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CASE STUDY

Spontaneous haemothorax caused by costal exostosis

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Spontaneous haemothorax caused by costal exostosis. K. Uchida, Y. Kurihara, S. Sekiguchi, Y. Doi, K. Matsuda, M. Miyanaga, Y. Ikeda. ©ERS Journals Ltd 1997.

ABSTRACT: We report a case of spontaneous haemothorax in a 19 year old boy with an exostosis of the left second rib. It may have been caused by nontraumatic rupture of markedly dilated pleural vessels, as a result of long-standing friction between the exostosis and the pleura.

This is the first report of spontaneous haemothorax, without penetrative injury to the pleura or the diaphragm, in a patient with hereditary multiple exostosis.

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Nontraumatic or spontaneous haemothorax is a rare condition, and sometimes occurs during anticoagulant therapy for venous thromboembolism and pulmonary embolism. Other causes of the condition are bleeding disorders, complication of spontaneous pneumothorax, pleural malignancy, and rupture of the thoracic aortic aneurysm [1]. Furthermore, costal exostosis also causes a haemothorax due to direct traumatic injury of the pleura or diaphragm by the intrathoracic tumour [2–5].

We present a case of spontaneous haemothorax in a patient with hereditary multiple exostosis (HME), and propose a new mechanism of haemothorax.

Case report

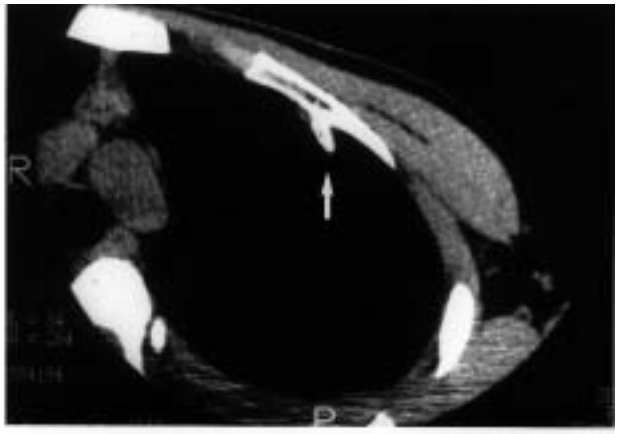
A 19 year old boy was admitted to hospital with a 6 day history of left-sided pleuritic chest pain. His history of recent trauma was unclear. There was no cough or fever. He had previous excisions of histologically proven cartilaginous exostoses. His father also had a history of multiple exostoses.

Chest radiography showed left pleural effusion and suspect exostosis of the right third rib, and of the left scapula. Thoracocentesis was performed and 20 mL of noncoagulated bloody fluid, with a haematocrit of 48%, was removed. No tumour cells were present. Gram stain of the fluid sediment showed no microorganisms, and cultures for bacteria and fungi produced no growth.

The patient's haemoglobin concentration was 128 $g \cdot L^{-1}$, haematocrit 36%, white blood cell count 4.4×10^9 $cells \cdot L^{-1}$, and there were 271×10^9 $platelets \cdot L^{-1}$. Clotting studies produced normal results.

Although the exostosis of the left second rib was not evident on the chest radiograph, thoracic computed tomography (CT) scan showed the exostosis from the left second rib anteriorly, with a long bony spicule projecting inward to the lung (fig. 1a). A parapleural cystic lesion

a)



b)

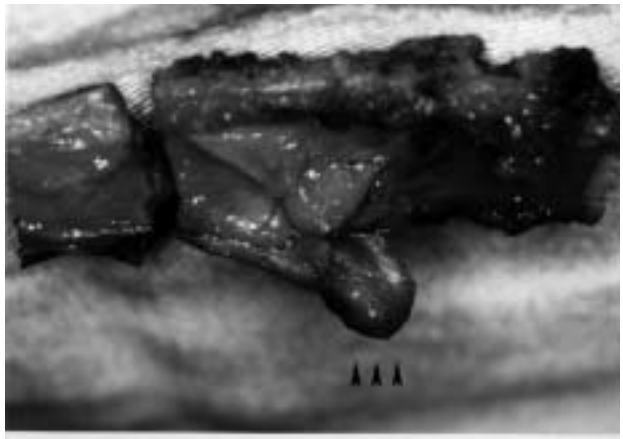


Fig. 1. – a) Computed tomography (CT) scan showing exostosis from the left second rib (white arrow). b) The resected rib with the exostosis (arrowheads).

