A 5-year-old Boy with a Large Hereditary Multiple Exostoses Lump Grown into Thoracic Cavity

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Introduction

HME is a dominant disorder characterized by multiple benign cartilaginous bone tumors that grows outward from the metaphyses of long bones. Exostosis primarily involves the long tubular bones of the extremities, such as the femur, tibia, fibula, humerus, radius, and ulna. Other bones such as ribs, scapulae, pelvis, and vertebrae are less often involved.1) The prevalence of HME was reported from as high as 1% of a small population in Guam, to 1 case in 100,000 in European populations.2) Complications of exostosis such as hemothorax, reactive tenosynovitis, popliteal artery pseudoaneurysm, central or peripheral nerve compression, bladder irritation and hematuria or urinary obstruction, are uncommon. They are caused by interference from surrounding anatomical structures. The patient in this report is the first child to present with a large chest lump due to costal exostoses. Our patient received video-assisted thoracoscopic surgery (VATS) and a completely resection of the lump and 1 cm ribs involved, resulting in a very positive prognosis.

Case Report

A 5-year-old boy was admitted to our hospital with a 1 week history of chest distress and right chest pain. He had no preceding traumatic history and the chest pain was gradually increased. There was no sign of coughing, sputum, vomiting, palpitation or fever. His medical history showed that 2 years previously he was found to have multiple painless lumps in knee joints which did not affect his physical activity. The boy did not receive any medical treatment for these lumps. His grandmother and father were also found having multiple arthral painless lumps which did not affect their physical activities. Symmetry nodules were found in their bilateral scapulas, elbow joints and knee joints.

Chest radiography revealed a mass in the right thoracic cavity. Computed tomography (CT) showed the right third and fourth ribs were irregular, and a 4 cm × 3.5 cm
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A high-aniso-density mass was identified in the posterior margins, projecting toward the middle lobe of right lung (Fig. 1). His laboratory values including hemoglobin level, hematocrit, electrolytes, liver enzymes and coagulation parameters which were all normal.

During the operation, a 4 cm irregular firm osseous lump from the right fourth costal cartilage was found to be tightly adhered to the visceral pleura, however not invaded into the parenchyma of the lung. A 4 cm × 3.5 cm lump was completely removed and 1 cm of the fourth ribs were partially resected under VATS. The clinic diagnosis of exostoses was confirmed pathologically (Fig. 2). During a 2 years follow-up, the patient remained in good condition without recurrence and complications.

Discussion

Complications of exostosis present in the chest cavity are rare. Cases of hemothorax as a result of costal exostosis with HME have been reported in literatures and costal exostoses are very small. The proposed etiological mechanisms are either the shearing of pleura or diaphragm by the relatively sharp, margins of the intrathoracic costal exostosis, or focal pleural changes induced by long-standing friction between the intrathoracic exostosis and the visceral pleura due to respiratory motion, leading to rupture of the dilated vessels. This 5-year-old boy is a rare case presented with a large chest lump over 2 cm in diameter in child owing to HME, which revealed that the exostoses could grow very fast and without hemothorax in child. Hereditary multiple exostoses was characterized by multiple osteochondromas. The exostosis of infant grows very fast and stop growing end of the body growth. Some cases of patient showed significant growth in adulthood, the incidence of about 25%.

It’s generally agreed that when there are no symptoms present, a HME patient could either avoid any treatment or postpone operation until adolescence. However in this child the large costal cartilage exostosis intruded into the pleural cavity and leads to chest pain and chest distress, so a minimally invasive VATS was given. Although HME is benign in nature, exostoses can sometimes transform into malignancy (5% of cases) in adulthood. In the previously reported cases involving chest cavity, they were mostly approached with excellent results via formal thoracotomy or thoracoscopic approaches with resection of either the involved rib segment or the exostosis itself. The outcome of surgery was satisfied with no significant complications in all these reported patients.

A three-layered structure, the bony stalk, the cartilaginous cap and the cartilaginous membrane from the...
inside to the outside of this patient’s lump, was demonstrated histologically and pathologically. The cartilaginous cap was 1 to 5 millimeters thick. Regardless of the benign nature of this lump, due to its rapid growth in the thoracic cavity a follow-up should be carried out routinely. The suspicion of secondary chondrosarcoma after puberty, the presence of pain, or an increase over 1 cm in the thicknesses of the cartilaginous cap, are the main points for future observation.

Disclosure Statement

The authors report no conflicts of interest in this work.

References