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Case Report

An Unusual Case of Spontaneous Hemothorax in an Adolescent Affected by Congenital Exostosis: Case Report and Literature Review

Giangiacomo Nicolini[1*], Giulio Bove[2], Monica Micera[1], Alessandra Bet[3] and Fabio Ricagna[4]

[1]*Unità Operativa Complessa di Pediatria e Patologia Neonatale, Ospedale San Martino, Belluno, Italy*

[2]*Dipartimento Salute della Donna e del Bambino, Scuola di Specializzazione in Pediatria, Università di Padova, Padova, Italy*

[3]*Country Paediatrician, Italy*

[4]*Unità Operativa Complessa di Chirurgia, Ospedale San Martino, Belluno, Italy*

***Corresponding Author:** Giangiacomo Nicolini, Unità Operativa Complessa di Pediatria e Patologia Neonatale, Ospedale San Martino, Belluno, Italy.*

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Abstract

We present the case of a male adolescent with a haemothorax caused by pericardial rupture secondary to costal exostosis. The patient is affected by multiple congenital exostosis and in 2014 he underwent a bone spur resection of the right femur. An X-ray performed at presentation was interpreted as suggestive of pneumonia and the patient was therefore treated with antibiotic without any clinical improvement. A thoracic ultrasonography performed 7 days later showed a massive haemothorax, confirmed by a CT scan on which a pericardial lesion caused by the growth of three osteocartilagineous spurs originating from the 5th, 7th and 8th rib was detected. The boy underwent surgery and recovered completely. The thoracic ultrasound and the subsequent CT-scan performed on the patient due to his lack of clinical improvement directed us towards a rare disease that the X-ray had not helped diagnose; this entailed the prompt surgical resolution of a possibly severe, maybe even fatal, condition.

***Keywords:** congenital exostosis, hemothorax, thoracoscopy, pericardial rupture*

Introduction

Most cases of haemothorax are related to blunt trauma, procedures, neoplasm such as schwanommas in von-Recklinghausen disease and soft-tissue tumours, and vascular ruptures. Spontaneous pneumothorax is also a cause of spontaneous haemothorax. It is rarely due to costal exostosis; in these cases, all affected individuals are usually diagnosed by age 12 years, but the median age of diagnosis is 3 years [1-3, 5-6, 9-12, 16-19, 21-23]. Haemothorax may result in respiratory distress, respiratory failure, retained clot, fibrothorax, empyema and extended hospitalization.

A costal exostosis is a benign tumoral growth capped by cartilage, which protrudes from a rib. It may sometimes require emergent surgery 21 due to an associated complication such as damage to an intrathoracic organ (e.g. lung, pleura, pericardium, diaphragm)[16–23]. Some cases of evolution into chondrosarcoma have also been reported [24].

In this article we report a case of spontaneous haemothorax due to a pericardial lesion caused by bone spurs in the context of multiple costal exostosis.

Case Report

A 13-year-old boy (weight 45 Kg, height 162 cm) with no previous history of trauma was seen by his general practitioner for a complaint of pain in the left hemithorax during respiratory acts (no fever and no cough were reported) and after a physical examination she recommended a chest x-ray which seemed to show left pneumonia with effusion (Fig.1). The patient started antibiotic therapy with a high dose of amoxicillin and after one week he was seen again by his paediatrician: he had no fever or cough but he looked pale and weak, and lung examination still revealed a reduced air entry; heart rate, blood pressure and pulse oximetry were normal.