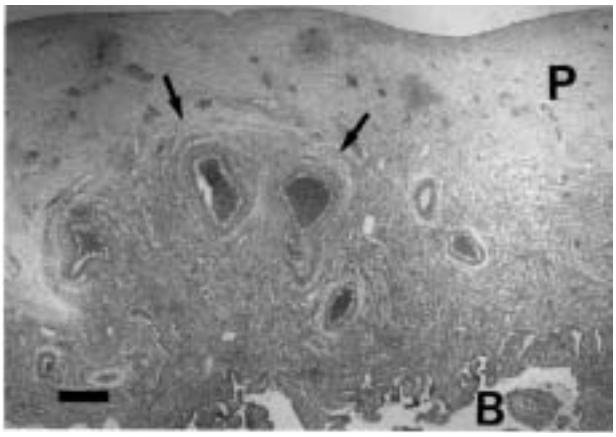


Fig. 2. – Dilated arteries (arrows) are seen in the thickened pleura. P: pleura; B: bronchiolectasis. (Haematoxylin and eosin stain; internal scale bar=250 μm).

and thickened pleura were also seen near the exostosis. Further radiological investigation showed other exostoses in the right femur, tibia, fibula, ulna and both phalanges.

Ten days later, chest roentgenograms showed less fluid, without repeated thoracocentesis or tube drainage. Surgery was necessary to establish the diagnosis, and to prevent further problems in the future. During the operation, a bony exostosis was found arising from the posterior aspect of the anterior portion of the second rib. There was no clot or active source of bleeding, but marked thickening of the visceral pleura was noticed, with several dilated vessels. The part of the rib with the inward projecting exostosis, which was a 1.5 cm length mass of bone covered by a cap of cartilage, was resected (fig. 1b).

Histologically, the tumour was osteochondroma, and there was no evidence of malignancy. In the partially resected lung tissue, there was marked fibrosis of the pleura and subpleural lung parenchyma, in which dilated muscular arteries and bronchiolectatic cysts were seen (fig. 2). In the postoperative period, the patient recovered completely and was discharged 8 days after operation.

Discussion

HME is an autosomal dominant condition, characterized by multiple exostoses, usually seen in the long bones [6]. There are a few reported complications, including popliteal aneurysm, haemarthrosis, central or peripheral nerve compression, and urinary obstruction. Haemothorax is a rare complication and has only been described in eight patients (six HME, and two solitary) in the English literature [2–5, 7, 8]. The aetiological mechanisms proposed by most authors are the shearing of the pleura or diaphragm by the relatively sharp margins of the intrathoracic exostosis.

The present case suggested another cause of haemothorax associated with costal exostosis. Surgery revealed no active source of bleeding; however, there were several dilated vessels with prominent thickening of the visceral pleura facing the exostosis. The parietal and visceral pleurae were neither lacerated nor punctured. We thought focal pleural changes were induced by long-standing friction between the intrathoracic exostosis and the visceral pleura due to respiratory motion, and spontaneous rupture of the dilated vessels might result in a haemothorax.

Although exostosis is an obvious pathology of the thoracic bony cage, it is not easy to identify on the plain chest radiograph whether the exostosis has intrathoracic direction or extrathoracic direction. In the present case, thin-section CT scan clearly showed that the exostosis pointed toward the lung, and revealed additional information, such as thickened pleural and bullous cysts adjacent to the exostosis.

According to previous reports [2–5, 7, 8] all patients with spontaneous haemothorax secondary to HME were less than 23 yrs of age. Differential diagnosis of the spontaneous haemothorax in young people revealed native coagulopathy, such as haemophilia, thrombocytopenia and von Willebrand's syndrome [1].

Hereditary multiple exostosis should be considered in cases of nontraumatic haemothorax, without coagulopathy, in young patients.

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