

CASE REPORT

Multiple Rib Exostoses in a Boy: A Rare Case Resulting in Surgery Secondary to Cosmetic Concerns

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Abstract

A seven year-old boy with several painless masses on the ribs and shoulder was referred to our hospital. The masses were so prominent that they prevented the child's sleep. Since the patient had been ridiculed by his friends due to the rib prominences, he had refused to attend school. After clinical and radiological evaluations, the masses were diagnosed as hereditary multiple exostoses of the shoulder and ribs. He underwent surgery for cosmetic reasons resulting in the patient's return to a normal life.

Key words: Hereditary Multiple Exostosis (HME), Osteochondroma, Rib exostosis, Rib tumors

Introduction

Osteochondroma, also known as exostosis, is the most common cartilaginous tumor (1). It is also known as the most benign bone tumor, which is estimated to account for 20 to 25% of all benign tumors (1).

About 3% of osteochondromas are estimated to occur in vertebrae and ribs; among them, 70% are accompanied by hereditary multiple exostoses (HME) (1-2). HME mostly appears in skeletal bone and has a multiple autosomal dominant heredity. Rib exostosis is rare and occurs in the area of costovertebral junctions or in the level of rib vertebrae (1-2).

Osteochondroma in the chest wall region may cause external pressure on the chest organs and result in subsequent symptoms such as shock due to massive hemothorax, recurrent chest infections due to emphysema, diaphragm damage, or skeletal deformities (3-9). The patient in this report is a seven year-old boy with multiple exostoses in the chest wall underwent surgery for cosmetic reasons.

Case report

A seven-year-old boy with multiple lesions in the

chest wall and extremities was admitted to our referral hospital. The only complaint of the patient was several solid prominences in the chest wall causing disruption during sleep as well as cosmetic problems for both the child and parents. The patient refused to go to school due to psychic problems given that other children mocked him and called him "boney boy". The prominences were found three years prior to admission and they grew in the recent years. There was no sign of chest pain, breathing distress, coughing, sputum, vomiting, palpitation or fever. The bumps were solid and painless, with the round border firmly attached to the patient's chest wall. Two of them existed in the middle and lower part of the left lateral chest wall, one in the middle of the right lateral chest wall, and the fourth attached to the upper portion of the scapula, moving with shoulder movements (Figure 1). Our clinical diagnosis was multiple exostoses. Family history also showed exostosis in the patient's father and uncle, as well, but the disease in them did not affect the chest wall nor their physical activity. In addition to rib exostoses, there were symmetrical exostoses in the upper limbs. Chest radiography revealed a large bony mass in the lower thoracic cavity below the

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Figure 1. Ribs involvement in a seven year-old boy resulted to unaccepted cosmetic appearance. A: anterior view. B: lateral view. C: posterior view.

diaphragm, attached to chest wall (Figure 2). A CT scan showed multiple bony structures attached to the ribs compatible with exostosis lesions (Figure 3).

The patient laboratory values such as white blood cell count, hemoglobin level, hematocrit, liver enzymes, electrolytes and coagulation parameters were all within normal ranges.

After consulting with the patient's parents and the patient himself, according to the cosmetic and psychiatric effects of the disease, we decided to excise all four prominent lesions from the chest wall, as well as the one below the diaphragm, in one anesthetic session. The lesions that penetrated to the thorax and diaphragm were removed by limited thoracotomy.

After surgery the patient went back to school. The

patient was followed up for 2 years and no complications or recurrences were reported.

Discussion

Despite the fact that an exostosis is a benign lesion both clinically and histologically, sometimes it may cause clinical problems such as pain, compression of the adjacent soft tissues, and aesthetic problems (such as in our patient).

