Giant Costal Osteochondroma in a Man With Multiple Exostoses
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We report the case of a 42-year-old man who presented with kyphosis resulting from a giant symptomatic costal osteochondroma around the left fourth rib. The osteochondroma on the left side of the back was 56 cm × 47 cm × 33 cm and was painful. The size and growth of the tumor suggested a malignant transformation of a large costal osteochondroma. Multiple osteochondromas were also found on the legs. The patient's family history revealed hereditary characteristics. This patient was clinically diagnosed as a case of multiple familial osteochondromatosis. Complete removal of the tumor relieved the symptoms, and histopathologic examination confirmed malignant transformation of chondrosarcoma. There was no recurrence after 16 months of follow-up.


Hereditary multiple exostosis (HME), a type of rare benign bone tumor, is an inherited autosomal dominant disorder characterized by the presence of multiple exostoses or osteochondromas [1]. Osteochondromas commonly develop during the first decade of a person's life but stop growing when the individual reaches skeletal maturity [2]. The tumor commonly occurs in long bones but rarely affects ribs. The occurrence of a huge costal osteochondroma is rare in adults. We report a case of giant costal osteochondromas in a 42-year-old man with a family history of HME.

A 42-year-old man presented with approximately a 40-year history of multiple osteochondromas on the legs and ribs and deformity of the left side of the back. The patient was 164 cm tall. He had undergone a biopsy 10 years before and a benign tumor was diagnosed (results not available). The patient reported a history of mild pain and a significant increase in mass and size of the tumor on the left side of his back in the past 2 years, which limited motion of the left shoulder. The patient's family history was notable for multiple osteochondromas in several paternal relatives. The patient's father died 5 years before because of multiple osteochondromas, and the patient's brother and son had been clinically diagnosed with multiple osteochondromas. The patient's left scapula was found to be laterally displaced, with a bony protrusion that resulted in a hunchbacked deformity on the left side of the back (Fig 1). Physical examination showed a tumor measuring 56 cm × 47 cm × 33 cm on the posterior part of the chest, with significant tenderness to palpation. In addition, bony masses were also found around the right knee joint. Motor, sensory, and vascular examinations showed no compromise of the distal extremities. The patient's intelligence was normal, and ultrasonography did not detect any anomalies of the heart or kidneys.

Laboratory tests revealed normal values for hematocrit, electrolytes, liver enzymes, and coagulation factors. Bony masses were further noted on the right distal femur and the right proximal tibia in roentgenograms. Chest computed tomography (CT) revealed a large extrapleural tumor arising from the posterior aspect of the third and fourth ribs. The exostoses protruded through the intercostal space and fused to the vertebral pedicle and facet, but no pleural and pericardial effusions were observed. Three-dimensional CT demonstrated a mushroom-shaped bony protrusion on the posterior surface of the body. No mass was found on the right side. The lesion, measuring 56 cm × 47 cm × 33 cm, was directed toward the chest wall.

Chest CT was used to decide the best approach to excise the exostoses. First, the patient underwent a left thoracotomy through a lateral thoracic incision. A broad-based osteochondroma was noted. It originated from the fourth rib and extended into the third rib, the fifth rib, and thoracic vertebrae (T3-4). The intrathoracic part of the mass was approximately 8 cm × 6 cm × 4 cm. The
mass and the involved ribs were excised. The pathologic assay of the intraoperative biopsy sample confirmed the diagnosis of osteochondroma with evidence of malignant transformation. Therefore an S-shaped skin incision was made along the medial border of the scapula. The trapezius and greater rhomboid muscles were separated and retracted. The bony mass was removed together with the involved ribs beyond the lateral extent of the lesion. The patient also underwent a T3 and T4 laminectomy followed by the removal of involved muscles (serratus anterior, trapezius, rhomboid, erector muscle of the spine, and latissimus dorsi). The chest wall was repaired and reconstructed.

A gross specimen of the osteochondroma exhibited a size of 39 cm × 24 cm × 17 cm and a weight of 6.7 kg (Fig 2). A lot of central punctate cartilaginous matrix and calcification components were found. Histologic examination confirmed a typical osteochondroma composed of monotonous proliferation of cartilage cells with evidence of malignant transformation (Fig 3). The patient was discharged in stable condition. The follow-up was 16 months and no evidence of deformity was found. The patient no longer complained of pain and regained full range of motion of the shoulder and scapula.

