VASCULAR AND CARTILAGINOUS HAMARTOMA (MESENCHYMOMA) OF THE KIBS IN INFANCY

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Vascular and cartilaginous hamartoma of chest wall, also known as mesenchymoma, is a very rare condition which arises exclusively in the chest wall of infants. Less than 20 cases have been reported. Its importance lies in the fact that sometimes it is mistaken radiologically and histologically for a malignant tumour. It is not only benign but also self limiting and its recognition is important if overtreatment is to be avoided. A case of chest wall hamartoma in a four month old baby is reported.

CASE REPORT

A four month old boy was admitted to the national Institute of Child Health, Karachi with a bulging mass on the right side of chest. The child was born at full term by normal delivery. Three days after birth, the mother noticed a swelling on the right side of the chest. There was no other complaint. On physical examination a firm mass, 6 cm in diameter was noted on the right lateral chest wall inthe mid axillary line. A chest X-ray showed a large soft tissue mass with irregular calcification in the right side of the chest causing deformity of the ribs (7th through 10th). Scoliosis of the dorsal spine was noted with convexity to the left (Figure 1).

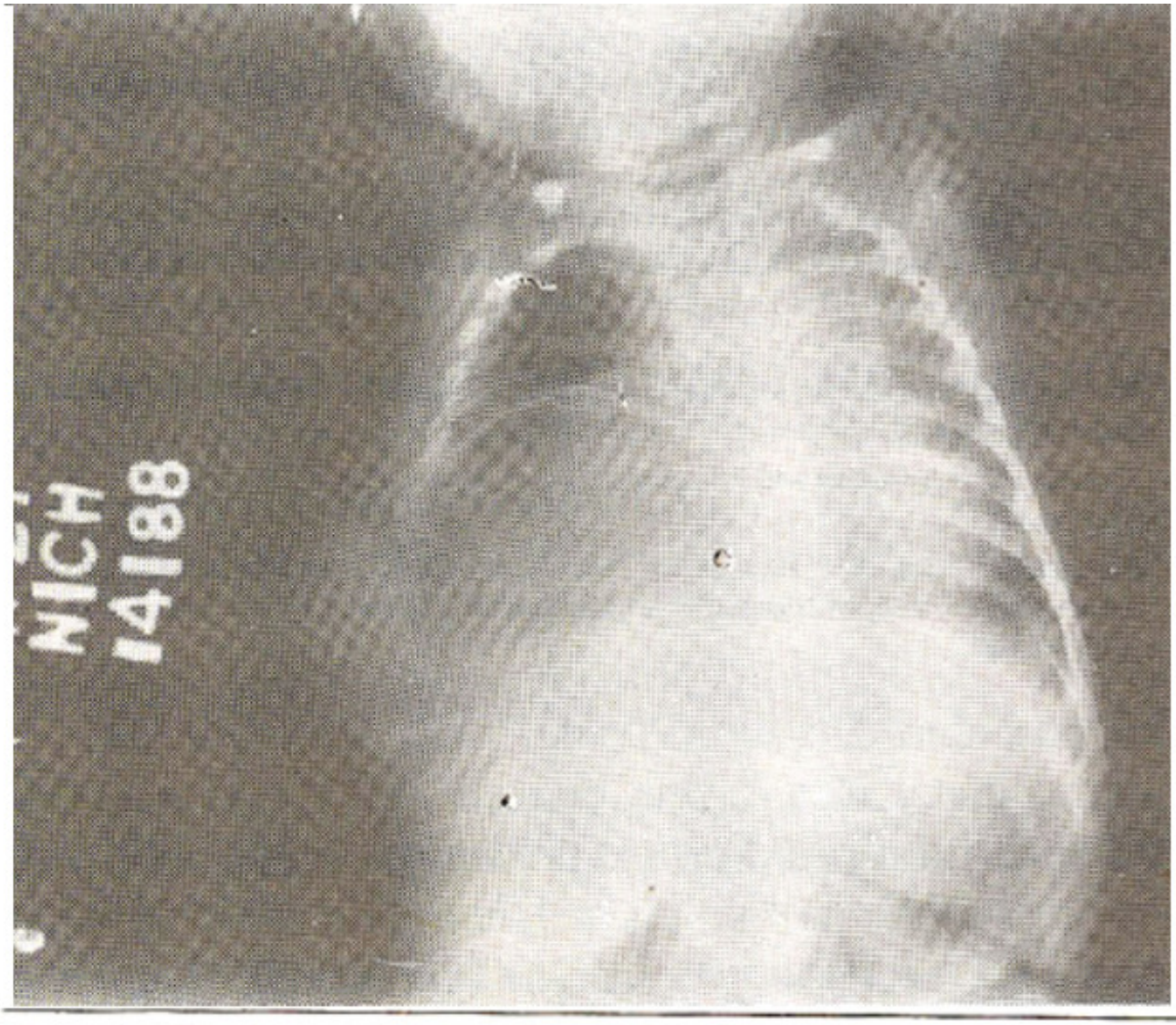

Figure 1. Chest x-ray showing a large soft tissue mass with irregular calcification in the right side of the chest. It is causing deformity of ribs (7th through 10th) with associated scoliosis of dorsal spine.

A clinical diagnosis of teratoma was made. At surgery a circumscribed extrapleural mass was found which was excised and sent for histopathology. Gross examination showed a greyish white smooth mass, partly cystic and partly solid measuring 9x6x5 cm in size. Cut section showed a cystic space 3 cm in diameter filled with reddish brown soft material. The rest of the mass was greyish white, very firm in consistency and at places cartilaginous on cutting. Microscopic examination showed islands of hyaline cartilage lying in a stroma which was very cellular at places and composed of closely placed spindle shaped cells. Areas of calcification and ossification were also seen. Some fragments were composed of large blood filled spaces separated by fibrovascular septae containing osteoclast type of giant cells (Figures 2 & 3).

