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CASE REPORT

Mesenchymal hamartoma of the chest wall in a 10-year-old girl mimicking malignancy: a case report

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Abstract

We herein report a rare case of mesenchymal hamartoma of the chest wall in a 10-year-old girl. She complained of chest pain and was diagnosed with a large chest wall tumor originating from the left fourth rib. Malignancy such as osteosarcoma or chondrosarcoma could not be ruled out with imaging studies. Therefore, we performed a core needle biopsy assisted by thoracoscopy, which revealed no malignancy. Therefore, extended resection with chest wall reconstruction was unnecessary, and thoracoscopy-assisted tumor excision with only the removal of the involved fourth rib was performed without chest wall reconstruction. The postoperative course was satisfactory with no thoracic deformity and no recurrence.

Keywords Mesenchymal hamartoma of the chest wall · Chest wall tumor · Thoracoscopic surgery

Introduction

Chest wall tumors are rare, with an incidence of less than 2% of the population, but approximately 50–80% of such tumors are malignant. In children, chest wall tumors become much rarer, accounting for only 1.8% of all solid tumors. Among them, mesenchymal hamartoma of the chest wall (MHCW) is extremely uncommon and is reported to account for 0.03% of primary bone tumors [1, 2]. MHCW is a chondro-osseous benign tumor in ribs that is usually diagnosed at birth or shortly afterward, which was first reported under this name in 1979 [3, 4]. Although it has a benign nature, it may be diagnosed as a malignant tumor like chondrosarcoma, Askin tumor, or osteosarcoma because of its aggressive appearance [5, 6]. In some cases, this aggressive appearance leads to unnecessary extended surgery with chest wall reconstruction, which may cause several complications, including deformity of the trunk and scoliosis [3].

In this paper, we experienced MHCW in a girl who was 10 years of age, which is far older than a peak age of diagnosis of this rare disease. Moreover, the imaging findings of her tumor mimicked malignant characteristics, and thoracoscopy-assisted core needle biopsy was useful to determine surgical strategy.

Case report

A 10-year-old girl presented with anterior chest pain and visited the clinic. She was referred to our hospital because X-ray at the previous hospital revealed a mass with calcification in the left upper thoracic area (Fig. 1). She was otherwise healthy and had no remarkable family history or personal medical history. Neither had she undergone an X-ray examination before this event.

When she first came to our hospital, the chest pain was spontaneously relieved. She had no palpable mass and no chest deformity. Contrast-enhanced chest (CT) showed a cauliflower-like tumor of 7 cm in diameter with sporadic calcification arising from left 4th rib associated with osteolysis (Fig. 2a). CT also revealed pleural effusion (Fig. 2b), and the 5th and 6th ribs were with bone deformities (Fig. 2c). Magnetic resonance imaging (MRI) showed a mass with

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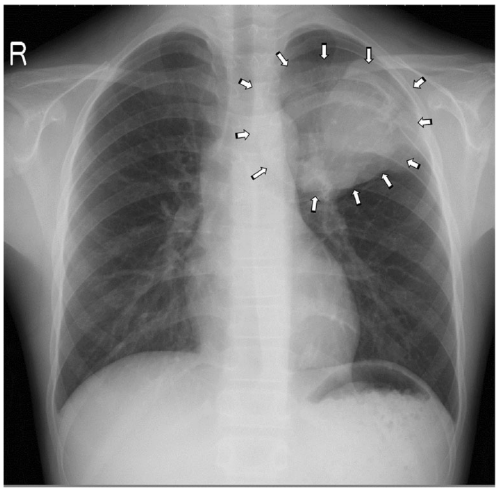

Fig. 1 Chest X-ray at admission showed a mass with calcification in the left upper thoracic area

internal heterogeneity on both T1- and T2-weighted images, including cystic lesions with fluid-fluid level (Fig. 3a). The tumor was enhanced reticularly with gadolinium (Fig. 3b). The differential diagnoses based on imaging tests included osteochondroma with or without chondrosarcoma. However, we lacked enough information to deny malignancy. If the tumor was chondrosarcoma, extended surgery with wide margin was necessary, as chondrosarcoma is refractory to chemotherapy [7].

