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Case Report

COUGH AND CHEST PAIN WITH A UNCOMMON CAUSE

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ABSTRACT

Multiple hereditary exostoses (MHE) is an autosomal dominant disordes with multiple cartilage capped bony outgrowths in tibia, fibula, femur, and sometimes the ribs and scapula. They may present with a variety of symptoms depending on the structures it compresses such as nerves, arteries or may lead to limb deformities or may cause bursitis, or may undergo malignant transformation. A 33-year-old male presented to our outpatient department with recurrent cough and left sided chest pain. On the evaluation, he was found to have multiple bony outgrowths in the scapula, fifth rib, and limbs. Similar bony outgrowths were also present in his father and grandfather. On clinical and radiological basis, a diagnosis of MHE was made. His symptoms gradually subsided with removal of the rib and scapular exostoses. Thus, the evaluation of bony structures should not be overlooked in cases of cough and chest pain.

Keywords: Cough, Chestpain, Multiple hereditary exostoses.

INTRODUCTION

Multiple hereditary exostosis (MHE) may present with solitary or multiple bony outgrowth with male preponderance. There is higher incidence of malignant transformation (10-20%). Presenting features depends upon site of bony growth. Here we present an interesting case of MHE in a middle aged male presented to our clinic with cough and chest pain due to rib exostosis. We documented MHE in male members of his family of three generation.

CASE REPORT

We present a case of cough and chest pain with an uncommon cause. A 35-year-old male patient presented at the outpatient department with a complain of cough since last 2 weeks. It was dry in nature and was not accompanied by any expectoration, hemoptysis, rhinorrhea, fever or weight loss. There was a history of similar episodes in the past 2 months. He had no history of exposure to pets or birds. He also had a history of chest pain which was localized to the back, on the left side since last 2 weeks. It was not aggravated by exertion, but it increased on moving the left arm. It was not associated with any shortness of breath or palpitation. There was no radiation to arms or neck. He is a bank employee, non-smoker, non-diabetic and non-hypertensive. There was no history of similar episodes of cough or chest pain in his family members.

On examination, his vitals were stable, and there was no pallor, cyanosis, clubbing or edema. On chest examination, there was a small bony, non-tender prominence over the left scapula. However, percussion did not reveal any abnormality. There was noisy respiration on auscultation. There were no crackles. Cardiovascular examination was within normal limit. Notably there were small bony prominences in few other sites like both upper arms and large ones on the back of both lower legs. On enquiring further, he said they were non-tender, progressing very slowly since childhood (Figs. 1 and 2). Significantly similar bony swellings had been present even in his father and grandfather.

On investigating, his hemogram and biochemistry were in the normal range. Hemoglobin 11.8 g/dl total count 8700/cm (60 N, 37 L, 3 E), erythrocyte sedimentation rate 38 mm in 1st hr, fasting blood sugar 90 mg/dl. Serum calcium 9.2 mg/dl (n=8.5-10), phosphorus 4.3 mg/dl (n=2-5), parathormone 41.1 pg/ml (n=12-72).

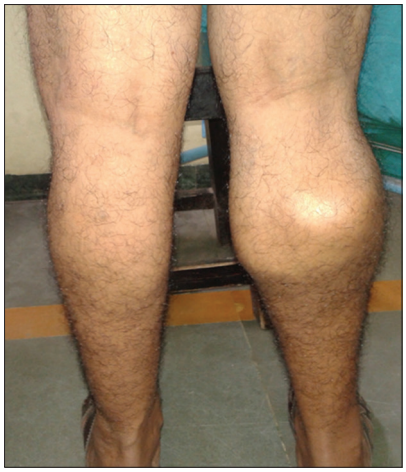
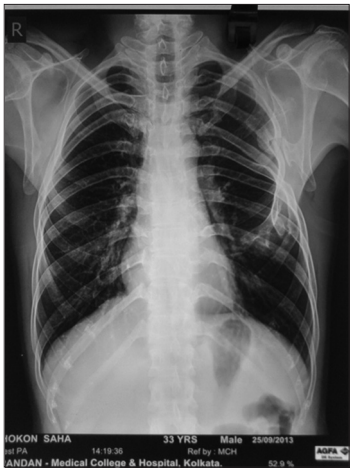
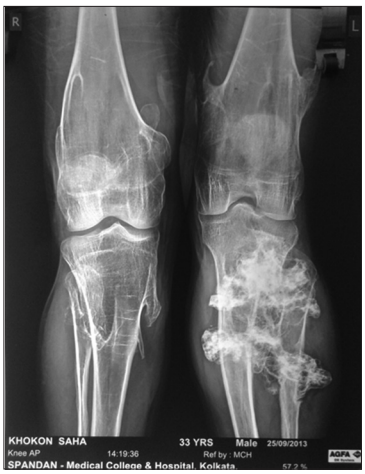


