# CARTILAGINOUS HAMARTOMA OF THE CHEST WALL WITH SECONDARY ANEURYSMAL CYST – LIKE AREAS IN AN INFANT*

## A Case Report

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**SUMMARY: Göre O, Kılıçalp A, Başdemir G, Özer E, Aktuğ T. (Department of Pathology, Dokuz Eylül University Faculty of Medicine; Department of Pathology, Ege University Faculty of Medicine; and Department of Pediatric Surgery, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey). Cartilaginous hamartoma of the chest wall with secondary aneurysmal cyst-like areas in an infant. Tur J Pediatr 1999; 41: 139-142.**

**A case of a four-month-old infant diagnosed as cartilaginous hamartoma of the rib is presented. This rare tumor usually presents at birth. The patient had respiratory distress syndrome. Swelling of the ribs was palpable on physical examination and the infant underwent surgery for excision of the ribs. Histopathologically, the tumor showed benign focal overgrowth of normal skeletal elements with cartilaginous, vascular and primitive-appearing mesenchymal elements. Additionally, secondary aneurysmal cyst formation coexisted with the tumor. The diagnosis was infantile cartilaginous hamartoma of the rib. In this entity, primitive-appearing mesenchymal stroma may be mistaken for a malignant condition. Usually a benign clinical course is expected and treatment is by block excision.** *Key words: cartilaginous hamartoma, benign mesenchymoma, rib.*

Infantile cartilaginous hamartoma (ICH) of the rib is an extremely rare neoplasm characterized by cartilaginous, vascular and primitive-appearing mesenchymal elements. It presents in newborns and infants. The tumor is associated with a benign clinical course and cure is by block excision. Mirra[1] pointed out that this entity might be mistaken for a malignant condition because of the primitive appearance of the mesenchymal component. The rarity of the tumor has been reported by McCarthy et al.[2] and McLeod et al.[3] under the designations infantile ICH or benign mesenchymoma of the rib. Since the entity never metastasizes and is curable with local resection, it is now clear that it is a congenital benign hamartomatous lesion of the rib.

## Case Report

A four-month-old female infant was admitted to Dokuz Eylül University Hospital with the complaint of respiratory distress. The initial chest radiogram and thoracal computed tomography revealed a large multicystic intrathoracic mass occupying

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almost the whole of the left hemithorax (Fig. 1). There was erosion on two ribs. At surgery, the entire mass along with portions of the left sixth and seventh ribs were completely, removed. On gross examination, the tumor measured 85x40x35 mm and showed two large nodules (Fig. 2). The inner aspect of the ribs was destroyed by the mass. The cut section showed firm whitish glistening cartilaginous tissue admixed with multicystic congested tissue. Microscopic sections of the tumor revealed a mixture of mesenchymal elements, including chondroid tissue with large endothelium-lined blood spaces resembling aneurysmal bone cyst with osteoclast-like giant cells and osteoid, and foci of calcification (Fig. 3). The cartilaginous component was composed of mature hyaline cartilage and prechondrocytes resembling chondroblasts. Some cartilaginous areas adjacent to the bone showed vacuolization and primitive formation of colums resembling that seen in epiphyseal cartilage (Fig. 4). The pathological diagnosis was infantile cartilaginous hamartoma. In the 20 months since diagnosis, the patient has grown normally with no evidence of recurrence.

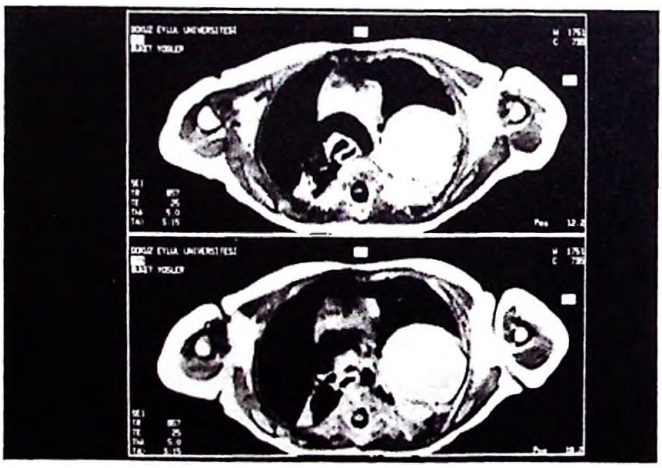

Fig. 1. Computed tomography of the thorax showing multicystic mass occupying the whole of the left hemithorax

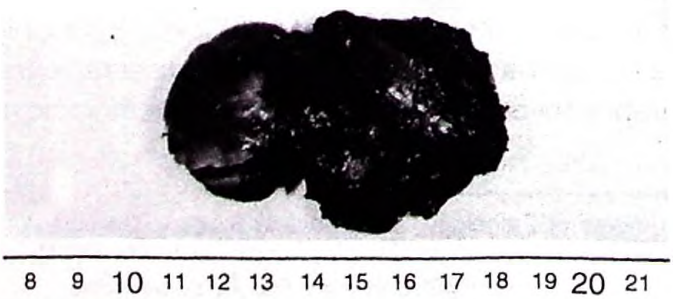


Fig. 2: Macroscopic appearance of the lesion. The lesion is composed of two nodules. The rib is partially destroyed.