However, as the patient was about to enter puberty, the aesthetic outcome was very important. Therefore, we planned a thoracoscopy-assisted biopsy, with the biopsy itself performed from the chest wall side, since a biopsy from the thoracic cavity side might cause pleural dissemination if the lesion turned out to be malignant. The patient was placed in the left lateral decubitus position under single-lung ventilation. Thoracoscopy revealed a tumor protruding from the fourth rib that had partly adhered to the lung (Fig. 4a). A small

Fig. 2 Contrast-enhanced CT showed a mass 7 cm in diameter, with calcification and cyst formation. The tumor arose from the fourth rib

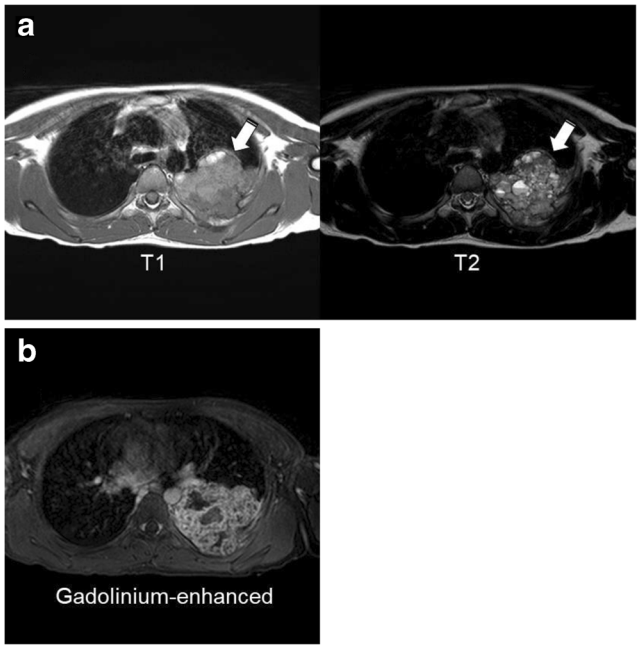

Fig. 3 MRI revealed a heterogeneous mass including cystic lesions with fluid-fluid level. It was low to iso in T1-weighted image and iso to high in T2-weighted image (**a**). The tumor was reticular enhanced with 0.2 ml/kg gadopentetate dimeglumine (Magnevist®) (**b**)

posterolateral incision was made, and we obtained two samples with an 8G bone marrow needle. The needle ruptured the tumor capsule and caused intrathoracic bleeding, which was stanched by a thoracoscopic procedure.

The pathologic examination revealed cartilaginous nodules with granulation tissue (Fig. 4b, c). The differential diagnosis now included MHCW and osteochondroma. A cytological examination of the pleural effusion revealed class 3. Since malignancy was denied, we planned a less-invasive approach for radical tumor resection.

We utilized thoracoscopy this time also to make the surgical margin as small as possible. The patient was again placed in the left lateral decubitus position under single-lung ventilation. Operative scar at biopsy was extended. The fourth rib was cut at both sides of the tumor (Fig. 5a). The third and fifth ribs were artificially fractured to obtain an adequate operative field. After verifying the edge of the tumor by thoracoscopy, the pleura was incised just outside the tumor. The adhesion with the lung was separated via a thoracoscopic procedure. The tumor was extricated and removed after being packed into an EndoCatch™II. Some bits of the tumor were scattered into the thoracic cavity, but they were all collected via thoracoscopy. There was no need for reconstruction of the chest wall because the defect was under the left scapula. The operative time was 9 h 28 min, and the intraoperative bleeding volume was 622 ml.

Macroscopically, the tumor was 6 cm in diameter and arose from the fourth rib. The cartilage, bone, and cysts containing blood were observed in a cross section. Microscopically, the

