Thoracic Outlet Syndrome Caused by Hydatid Cyst of the First Rib-Rare But Important

Dan Levy Faber · Lael-Anson Best · Daniela Militianu · Alon Ben Nun

Received: 30 July 2008 / Accepted: 2 January 2009 / Published online: 16 November 2010
© Association of Surgeons of India 2010

Abstract Hydatid cysts are usually located in the liver and lungs. Skeletal echinococcosis is relatively rare and that of the rib is exceptional. Less than 50 cases of costal echinococcosis have been reported in the literature so far. To our knowledge, only one case report of thoracic outlet syndrome due to echinococcal cyst in the first rib was described in 1995. Accurate pre-operative diagnosis is important but may be challenging in some cases. Reported here is a case of echinococcosis of the first rib in a young adult who was presented with thoracic outlet syndrome. Plain chest radiograph, CT scan and MRI were performed. The imaging features were suggestive of a solitary aneurysmal bone cyst and the differential diagnosis included echinococcosis of the first rib. The lesion was completely resected and the histopathological examination confirmed the diagnosis of echinococcosis.

Keywords Thoracic outlet syndrome (TOS) · Skeletal echinococcosis · Hydatid disease

Introduction

An 18 year old adolescent was admitted to our hospital suffering from painful swelling in the right shoulder. Her imaging workout demonstrates a lytic lesion on her first right rib. The findings match the diagnosis of thoracic outlet syndrome (TOS). The differential diagnosis was wide and included a variety of primary and secondary

tumors of the bone. Her preoperative provisional diagnosis was aneurysmal bone cyst. We performed a resection of the first right rib using a subclavicular approach. The operative findings led us to suspect the diagnosis of hydatid cyst. Histopathological examination confirmed this diagnosis.

Clinical History

An 18 year old adolescent, resident in the rural area, was referred to our hospital for painful swelling in the right shoulder. Medical history revealed that her symptoms began almost a year ago with occasional pain in her right shoulder and arm. She continued complaining of neck pain, positional right shoulder pain, right arm pain, and paresthesias, which became more intense a few weeks prior to her admission. No evidence for motor weakness was noticed. Thoracic outlet syndrome consists of compression of the subclavian vessels and brachial plexus at the superior aperture of the thorax [1]. Thoracic outlet symptoms usually involve nerves or blood vessels, and sometimes can involve both. The patient was clinically diagnosed with TOS. The patient's laboratory investigations were normal. Plain chest X-ray showed a lytic lesion in the first right rib and bone scan demonstrated a single "hot" spot in the anterior part of the first right rib. MRI revealed a solitary cystic lesion of $3.8 \times 3.2 \times 3.1$ cm on the anterior part of the first right rib with thinning of the bone cortex (see Figs. 1 and 2). It seemed that the cyst produced significant pressure on the brachial plexus. The cyst proximity to the subclavian vessels was typical of thoracic outlet syndrome. The provisional diagnosis was aneurysmal bone cyst.

D. Levy Faber (✉) · L.-A. Best · D. Militianu · A. Ben Nun
Rambam Health Care Campus,
Haifa, Israel
e-mail: d_faber@rambam.health.gov.il

Fig. 1 Transverse CT image with intravenous contrast enhancement demonstrating expansile cystic lesion of anterior aspect first rt rib. The lesion is causing anteriosuperior displacement of the subclavian vessels

Operative Procedure and Post Operative Details

A modification of the anterior supraclavicular approach reported by Harold c. Urschel, Jr was used [1]. This approach is usually saved for cases requiring arterial bypass therapy. The main idea of the original technique is using a transverse supraclavicular incision to gain a proximal subclavian arterial control as preparations for axillary or brachial anastomose. Since our problem was the mass anterior—medial location rather than vascular involvement, we preferred a Subclavicular incision. This incision allows us a relatively safe approach while dissecting towards the lesion. Inflammatory adhesions

Fig. 2 Sagittal MR image FSE T2 FAT SAT with gadolinium injection demonstrating expansile cystic lesion of rt first rib, causing compression of subclavian vessels and brachial plexus

Fig. 3 (a-b) pathology slides from the operation, **a**: parasite scolex and typical membrane; **b**: bone inflammation and destruction due to parasite membrane

made the dissection difficult and a limited anterior dislocation of the sterno-clavicular joint was utilized to gain better exposure. We continued to dissect along the anterior border of the first rib exposing and retracting the blood vessels and the pleura. After we exposed the lesion we resect the first rib with 2 cm border from each side of the lesion. During the operation due to the cystic membranous appearance of the lesion we suspected the diagnosis of echinococcal cyst. Histopathological examination confirmed the diagnosis of hydatid cyst (Fig. 3). The post operative period was uneventful. One month following the operation, the patient was healthy, pain free and started her shoulder physiotherapy.