He was referred to us to validate the hypothesis of pneumonia with thoracic ultrasound, but the imaging showed hypoechoic corpuscular pleural effusion that looked like blood. A second chest x – ray showed a worsening of the pleural effusion (Fig. 2). A more extensive medical history analysis revealed that the boy was affected by familial congenital exostosis, that 6 years earlier he had undergone a bone spur resection on the right femur and had a scheduled intervention for the following month on his left calf (Fig. 3). The boy's father, the paternal grandmother, great-uncle and great-grandfather presented with the same condition and received surgery mainly for aesthetic reasons. The father had also been operated for a lesion of the femoral artery caused by a bone spur. We therefore performed a chest CT-scan that confirmed our hypothesis and showed a small pericardial lesion caused by an abnormal growth of three ribs, 5th, 7th and 8 th (Fig 4,5). Biochemical examinations revealed anaemia with a hemoglobin level of 10 g/dl. A video – thoracoscopy was then performed to identify the site of the intrathoracic bleeding and treat the spontaneous hemothorax. The patient being under general anesthesia, with selective left lung exclusion and full right lateral decubitus, a 11 mm trocar was placed at the 8th left intercostal space along the posterior axillary line. Upon entering the pleural space, a large hemothorax was found that was evacuated with immediate improvement of the hemodynamic parameters and oxygenation. A 10 mm/30 endoscope was introduced into the pleural cavity. Three osteo – cartilagineous spurs originating from the 5th, 7th and 8th rib along the mammillary line were found to protrude into the pleural cavity (fig 6,7). The lowest spur caused a tear of the pericardial fat with active bleeding. Two more torcars were then placed under direct vision: a 5 mm one at the 4th intercostal space along the posterior axillary line and a 11 mm one at the 6th intercostal space along the mid axillary line. The haemostasis on the pericardial fat was secured with bipolar coagulation. All of the three osteo – cartilagineous spurs were removed using a long Kerrison rogeur and their bases were carefully cauterized. In the postoperative period a pneumothorax occurred that was treated with drainage for 2 days and progressively resolved without complications. On the 5th post-operative day he was discharged and an ultrasonographic follow-up after 2 weeks showed a complete reabsorption of the pneumothorax and normalization of the pulmonary picture. After one month ultrasound was still normal and the haemoglobin level was normalized.