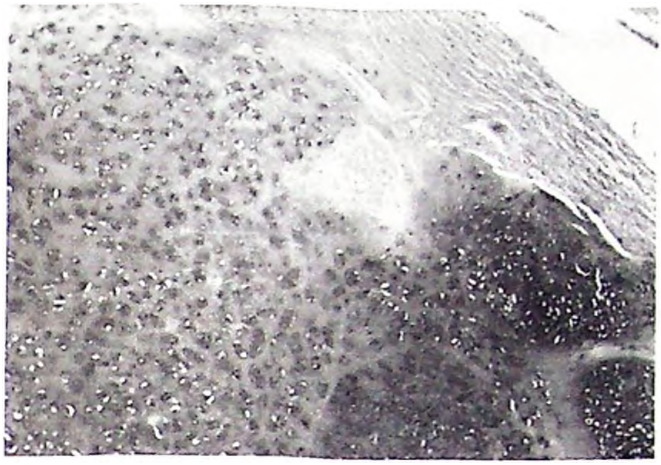

Fig. 3: Microscopic appearance of the lesion showing areas of large masses of hyaline cartilage, (H and E X40).

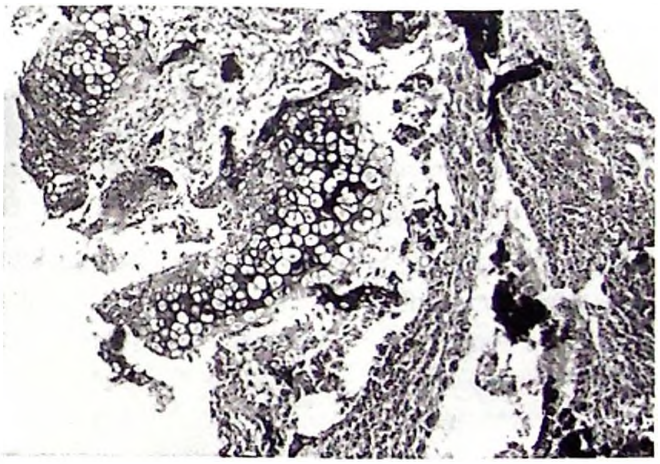

Fig. 4: Microscopic appearance of the lesion: Foci of woven bone and cartilage showing vacuolization and primitive column formation (H and E X40).

## Discussion

The report by McLeod et al.[3] reveals that infantile cartilaginous hamartoma, commonly known as benign mesenchymoma, is very rare in infants and usually presents at birth. The incidence is about 0.03 percent of primary bone tumors. About 25 cases have been reported in English literature[4-6]. As seen in our case, this is a solitary lesion of the rib with a size varying from 2 to 15 cm. Because the character of the lesion is benign, Mirra[1] and Cohen[7] noted that excision of the rib is the recommended procedure. Immunohistochemical localization of various collagen types is consistent with the non-neoplastic notion of character[6]. This lesion should be distinguished from primary aneurysmal bone cyst, chondroma, chondrosarcoma and other mesenchymal neoplasms.

This lesion is presented with either rib swelling or respiratory distress symptoms. In our case, respiratory distress symptoms were predominant. Rarely, incidental chest radiograms may reveal a large expansive mass of the rib, with or without spotty calcification characteristic of cartilaginous tumors. More than one rib may be involved and destroyed by the lobulated mass. The report by Blumenthal et al.[8] suggests that ribs adjoining the main lesion can be deformed or eroded by the extrinsic mass. The external surface of the rib is well circumscribed and smoothly expanded. The cut section shows variable-sized foci of chondroid to cartilaginous tissues with foci of calcification. In many cases, dilated blood-filled spaces similar to that seen in aneurysmal bone cysts may be observed[1].

Constant histological features in ICH are sheets of chondroblast-like mesenchymal cells without atypical mitoses or frank anaplasia, hyaline cartilage derived from hamartomatous chondroblast-like stroma which can be mistaken for a chondrosarcoma, and prominent vascularity for aneurysmal bone cyst. The variable histological findings are foci of calcification, woven bone production, reactive osteoclast-like giant cells, enchondral ossification similar to growth plate cartilage and fibroblastic hyperplasia, particularly in aneurysmal bone cyst-like areas. In 1972 two cases with atypical mitoses, frank anaplasia and prominent hypercellularity were reported as intrathoracic malignant mesenchymoma[8]. However, no evidence of malignancy was observed in the present case.

To conclude, we have reported a case of infantile cartilaginous hamartoma because of its rarity and constant histological features. There has been no evidence of recurrence, during the 20 months since the excision, consistent with its benign clinical course.

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