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Case Report 209

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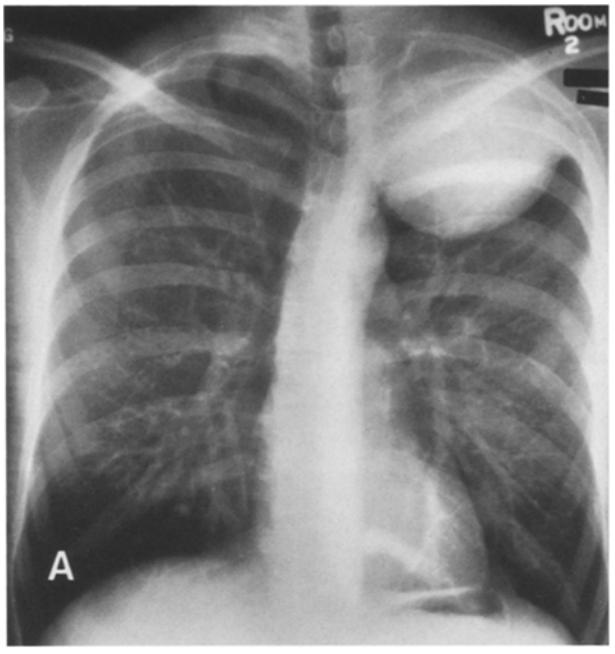

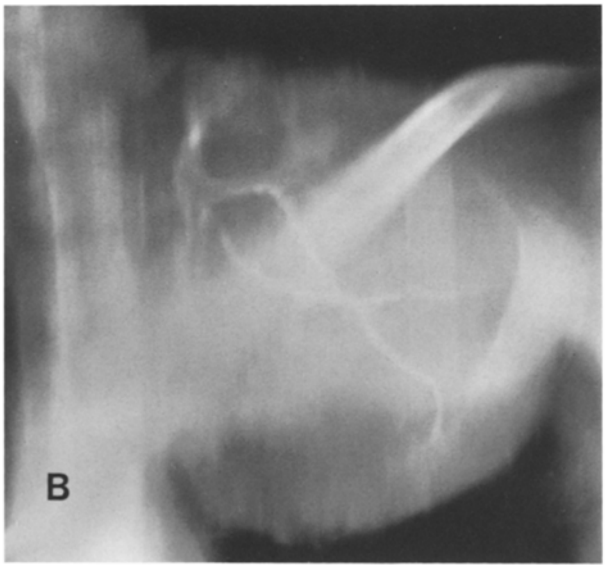

Radiological Studies

Fig. 1A, B. A A postero-anterior roentgenogram of the chest demonstrates a well-defined opacity, approximately 11 cm in diameter, overlying the left hemithorax. The left 1st rib is markedly thinned and expanded. Bony trabeculae are noted throughout the opacity. **B** A laminogram of the left upper hemithorax confirms the well-defined margins of the opacity and the central bony trabeculae. The lesion appears markedly expanded

History

A chest roentgenogram obtained in this 31-year-old man because of a flu-like illness, showed an opacity overlying the left upper hemithorax, which appeared to be originating in the left 1st rib. A history of discomfort in this area was obtained, following a "pulled" muscle seven years previously. The discomfort had persisted since that time.

On examination slight fullness in the left supraclavicular fossa was noted. No evidence of adenopathy or any other abnormality in the shoulder joint was demonstrated. The remainder of the examination was unremarkable.

Hematological and other laboratory studies were normal.

The chest roentgenogram and a laminagram of the left first rib are demonstrated (Fig. 1A and B).

Angiography showed the lesion to be mildly vascular, with some abnormal vessels noted both in the peripheral and central areas. The subclavian artery and vein were displaced, but were neither encased nor invaded.

A biopsy followed by a surgical procedure was performed.

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Diagnosis: Chondromyxoid Fibroma of Left First Rib

The differential diagnosis includes fibrous dysplasia, "brown tumor" of hyperparathyroidism, hamartoma (rare) an infective process (e.g. tuberculosis, fungal disorder), chondrosarcoma, plasmacytoma and metastasis (the last two unlikely because of age of patient).

Discussion

Following biopsy the left first rib was resected from the costochondral junction anteriorly to the costovertebral articulation posteriorly. The biopsy showed a typical chondromyxoid fibroma (see Fig. 2A and B).

Chondromyxoid fibroma, a relatively uncommon benign tumor of cartilaginous derivation, comprising less than 1% of all skeletal neoplasms, was first described as a distinct pathological entity by Jaffe and Lichtenstein [2], who separated the lesion from chondrosarcoma because of its generally benign nature and distinctive histological appearance. However, about 20% of chondromyxoid fibromas may recur within two years of surgery, particularly in patients younger than 15 years of age and in lesions which are predominantly myxoid in nature [3, 4]. Malignant transformation of chondromyxoid fibroma has been reported as a very rare finding and even in these cases it is probable that such lesions were malignant from the onset [6].

Most authors report this neoplasm to be more common in males than females. Although the lesion has been reported in most skeletal areas the more common sites are the proximal end of the tibia (approximately one-third of all cases), distal end of the femur, fibula and, less commonly, the small tubular bones and flat bones. Mainly occurring in the first three decades of life, the lesion may also be observed in the fourth through the sixth decades. Usually the tumor is painful and commonly associated with localized swelling, with patients frequently giving a history of several years of discomfort before initial examination [5].