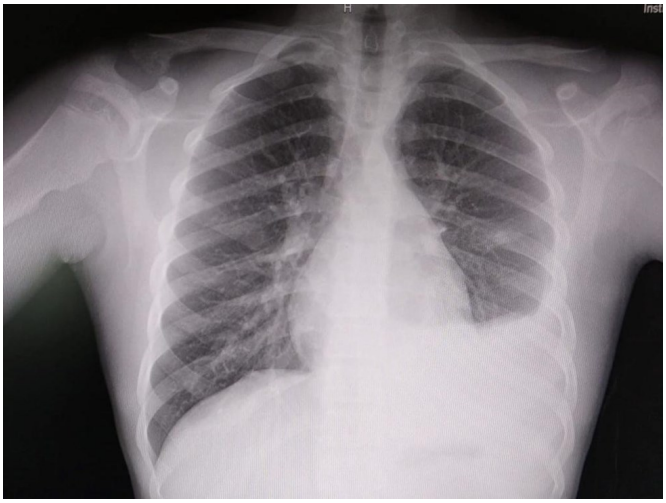

Fig 1: *Pulmonary thickening, reported as pneumonia.*

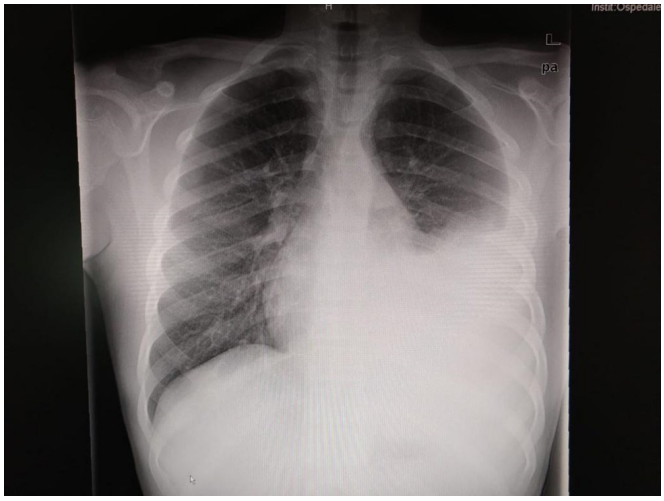

Fig 2: *Worsening of pleural effusion.*

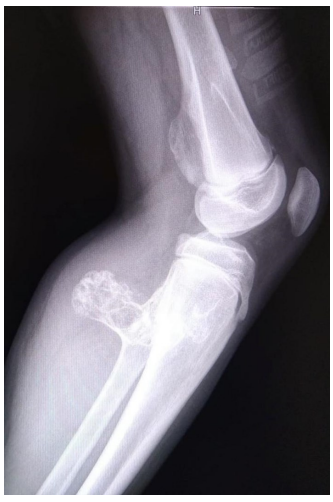

Fig 3: *Left calf exostosis.*

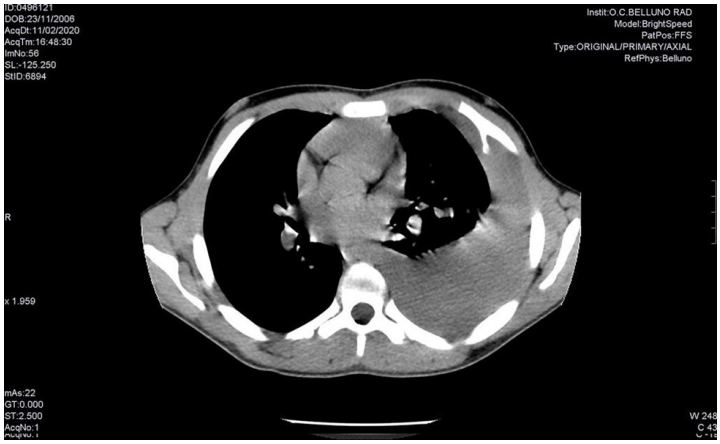

Fig 4: *CT scan showing pericardial lesion caused by an abnormal growth of three ribs, 5th, 7th and 8th.*

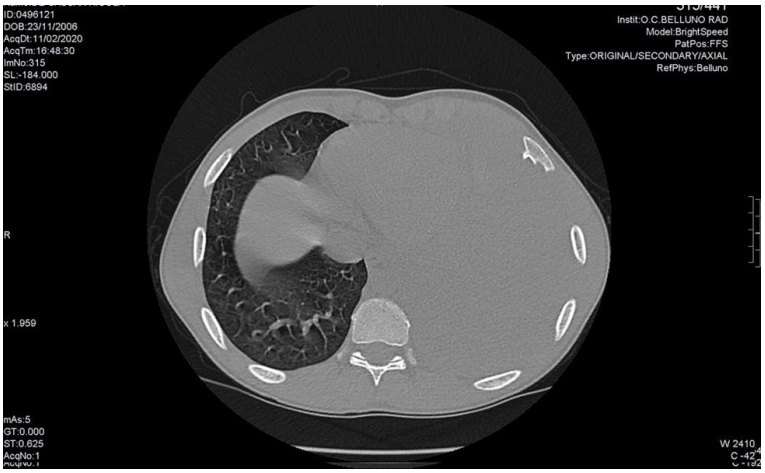

Fig 5: CT scan showing hemothorax.

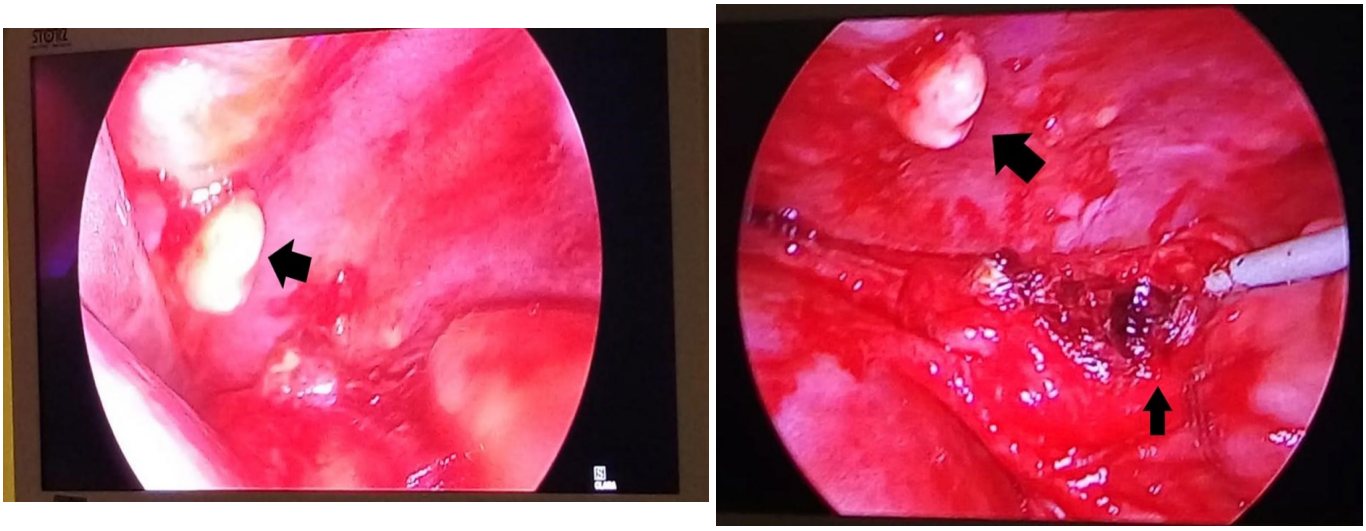

Fig 6&7: Thoracoscopy showing bleeding and exostosis.