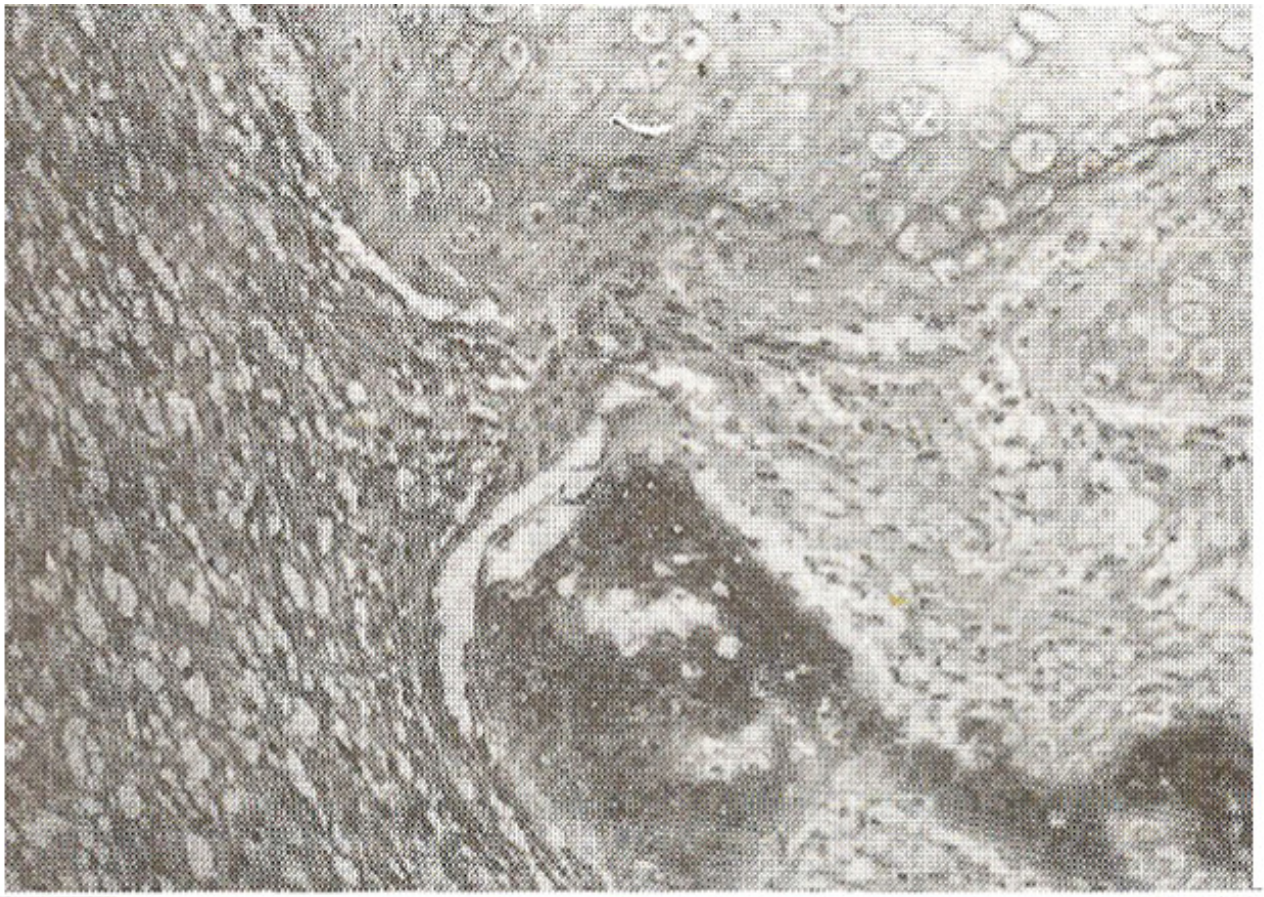

Figure 2. Photomicrography showing islands of hyaline cartilage and osseous trabeculae lying in a cellular fibrous tissue strom. (H&Ex700).