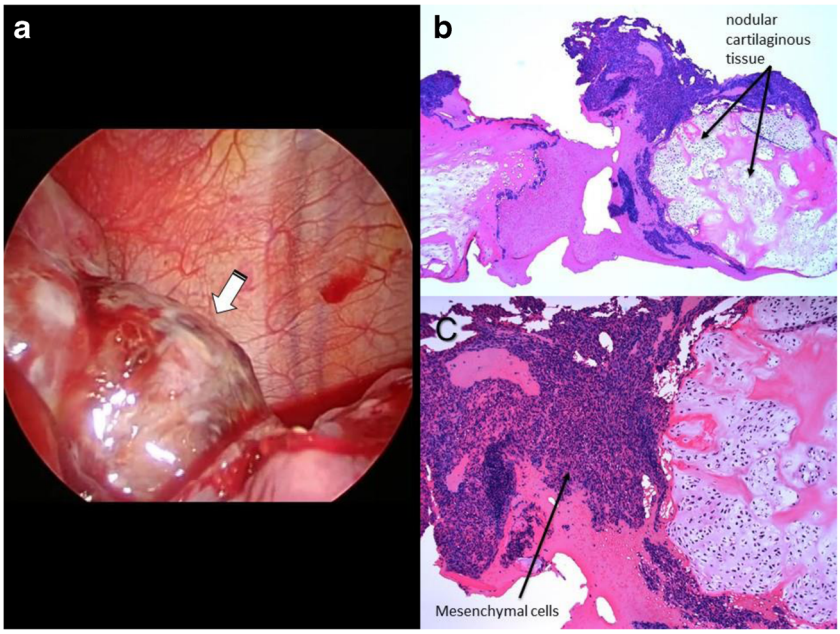

Fig. 4 Thoracoscopy showed a protruded tumor at the biopsy. The tumor had partly adhered to the lung and pleura (**a**). H&E staining of the biopsied samples showed nodular cartilaginous tissue and well-differentiated bone trabeculae. The stroma consisted of oval mesenchymal cells, and aneurysmal bone cysts were observed. No malignancy was noted (**b**, ×40, **c**, ×100)

tumor has islands of cartilage, ossification, and aneurysmal bone cyst. Spindle cells were growing in the background (Fig. 5b, c). The tumor was ultimately diagnosed as a MHCW. The postoperative course was satisfactory. The patient underwent rehabilitation during hospitalization and was discharged 13 days after surgery. No tumor recurrence and no scoliosis were noted (Fig. 6), and she had full upper limb range of motion at 2 years after surgery.

Discussion

MHCW is a benign tumor, and physicians should endeavor to make the correct diagnosis preoperatively and avoid overtreatment. Although MHCW is a rare tumor, its diagnosis is not typically difficult in neonate cases thanks to its specific imaging findings. X-ray usually shows an expansive mass involving one or more ribs with calcification and destruction