Discussion

We described the case of a young male patient with a hemothorax, caused by a rib exostosis that was initially visualized on a CT-scan and then confirmed during videothoracoscopy.

Exostosis accounts for 50% of primary benign bone tumors and occurs at the metaphysis of long bones or originates from the surface of flat bones [4]. It generally occurs at the proximal femoral or distal tibial metaphysis [1]. Exostosis of the rib constitutes 1 – 2% of all the cases of exostosis [4].

Primary exostosis occurs as solitary or multifocal lesions. Multiple exostosis disease is a rare hereditary disorder that occurs in infants [5, 6] and is characterized by the formation of exostoses with many localisations. It is associated with mutations in the EXT genes and presents with skeletal deformities caused by abnormal bone growth [5]. The pathogenesis of costal exostoses is unclear, but it appears that developmental growth defect of the fibrous tissue (perichondrium) covering the epiphyseal plate may result in the lateral growth of the epiphyseal cartilage plate instead of the normal downward growth towards the metaphysis. This abnormal growth leads to an inward protrusion of the rib cartilage [3]. In our case both the father, the paternal grandmother, the brother of the paternal grandmother and the grandfather of the father were affected by similar conditions; none of them had performed a genetic test but we think it's very likely to be a congenital familial exostosis.

Solitary exostosis occurs in both infants and adults and is generally asymptomatic. In our case, the X ray suggested that the boy had a complicated pneumonia, leading us to a wrong diagnosis. An ultrasonography and a chest CT-scan could be useful to clarify the clinical picture. Symptomatic cases of costal exostosis have presented with swelling, hiccups [7], chest pain [7], pneumothorax [4, 8], or hemothorax [1, 2]. Hemothorax is thought to result from trauma caused by the costal exostosis to the pleura [6], diaphragm [1, 9], lung [10, 11], or heart. Patients at risk of life-threatening damage to the intrathoracic organs should receive (o undergo) surgical resection [12].

Past reports of organ damage due to costal exostosis have described longitudinal lacerations of the diaphragm and pericardium adjacent to the tip of the exostosis that were caused by respiratory movements[13].

Malignant transformation of an exostosis to chondrosarcoma [14] is relatively rare in adult and extremely rare in children; the risk of a solitary exostosis transforming to chondrosarcoma is 1–2 %, and for multiple osteocartilaginous exostoses is 5–2 %. Chondrosarcoma should be suspected if a lesion continues to grow after puberty or localized pain develops [4].

In our case, the costal exostosis caused the lesion of the pericardium, which led to bleeding and progressive deposition o build-up of blood in the pleural space.

We found that 22 cases of pediatric hemothorax caused by costal exostoses have previously been described in literature (tab 1). The patients ages ranged from 3 to 17 years (16 were male and 6 were female). Five patients had bleeding caused by a punctured diaphragm, 6 had a punctured lung/pleura, 3 had a punctured pericardium, 1 a tear in pericardium and diaphragm, 2 lung/diaphragm punctures and in 5 patients the exact source of bleeding was not found. In most cases, the onset symptom was chest pain (9 patients), sometimes accompanied by dyspnoea (9 cases), 1 case presented with dyspnoea and 2 cases were asymptomatic.

Table 1: Congenital exostosis: cases described in literature.