Discussion

Hydatid cyst is a parasitic infection caused in most cases by the tape worm *Echinococcus granulosus*. The disease is typical in residents of rural areas especially those who come

in close contact with farm animals [2]. The definitive hosts, usually dogs, eat viscera containing larvae of the parasite. Eggs, created by the larvae in the definitive host's gastrointestinal tract, find their way to the animal feces. The intermediate hosts, usually sheep, ingest the eggs and enzymatic activity releases the parasite from the eggs. The parasite perforates its way to the lymphatic and blood circulation, and continues its movement to various locations. The parasite creates a cyst in which the larvae reproduce. Humans are infected as incidental host by ingestion of contaminated water, food, or by close contact with definitive or intermediate hosts. In humans the liver is the most common site of echinococcal cyst, (50–77%), followed by the lungs. Bone cysts are rare and represent approximately 0.5–2% of all cases [3, 4]. The lesion tends to appear in the more vascularized areas of the bones [4]. We do not know what causes the parasite to enter the bone. The process starts when blood borne scolex reaches the skeletal system. The parasite grows in the bone and replaces the normal tissue until it destroys the bone cortex and infiltrates the surrounding tissue. Rib cysts are only about 6% of all the bone involvement [3]. Rib involvement is usually on the posterior segment of the bone. It can be either intra or extra osseous in nature [3]. The patient presented here suffered from TOS caused by an intra osseous lesion in the anterior medial segment of the first rib. According to a computerized literature search this case is the second description of echinococcal cyst in the first rib and both are presented by TOS [5].

The diagnostic process for suspected hydatid cyst usually involves imaging techniques and serological tests. The typical characteristics of hydatid cysts in the lung and liver are contrast enhancement, calcifications, germinal membrane detachment and daughter cysts [3]. Unfortunately these specific manifestations of the cysts are absent in the bone lesions. Findings such as pathological fractures, cortical thinning, sclerosis, honeycomb appearance, bone expansion and soft tissue extension may be evident but are not pathognomonic [3, 4]. Because there are no typical appearances of echinococcal bone cysts the differential diagnosis is wide and includes fibrous dysplasia, tuberculosis, aneurysmal and simple bone cyst, plasmacytoma, osteosarcoma, chondrosarcoma

and a variety of other primary and secondary tumors of bone [3].

The treatment of echinococcal bone cysts is similar to the treatment of malignant lesion, much more than to the treatment of infectious diseases. A radical removal of the lesion with wide excision is the main component of treatment. The uncommon location of the lesion, with brachial plexus compression and proximity of major blood vessels creates a surgical challenge in this case. We used an anterior subclavicular approach. This approach allows a safe removal of the first rib with optimal exposure of the subclavian vessels and the brachial plexus. We preferred this approach over the more common transaxillary one due to the antero-medial location of the lesion [1].

The transaxillary approach allows less retraction of the brachial plexus or blood vessels, but since the surgical wound is deeper it offers less exposure of the lesion in our case.

Conclusion

This unique case is a good example of the obscure nature of echinococcal bone cysts which requires a high index of suspicion during the preoperative diagnostic stage. Hydatid tends to destroy the bone matrix and to infiltrate the adjacent tissues [2]. In this case complete resection of the affected rib with careful protection of the neurovascular bundle lead to an excellent outcome.

References

1. Urschel HC (2005) Thoracic outlet syndrome. In: Shields TW, Locicero J (eds) General thoracic surgery, 6th edn. Lippincott Williams & Wilkins, Philadelphia, pp 689–697
2. Nurettin K, Metin G, Atilla E (2001) Hydatid disease of rib. *Ann Thorac Surg* 71(1):372–373
3. Tuzun M, Hekimoglu B (2002) CT findings in skeletal cystic echinococcosis. *Acta Radiol* 43(5):533–538
4. Gossios KJ, Kontoyiannis DS, Dascalogiannaki M, Gourtsoyiannis NC (1997) Uncommon locations of hydatid disease: CT appearances. *Eur Radiol* 7(8):1303–1308
5. Alaoui-Yazidi A, Sibai H, Kafih A, el Meziane A, Bartal M (1995) Pancoast—tobias syndrome of hydatid etiology. *Rev Mal Respir* 12 (1):49–52