endothelial lining is flat and mature, and the reticulin wall is very delicate.

The differentiation between hemangioma and lymphangioma may be problematic: blood can be present in the lymphangioma cavities due to haemorrhage, or, on the contrary, the cav-
Fig. 37-2. Cystic angiomatosis, in a girl aged 9 years. The lesion, histologically proved as lymphangioma, involved the cervical spine and mediastinum.

erns of hemangioma can be empty due to blood escape during surgery or histological preparation.

Also rare is cystic angiomatosis (lymphangiomatosis, hamartomatous haemolymphangiomatosis), extended to several bones, both of the axial skeleton and of the limbs (not affecting hands and feet). This lesion is usually seen in children and exceptionally has shown familiarity. The skeletal involvement is often associated with soft tissue (fig. 37-1, 37-3) or visceral lymphangiomas, occasionally with pleural or pericardial chylous effusions.

Imaging shows multiple round to oval osteolytic lesions, mono or multilocular (from “honeycomb” to “soap-bubble”), sharply outlined, with or without a sclerotic rim. Lymphangiography may show dilated lymphatics within the soft tissues and viscera, and (one to a few days after the injection of the contrast medium) accumulation of the dye even in the affected bones.

Pathologic fractures are frequent.

Histologically, The features are the same as lymphangioma, with large endothelial lined spaces, with very delicate walls not surrounded by any tumoral or reactive tissue.

The lesion tends to eventually stabilize around the puberal age and even spontaneously regress (progressive sclerosis and cyst obliteration).

Treatment is only aimed at preventing - correcting the pathologic fractures and deformities.

It is sometimes difficult to establish whether the lesional vascular spaces are of blood or lymph vessel origin. The two may even associate.