Aneurysmal bone cyst presenting as an intrathoracic mass in an infant

Maninder S. Kalkat*, Frank J. Collins

Department of Thoracic Surgery, Birmingham Heartlands Hospital, Bordesley Green East, Birmingham B9 5ST, UK

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Abstract

A 9-week-old child presented with a rare aneurysmal bone cyst of the rib masquerading as a left intrathoracic mass displacing the mediastinum and diaphragm. This was managed successfully with excision of the mass and reconstruction of chest wall.

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1. Introduction

Chest wall tumours are rare in infants. They are usually highly malignant and mostly consist of sarcomas. Primary aneurysmal bone cyst occurs most commonly in teenagers (80%) and rarely involves the ribs [1]. The management of chest wall masses involves en bloc excision of the tumour with the adjacent chest wall and reconstruction of the resultant defect.

2. Case report

A 9-week-old baby girl was discovered to have hepatosplenomegaly on routine examination and was referred to the local children’s hospital for further management. The child had normal antenatal and natal history. She was discovered to be tachypnoeic with mild subcostal recession. The lower borders of liver and spleen were two to three fingers breadth below the costal margin. No other abnormality could be detected on clinical examination. The ultrasound examination revealed normal sized liver and spleen without any focal abnormality. The X-ray and ultrasound examination of chest was suggestive of a mass in the left thoracic cavity. CT scan showed a fairly rounded mass lesion within the left hemithorax displacing the heart to the right (Fig. 1). The mass was of mixed attenuation with cystic, solid and calcific components and associated with destruction of axillary portion of left third rib. Minimal enhancement at the periphery of mass was seen after the injection of contrast. A presumptive diagnosis of intrathoracic mesenchymal tumour was made and the child underwent surgery.

A left posterolateral thoracotomy was carried out and chest entered through fifth intercostal space. An 8 cm × 4 cm, hard lobulated mass was seen arising from the inner surface of the lateral chest wall in the region of the third and fourth rib, encroaching on to the fifth one. It was adherent to the lung parenchyma, displacing the heart to right and pushing the diaphragm down. The lower part of the mass was multicystic and apparently full of old dark blood.

A 5-cm block of the lateral bony cage of chest wall including third to fifth ribs were excised along with the mass (Fig. 1 inset). The defect in the chest wall was closed using methylmethacrylate cement sandwiched between the two layers of vicryl mesh and securing this composite material with interrupted mattress sutures of vicryl 1-0 suture to the surrounding bony and soft tissue structures. The composite graft was tautly stretched at the time of insertion and was covered with overlying muscle, subcutaneous tissue and skin. The patient made an uneventful recovery and was discharged home in about a week’s time. The chest X-ray performed later demonstrated well expanded lungs and the composite graft incorporated into the chest wall (Fig. 2).

Microscopic examination of the specimen revealed multiple, large blood filled spaces surrounded by multinucleate giant cells with foci of haemosiderin deposition
and fibrosis. Cartilage was seen in areas with a pattern of maturation, calcification and early osteoid formation. The lesion was surrounded by fibrous capsule and was arising from the 4th rib segment. These findings were consistent with an aneurysmal bone cyst of rib.

3. Discussion

In children, tumours of the ribs, sternum and extrapleural soft tissues of the chest wall are rare occurrences. These are usually highly malignant lesions, the majority being comprised of sarcomas [2]. Aneurysmal bone cyst is a benign non-neoplastic proliferative tumefaction of bone characterized by presence of channels and spaces of varying sizes surrounded by delicate walls. This term coined by Jaffe and Lichtenstein, however, is a misnomer as this lesion is neither an aneurysm nor a cyst [3]. The walls of the multiple channels and loculi are composed of fibrous tissue, do not contain the elastic tissue and muscle found in the blood vessels. The aetiology of these cysts is unknown, but is believed to be reactive lesions in response to some haemodynamic disturbance in the rich capillary network of the host bone resulting in an expansile destructive process [4]. It can manifest as a primary form in a normal bone or a secondary form, which is associated with various bony neoplasms. These cysts occur in older children, adolescents and young adults and are very rare in children under 4 years of age. Aneurysmal bone cysts can involve any bone, the most common site being ends of long tubular bones of limb followed by the vertebral column. They have not been very commonly reported from the ribs.

Local pain is the most common presenting symptom and if the involved bone is superficial, a tender swelling may be palpable. A bruit may be transmitted from a large aneurysmal cyst. The progressive enlargement of cyst in the vertebral column can cause cord and root compression leading to neurological deficit.

Although the lesion is benign, it is locally destructive with a high propensity for recurrence. Recurrence occurs in 10–20% of cases and the incidence decreases with increasing age of the patient. The treatment consists of curettage, marginal excision, or rarely, wide excision. In bones deemed expendable, such as ribs or fibula, wide resection is recommended [5].

Apart from chest wall tumour in an infant being highly suspicious of malignant lesion, an untreated bulky chest wall tumour may also interfere with lung function due to a mass effect [6]. Hence it requires excision of mass en-bloc with chest wall involved. At the time of chest wall resection, even though large segments of bony cage are removed, appropriate reconstruction results in reasonable chest wall stability [6]. A variety of techniques have been developed for this purpose and include autogenous tissues like fascia lata, rib grafts, omentum and latissimus dorsi muscle flap [7]. Prosthetic materials have been used and include metallic plates, acrylics and synthetic mesh fabrics (Marlex, Prolene, Gortex and Vicryl) [7]. These mesh fabrics can be used alone or as a composite graft in conjunction with methyl methacrylate or bone cement to provide extra stability to the chest wall [8]. To establish appropriate chest wall stability for adequate independent breathing, the synthetic mesh should be tautly stretched at the time of insertion by securing to the surrounding structures [9]. It is essential to obtain soft tissue coverage of the synthetic material with overlying muscle, subcutaneous tissue and skin. The prosthesis is well tolerated and gets incorporated in the chest wall tissues as the child grows.

Fig. 1. The preoperative CT scan of thorax depicting intrathoracic mass and the resected specimen (inset).

Fig. 2. Postoperative chest X-ray with composite graft in situ.
4. Conclusion

The management of the aneurysmal bone cyst arising from ribs in children, a rare entity, or any other tumour in relation to chest wall consists of adequate wide resection and reconstruction of the chest wall. The aim is complete removal of the tumour, restoration of adequate protection to the thoracic viscera, restoration of physiological function, providing for adequate lung and chest wall growth and an acceptable chest wall appearance [10].

References