

MASSIVE PLEURAL EFFUSION AS AN RESULT OF HEREDITARY MULTIPLE EXOSTOSES

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Abstract

Hereditary multiple exostoses (HME) is a rare, benign autosomal dominant genetic disorder, but in very rare cases they can have malignant transformation.

Exostoses can cause pain, pressure on some nerves, blood vessels and damage on surrounding tissues. Chest CT and CT angiography play an indispensable role in the diagnoses and management of exostoses.

We report an eleven-year-old boy with massive pleural effusion caused by costal exostoses on the right ninth rib. Costal exostoses represent an unusual cause of pleural effusion.

Key words: Costal exostosis, hereditary multiple exostoses, osteochondroma, pleural effusion.

Introduction

Hereditary multiple exostoses (HME) also known as hereditary multiple osteochondromas or diaphyseal aclasis is a rare autosomal dominant genetic disorder which is characterized by multiple osteochondromas that grow near juxta epiphyseal region of the bones such as the ribs, pelvis, vertebrae and markedly long bones.

The exostoses can be divided in: sessile or pedunculated. Sessile exostoses are permanently attached and broad, while pedunculated exostoses are conjoined by a narrow stem.

The most common appearance of the exostoses are around the knees and proximal humerus [1]. HME incidence is approximately 1:50000 in general population.

The genes responsible for MHE are EXT1 and EXT2 [2].

Generally all of affected individuals are diagnosed by age 12 years, the middle age is 3-4 years. Even though osteochondromas are benign in some cases they become malignant [3]. The exostoses become more prominent as the child grows but stop developing once a child hits maturity.

These patients seek for a medical help when they notice a palpable mass near the joints. Osteochondromas can cause pain, pressure on some nerves, blood vessels and surrounding tissues. [2,3].

The purpose of this paper is to report a rare case of pleural effusion caused by a multiple exostosis of the ribs.

Case report:

An eleven year old boy was admitted to ER after nausea and collapse.

His father reported that he had a spontaneous right sided chest and back pain lasting for over 2 months which in the last three days had irradiated to the right arm, and this was his only complain with no history of chest trauma.

Physical examination was done and decreased breath sounds were noted at the right sided chest.

Blood tests showed a little bit high level of leukocytes.

After that a chest CT was ordered where it showed right sided massive pleural effusion with consolidation and a costal exostosis which were comprising the heart and mediastinal structures to the left.

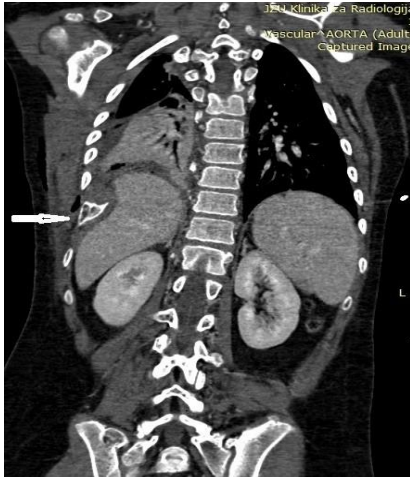


Figure.1 Chest CT coronal view



Figure2. Reconstructed chest CT image
Sharp-edged costal exostosis from 9th rib b (white arrow)

When a costal exostoses were noted ,a CT angiography was done to evaluate if there are other exostoses and to evaluate the relationship with the neighboring tissues. On the CT angiography was noted a big exostosis on the ninth rib right with intrathoracic prominence which was the cause of the right sided pleural effusion.

A lot of exostoses were noted on the body, where the one on the third rib left was with a very close relation to the hearth which can cause a pericardial effusion



Figure3. Chest CT,axial image. costal exostosis near the heart (black arrow);

Discussion

Hereditary multiple exostoses(HME) is autosomal dominant genetic disorder which is represented by exostoses of long bones usually appearing in the first decade of life and don't grow after puberty, actually after closure of the growth plate [2,4,5].

Generally the exostosis are located around the big joints, like distal femur, proximal fibula, tibia and humerus. Exostoses are usually asymptomatic but depending on their localization they can cause different symptoms, inflammation of the surrounding tissue like neurovascular compression, pain, fractures which some of this symptoms require surgery.

Those individuals with MHE who have clinical symptoms move up from approximately 5% at birth to 96% at age 12 years [6]. Exostoses can progress to a chondrosarcoma alteration (0.5%– 5% of patients),⁸ which can be notified by an increase in pain or size of the exostoses.

A plain radiography and CT scan can most often confirm the diagnosis, but requiring a MRI study can differentiate a benign lesion from a malignant cartilaginous tumor with measuring the

cartilaginous cap. A cartilage cap of over 1.5 cm in thickness is suspicious for malignant transformation [7].

Also a CT angiography should be done to be noted the correlations with the surrounding tissues.

Rib exostosis is rare and occurs in the area of costovertebral junctions or in the level of rib vertebrae [9,10].

Ribs exostoses are described between 35% and 44% of cases, depending on genotype and are usually asymptomatic [11]. However, rib exostosis can occasionally be associated with hemothorax, pneumothorax, diaphragm or pericardial injuries [12].

Repetitive irritation and stimulation of the exostosis during breathing and destroying neighboring tissue may cause fatal condition due to lung damage [9,10]. Chest pain may be the only symptom of an intrathoracic exostoses localization¹³ which can lead to serious complications as in the case we are presenting.

Conclusion

When a patient complains of a chest pain without a trauma and, we should be suspected for a rib localization of an exostosis. Chest CT must be done to prove the existence of exostosis and to evaluate relationship with the neighboring tissues.

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