Although exostosis in the long tubular bones of the extremities is a well-known tumor, it is a rare condition in the ribs and thoracic wall (1-2, 9). The differential diagnoses of exostosis in the ribs include enchondroma, osteoblastoma, osteoid osteoma, chondroblastoma,

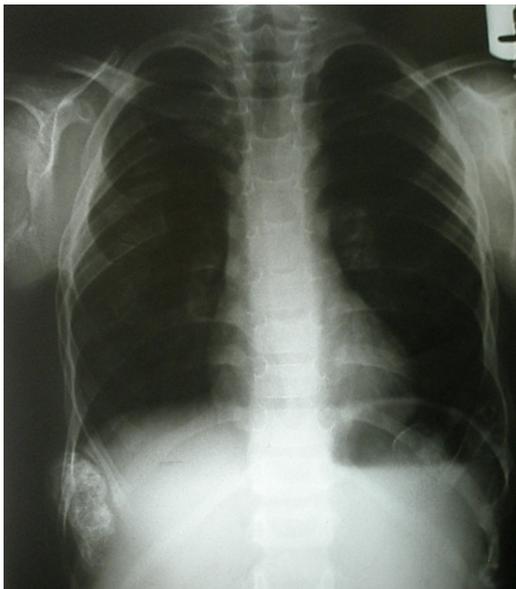


Figure 2. Chest X-ray of a seven year-old boy with heredity multiple exostosis. The massive exostosis compressed the diaphragm and liver which may lead to further complications such as hemothorax in future.

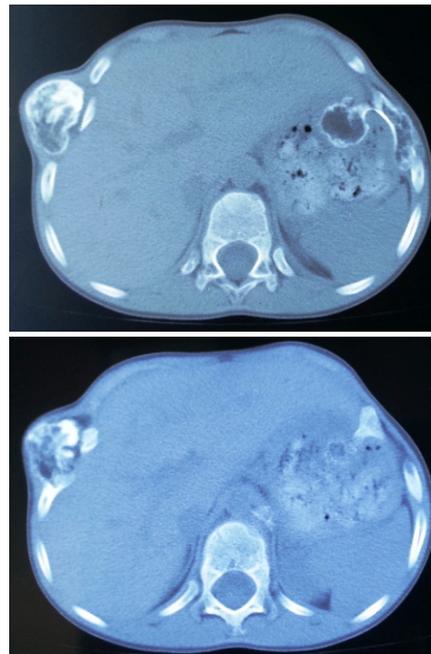


Figure 3. CT scan of the patient. Multiple exostoses are seen involving the ribs.

hemangioma and osteochondroma. A plain radiography and CT scan can most often confirm the diagnosis, and conducting a MRI study can differentiate a benign lesion from a low grade malignant cartilaginous tumor (1-2).

In most cases, exostosis is asymptomatic, but mechanical pressure of adjacent anatomical elements and the resulting degeneration might cause a symptom. The chest wall has a dynamic structure, so continuous friction against and stimulation of the exostosis during breathing and destruction of adjacent tissues may cause fatal conditions such as pneumothorax due to lung damage or hemothorax caused by damage to diaphragm, pleura, or the heart (3-4, 7-9). Takata et al. reported a case of life-threatening hemothorax caused by rib exostosis and advised that even if no complication is detected, resection should be performed after diagnosis (8).

Another 42 year-old man was reported who presented with kyphosis resulting from a giant rib exostosis (6). Huang et al. reported a 5 year-old boy with chest distress and pain similar to asthma that resolved after mass resection (5). Laceration to the diaphragm, recurrent phnomothorax, hemothorax and pricardial effusion were other complications reported in literature (3-4, 7-9). It is interesting that most of the reported patients had MHE and were male (3, 5-9). All symptoms were resolved after tumoral excision in all cases.

In the current article we present a boy who had multiple exostoses on the chest wall--an ucommon

situation. In the evaluation of medical literature, we could not find such a case with hereditary multiple exostoses involving multiple ribs in a patient who finally underwent surgery due to aesthetic and psychic concerns of the patient and family. In our case, in order to convince the child to return to school with a normal appearance, all under-skin lesions were removed. Since previously reported cases showed the probability of fatal complications following intrathoracic exostosis, we decided to remove the lesion as well in the same surgical session after consultation with the parents.

In conclusion, we not only recommend the exostosis removal for prominent clinical symptoms, but also for cases complicated by psychiatric problems, especially in child patients.

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