Radiologically, the typical appearance is that of an eccentric, metaphyseal lucency, with the long axis parallel to the long axis of the bone. A predilection exists for the diametaphysis in lesions affecting long bones. The medullary margin is usually scalloped, well-defined and frequently sclerotic. This feature is in contrast to the periosteal margin which may be attenuated and radiologically indistict. When occurring in small tubular bones the tumor generally occupies the entire width of the affected bone, producing expansion (which may be extensive), extreme thinning of both cortices and often, a trabecular framework within the lesion.

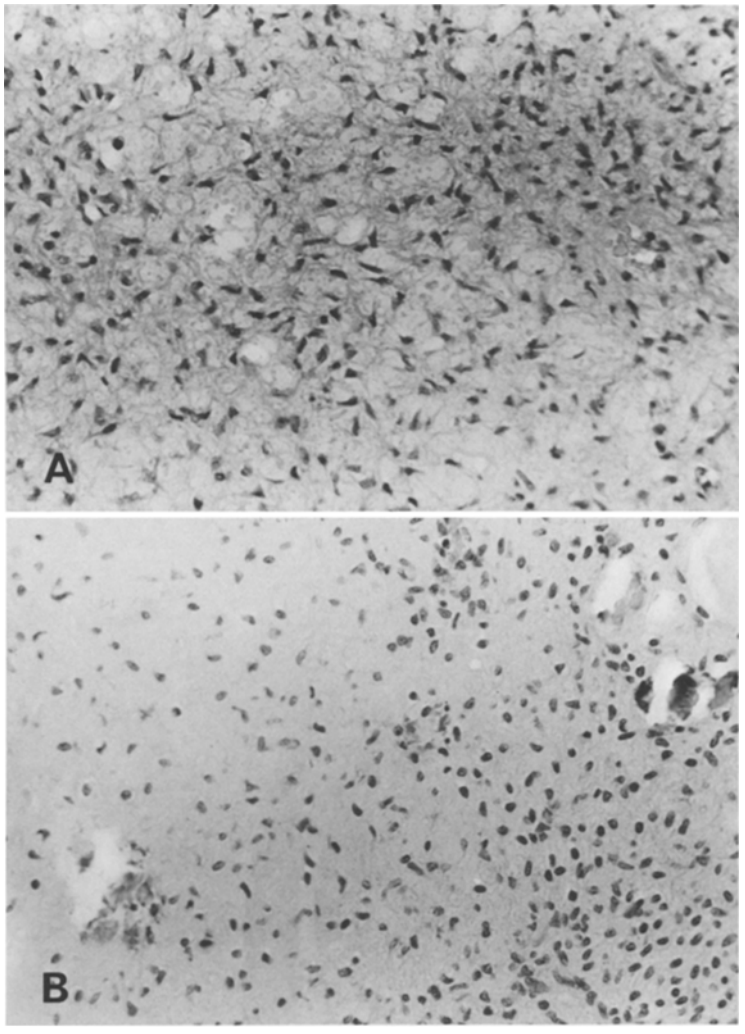
The gross appearance of chondromyxoid fibroma is usually that of a circumscribed tumor which on cut section demonstrates a lobular nature Chondroid, myxoid, and fibrous areas are characteristic. Cystic or hemorrhagic regions occur less frequently. Microscopic sections confirm the triphasic chondroid, myxomatous and fibromatoid matrices in a generally lobular arrangement (Fig. 2A). Any single tumor, however, may have a predominance or a paucity of only one or two of the matrix types. Between lobules, cellularity tends to be increased and benign giant cells may be scattered among the spindle and stellate-shaped cells (Fig. 2B).

Pleomorphism is uncommon, but may occur at the periphery of some of the tumor lobules.

As its name implies, chondromyxoid fibroma is a heterologous tumor which can be mistaken easily for other lesions by gross and microscopic examination. Distinction from chondroblastoma, chondroma and chondrosarcoma may be, on occasion, particularly difficult. The presence of the highly characteristic lobulated pattern with chondroid lobules showing various degrees of collagenization and separation by cellular myxoid tissue is the key to the diagnosis. Unfortunately this pattern can be mimicked focally by the other lesions just named, particularly chondroblastoma, and adequate sectioning is mandatory. The occasional occurrence of pleomorphic or multi-nucleated cells at the lobular peripheries can lead to the mistaken diagnosis of chondrosarcoma. Such cells observed only at the edge of lobules in a tumor having the above characteristics can, however, be assumed to be occurring in chondromyxoid fibroma.

Chondromyxoid fibroma arising in a rib is uncommon. First noted in 1951 [1] the lesion classically arises posteriorly, consistent with an origin adjacent to the physis. As with all rib lesions the skeletal origin may be actually overlooked; a recent case report described such a case of a chondromyxoid fibroma of the rib, initially thought to be a pulmonary lesion [7].

In summary, a large, expanding lesion of the left first rib in a 31-year-old man has been presented, proving histologically to represent a chondromyxoid fibroma in an unusual site. The radiological and histological features, the usual sites of



Pathological Studies

Fig. 2A, B. A Pleotomicrograph from specimen in this case shows myxoid area with small, spindle-shaped cells in a stellate arrangement within abundant ground substance (H and E stain, intermediate power). **B** Periphery of a chondroid lobule demonstrating increased cellularity in the right side of the field in this photomicrograph from same specimen (H and E stain, intermediate power)

involvement of the skeleton and the differential diagnosis of chondromyxoid fibroma have been stressed. Of particular importance is the close similarity, on occasion, to chondroblastoma, chondroma and chondrosarcoma.

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