Fig. 1: Bony swellings in both lower legs

Fig. 2: Bony swellings in right elbow and wrist



X-ray plate 1: Bony swelling at left fifth rib and left scapula



X-ray plate 2: Bony swellings in both lower legs

Skiagram of chest showed the bony deformity of the left scapula and the fifth rib (X-ray plate 1) and skiagram of the legs showed the calcified bony growth of the proximal part of both tibia and fibula as well as the femurs (X-ray plate 2). Electrocardiogram and cardiac enzymes were within normal limit. It was followed by an echodiography, which was also normal.

On the clinical and radiological basis, a diagnosis of multiple hereditary exostosis (MHE) was made. His cough subsided with cough suppressants, but to recur back again along with chest pain. Hence, he was referred to the surgery department where the rib and scapular bony outgrowth were excised. Thereafter, gradually his pain in chest and cough subsided.

DISCUSSION

Osteochondroma is an outgrowth of medullary and cortical bone. A portion of the cartilaginous growth plate grows outward instead of longitudinally and forms the osteochondroma/exostosis (EXT) (like a branch on a tree). It consists of bone covered with cartilaginous cap EXT. These are benign, non-neoplastic conditions. They occur as a solitary lesion or as multiple EXT associated with a hereditary condition known as MHE. It is an autosomal dominant condition with near complete penetrance and has been associated with mutations in at least three different genes termed EXT genes [1]. It has a male predominance (3:1) and an incidence of [2] in 50,000 [2]. There is the history of hard swellings for many years. It may be bilaterally symmetric, or one side may predominate. As in our case, the EXT were present in three generation of males. And they were mostly left sided. Typically five or six EXT are found in upper and lower limbs. Most common locations are distal femur (70%) proximal tibia (70%) humerus (50%) proximal fibula (30%) [3]. Depending on their location the EXT can cause the following problems pain or numbness from nerve compression, vascular compromise, inequality of limb length, irritation of tendon and muscle, Madelung's deformityas well as a limited range of motion at the joints upon which they encroach. MHE can lead to the shortening and bowing of bones affected individuals often have a short stature. Osteochondroma of the scapula is a rare entity whose prevalence has been estimated in <5% of all ostrochondromas [4]. When settled on the dorsal surface, it may lead to palpable EXT and thorax asymmetry when settled on the ventral surface, it may result in limited motion range, winging of the scapula or snapping noise [5]. An overlying bursa may form and result in a bursitis, particularly these bursae may become inflamed and painful at sites of friction around the scapula. Rib EXT may present with cough, pneumothorax, hemothorax, etc. As in our case which presented with chest pain and cough [6,7].

There is a higher incidence of malignant transformation (10-20%) of osteochondromas that develop in MHE. Signs of transformation being sudden growth and pain [8]. This occurs most frequently in patients with an EXT1 mutation or with a tumor affecting the scapula rather than other structures [9].

Most commonly a secondary low grade chondrosarcoma develops. Diagnosis is mostly clinical and radiological. Surgery, physical therapy and pain management are currently the only options available to MHE patients. They may undergo numerous surgical procedures throughout their lives to remove painful or deforming EXT correct limb length discrepancies or improve range of motion. Our case too responded to the surgical removal of the scapular and rib EXT.

Thus, needless to say it is very important to evaluate bony deformities carefully for the cause of cough and chest pain apart from heart and lung pathologies.

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