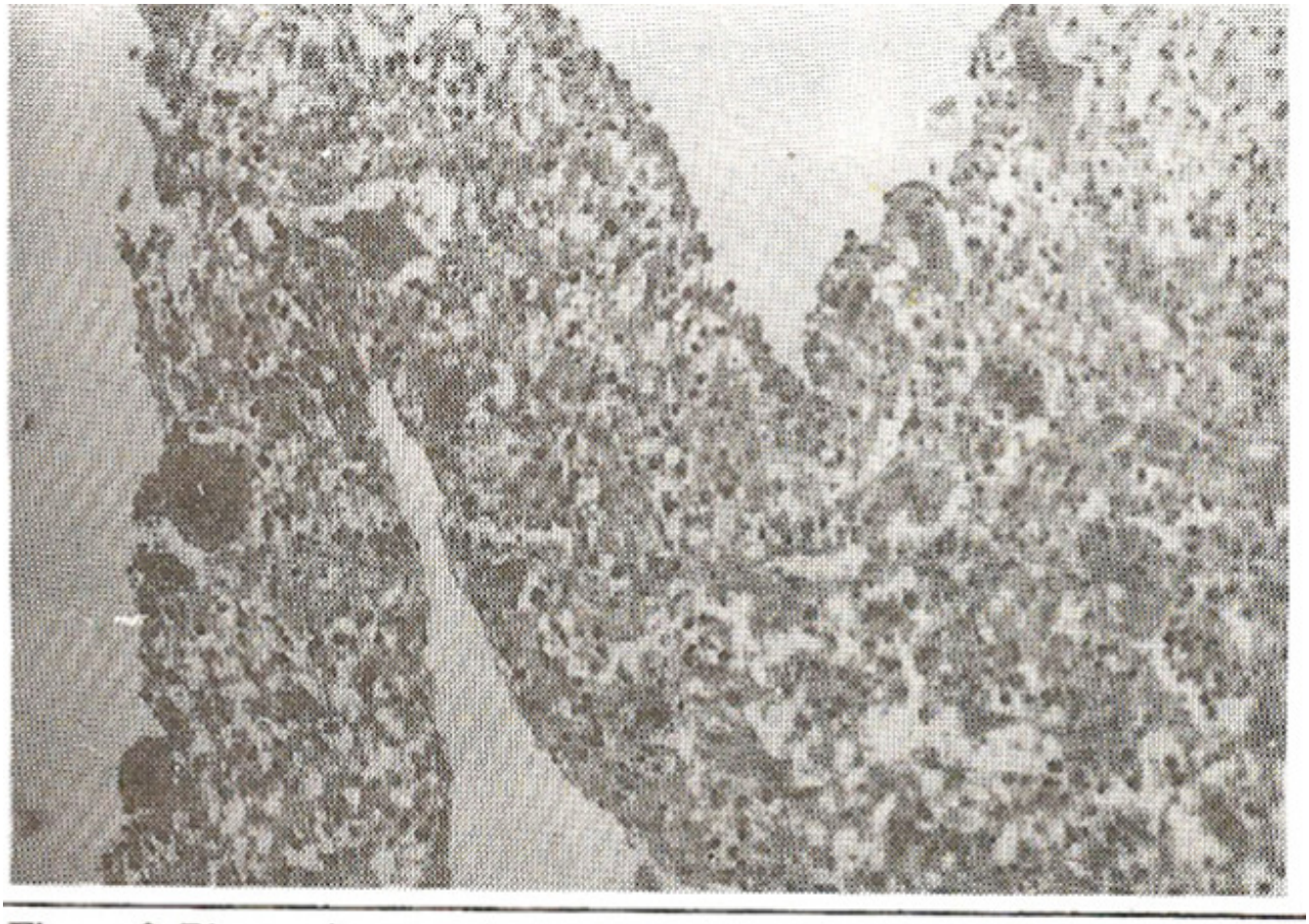

Figure 3. Photomicrograph showing portions of large dilated vascular spaces separated by fibrovascular septae containing osteoclast type of giant cells (H&E x 350).

DISCUSSION

Chest wall hamartoma is a very rare condition, presenting in infants at, or shortly after birth. Review of literature provides reports on less than 20 documented cases[1-5]. It is now mentioned in most textbooks of bone tumours and is twice as common in male, as in female infants. It has been diagnosed in utero[1] and is probably always present at birth, most cases being recognised in the first year of life. The mode of presentation ranges from an incidental fmding on a chest radiograph to severe respiratory distress due to compression of the lungs by the large extrapulinonary mass. However, they most commonly present as a chest wall lump possibly with a dorsal scoliosis and mild respiratory symptoms[1]. This bamartoma always arises in the lateral part of the chest wall from the main body of the rib distant from both costochondral and costovertebral junctions. Several ribs maybe involved. Two separate lesions have been reported in the same patient[5]. The size of the hamartoma varies greatly. Usually the subcutaneous swelling is small, the bulk of the lesion bulging into the thoracic cavity as a well circumscribed reddish brown mass covered by pleura. On section the hamartoma is soft, sometimes with sponge-like blood filled spaces and islands of cartilage may be recognised if large enough. Microscopy shows a mixture of hyaline cartilage, endochondral ossification, calcification and large number of vascular spaces resembling aneurysmal bone cyst in a background of vascular spindle celled

tissue. Very cellular areas in which spindle cells are associated with irregular woven bone trabeculae may mimic osteoblastoma. Most of the 20 or so reported hamartomas have been treated by surgical resection and none has recurred locally or metastasized. Chest wall hamartoma has also been called mesenchymoma. McLeod and Dahlin propose the name hamartoma rather than mesenchymoma which implies a benign or malignant neoplasm[4]. The histological features as well as clinical presentation at birth or early infancy and the benign behaviour suggests a bamartoma rather than a neoplasm.

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