First Author	Sex	Age	Clinical presentation	Imaging	Treatment	Damaged tissues
Kameda T [16]	M	16	asymptomatic	Rx, TC	Thoracotomy	Pericardium
Lin CY [17]	M	12	Pain + dyspnea	Rx, TC, Thoracentesis	Thoracotomy	Diaphragm
Martino A [18]	F	13	Pain	RX, TC, Thoracentesis	Thoracotomy	Lung, Diaphragm
Matsushima K [19]	M	13	Pain + dyspnea	Rx, TC, Thoracentesis	Thoracotomy	Lung, Diaphragm
Mann E [20]	M	17	Pain	Rx, TC, Thoracentesis	Thoracotomy	pleura
Ravindran R [21]	M	14	Pain, cough, intermittent fever, dyspnoea	Rx, ultrasound, Thoracentesis	Thoracotomy	-
Assefa D [22]	F	14	Pain + dyspnea	Rx, TC, Thoracentesis	Thoracotomy	Diaphragm
Assefa D [22]	F	6	Pain	Rx, TC, Thoracentesis	Thoracotomy	Pericardium
Matsuno Y [23]	M	3	Pain + anemia	Rx, TC	VATS	Pericardium
Takata K [24]	M	4	Pain	Rx, TC, Thoracentesis	Thoracotomy	Pleura, lung
Huang HR [25]	F	9	Pain	Rx, TC, Thoracentesis	-	-
Cowles RA [26]	F	6	Pain + dyspnea	Rx, TC, Thoracentesis	VATS	Diaphragm, pericardium
Dendale J [27]	M	12	Pain	Rx, TC, Thoracentesis	-	-
Teijeira FJ [28]	M	7	dyspnea	Rx, TC, Thoracentesis	Thoracotomy	Diaphragm
Kuo SM [29]	M	15	Fever, Pain, cough, dyspnea	Rx, TC	VATS	-
Huang L [30]	F	5	Pain + dyspnea	Rx, TC	VATS	pleura
Pham-Duc ML [31]	M	15	Pain + dyspnea	Rx, TC, Thoracentesis	VATS	pleura
Simansky DA [32]	M	17	Dyspnoea, syncope	Rx, TC, Thoracentesis	VATS	Diaphragm
Tomares [33]	M	3	Asypomatic	Rx, TC, Thoracentesis	VATS	Lung
Reynolds [34]	M	14	Pain + dyspnea	Rx, Thoracentesis	Thoracotomy	Diaphragm
Propper [35]	M	16	Pain	Rx, Thoracentesis	observation	-
Propper [35]	M	9	Pain	Rx, Thoracentesis	Thoracotomy	Lung

Resection was performed in 12 patients by thoracotomy, in 7 patients by thoracoscopy, and 3 patients were observed without surgery.

To the best of our knowledge our case is the fifth report of costal exostosis causing a hemothorax as a result of injury to the pericardium, and he was the eighth patient treated with a thoracoscopic approach.

The etiological mechanisms proposed by most authors are shearing of the pleura, diaphragm or pericardium by the relatively sharp margins of the intrathoracic exostosis. The present case revealed no active source of bleeding; there was however a prominent thickening of the pericardium facing the exostosis. The pleura and diaphragm were neither lacerated nor punctured. We thought the focal pericardial changes were induced by the long-standing friction between the intrathoracic exostosis and the pericardium due to respiratory motion, and spontaneous rupture of the pericardial vessels might have resulted in a hemothorax. Although pre-operative diagnosis on plain chest radiograph is usually difficult, careful examination of a chest CT scan may allow the presence of a bony spur to be detected. Magnetic resonance imaging may be used as an alternative mean of imaging and is particularly useful in cases of Hereditary multiple exostoses where many instrumental studies are necessary and several CT scans may expose a young patient to high doses of radiation. Magnetic resonance imaging is also preferred when an osteochondroma causes spinal cord compression because of its ability to evaluate the size of the lesion and its relationships with neural structures. Disadvantages of routinely using magnetic resonance imaging to evaluate these lesions include its high cost, limited availability, and contraindications in some patients who are claustrophobic or have implanted ferromagnetic foreign bodies [4].

In the present case the chest CT scan with 3-dimensional reconstruction clearly showed that the exostosis pointed inward to the thoracic cavity, thus proving to be not only a useful diagnostic tool but also of assistance in the clinical management (e.g. thoracoscopic vs surgical vs conservative approach).

Conclusion

This article presents the case of a male adolescent with hemothorax secondary to pericardial rupture caused by costal exostosis that was initially misdiagnosed as pneumonia with associated pleural effusion.

Because of the small number of paediatric cases reported such an error may be very easy to make. In our case, an approach based on the use of thoracic ultrasound and CT-scan after the lack of improvement with the first-line therapy, proved to be crucial in consideration of the potentially lethal consequences of a further delay in the diagnosis.

Conflict of Interest

The authors declare no conflict of interest.

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