Comment

Osteochondromas, also called exostoses, are the most common benign tumors of the bones, with an estimated rate of about 36% in all benign bone tumors and 8.5% in all bone tumors [3]. Osteochondromas may be solitary and multiple as a sporadic event or secondary to traumatic insult. Multiple osteochondromas can be sporadic or familial such as HME, which is a relatively rare disease [4]. The metaphyses of long bones of the extremities, predominantly around the knee, are commonly affected. The scapulae, vertebrae, and ilium may also be affected, but ribs are rarely involved [5, 6]. The occurrence of a huge costal osteochondroma in an adult is rare. Given his family history, as well as the nature, onset, and duration of symptoms, the patient described here was diagnosed with HME, although no specific genetic testing was available. To our knowledge, there was no previous report of multiple exostoses with such a large costal osteochondroma.

Costal osteochondromas are usually asymptomatic. Symptoms occur depending on the size and nature of the lesion. Pain, functional problems, deformities, and disturbance of blood circulation because of blood vessel compression can occur. Additionally, complications can result from mechanical injury to adjacent anatomic structures such as the pleura, the diaphragm, the heart, the lungs, and the vertebrae [7]. Complications of costal osteochondromas are uncommon, but spinal cord compression [8, 9], brachial plexus palsy [10], hemotherax [11, 12], pneumothorax [13], hiccup [14], snapping scapula [15], thoracic outlet syndrome [16], hemopericardium [17], and diaphragmatic rupture [18] have been occasionally reported. Spinal cord compression and disturbances of motor and sensory function of adjacent nerves may

Fig 2. (A) Macroscopically, the lesion was removed and measured 39 cm × 24 cm × 17 cm. (B) A cross-section showed cartilaginous tissue and calcified areas.

Fig 3. High-powered photomicrograph (original magnification, ×200; hematoxylin and eosin stain) showed hyperplasia of cartilage tissue, which was evidence of malignancy.
occurs. Although rare, malignant transformation to chondrosarcoma is the most severe complication. However, malignant transformations can occur in 0.5% to 10% of patients with HME [19]. Rapid growth of the tumor after puberty and/or the presence of pain is a possible sign of secondary chondrosarcoma [20]. Because of this risk, patients with osteochondromas, especially patients with HME, should undergo early assessment of chondrosarcoma transformation.

The diagnosis of costal osteochondroma is relatively easy in a patient with HME. In contrast, diagnosis of costal osteochondroma is usually difficult in a patient without a family history of the condition. Although plain roentgenography is routinely used to diagnose osteochondromas, CT is more sensitive and should be considered when a costal osteochondroma is suspected. CT is able to reveal the nature and location of the lesion as well as its topographic relationships with neighboring structures [21]. Three-dimensional CT can provide anatomic information for surgical removal of the mass. Magnetic resonance imaging, however, is more suitable for observing the true thickness of the cartilage cap, which shows high-signal intensity on magnetic resonance images.

Treatment of osteochondroma is usually conservative unless symptoms such as pain occur, the lesion is growing progressively, the tumor enlarges after skeletal maturation, or malignant transformation is suspected. Surgical resection should be considered if the costal osteochondromas have inward-protruding bony spicules or lead to complications and severe deformities. Most patients with these complications should undergo surgical resection. Complete resection with adequate margins is essential in patients with broad-based osteochondroma to reduce recurrence. A long period of follow-up is required to check for local recurrence.

In conclusion, we report a rare case of giant costal osteochondroma found in an older man with hereditary multiple exostoses. Malignant transformation was diagnosed in our patient. Surgical resection of the mass together with the involved ribs beyond the lateral extent of the lesion was performed. Follow-up at 16 months showed no evidence of recurrence. Our study suggests that tumors arising from the rib cartilage should be resected as early as possible if the costal osteochondromas are still growing and pain is present after puberty, leading to complications or severe deformities.

References


BRCA1 Gene Mutation in Thymic Malignant Melanoma

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We present a patient with triple primary malignancies: thyroid cancer, ovarian cancer, and thymic malignant

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