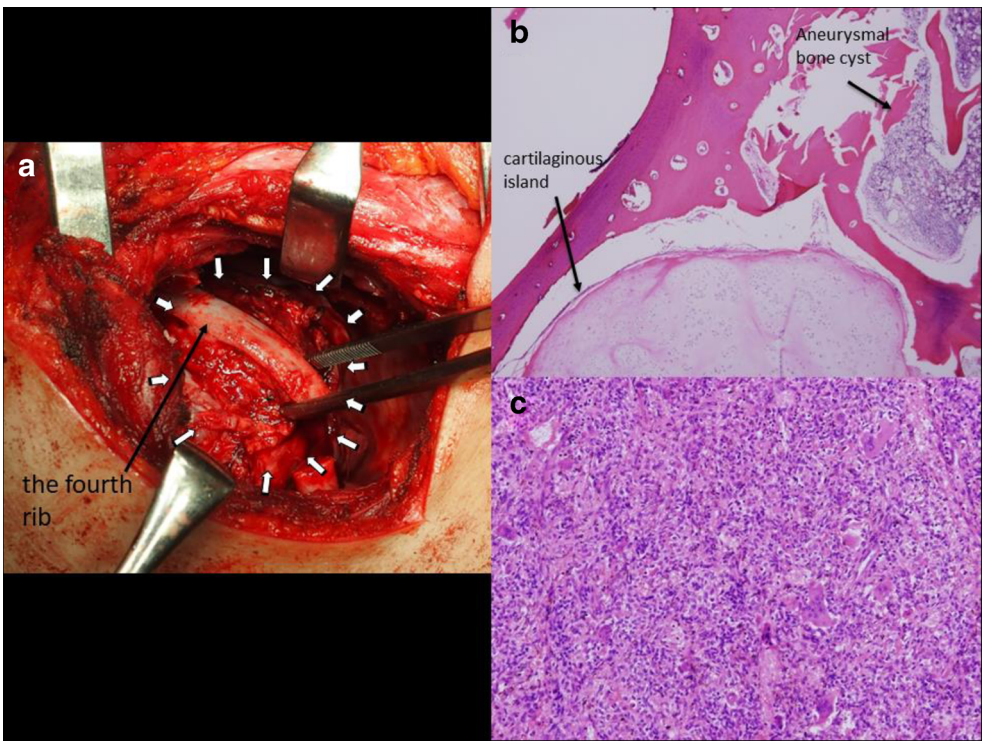

Fig. 5 At radical surgery, the tumor (*white arrows*) arose from the fourth rib. The intrapleural adhesion was dissected by a thoracoscopic procedure. The third and fifth ribs were artificially fractured to obtain an adequate operative field, and the fourth rib was removed with the tumor (**a**). Microscopically, the tumor has islands of cartilage, ossification, and secondary aneurysmal bone cysts. Spindle cells were found to be growing in the background with H&E staining (**b**, ×40, **c**, ×100)

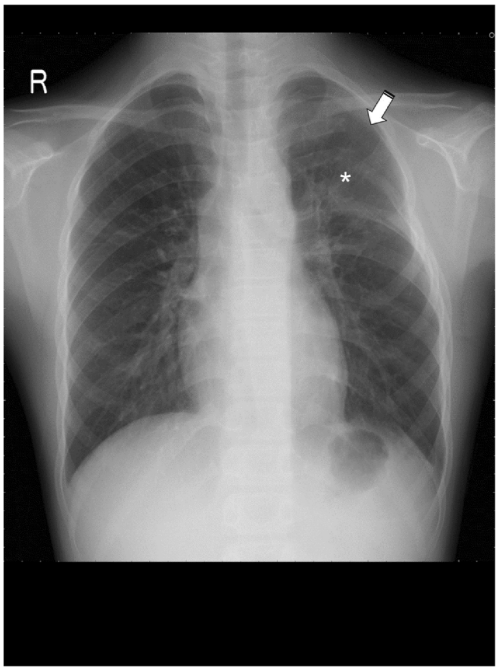

Fig. 6 Postoperative chest X-ray showed no recurrence and no scoliosis or chest wall deformity (2 years after surgery)

of the ribs [8, 9], and CT shows (1) rib origin, (2) osseous expansion, (3) intratumoral calcification (seen in nearly 100% of lesions) and (4) hemorrhagic cyst with fluid-fluid level (secondary aneurysmal bone cysts). MRI shows almost the same features as CT but is superior in its demonstration of aneurysmal bone cysts [10]. Heterogenous mass with an echogenic capsule and calcification by prenatal ultrasonography is also reported to be a key feature [11]. Additional imaging examinations reported for MHCW include PET-CT and bone scintigraphy, both of which show an uptake in the tumor [1, 10, 12].

However, an accurate pre-operative diagnosis is much more difficult in the older population, even with the typical image findings. Indeed, no patients were diagnosed correctly in later childhood or in adults despite their imaging examinations clearly showing MHCW-like features, such as calcification in the lesions or cysts with fluid-fluid level [3, 5, 13, 14]. In these cases, patients suffered from symptoms like chest wall deformity, chest pain, and Horner's syndrome. This difficulty in obtaining a pre-operative diagnosis led to a biopsy in two of five cases to obtain more information for a diagnosis. A CT-guided needle biopsy in a 39-year-old woman, which failed to obtain enough sample for a diagnosis, suggested the possibility of chondrosarcoma and resulted in unnecessary extended surgery with chest wall reconstruction [1]. In contrast, a thoracoscopy-assisted core needle biopsy in our case successfully denied malignancy by obtaining a greater amount of sample than could have been achieved with a fine-needle biopsy. Owing to this biopsy information, we were able to avoid extended surgery and resected the tumor with a small safety margin, affecting only one rib. In addition, we were able to control bleeding from the biopsied site with thoracoscopy. This is a great merit of thoracoscopy-assisted biopsies, as a percutaneous needle biopsy tends to cause massive bleeding in MHCW [15].

Regarding the treatment strategy, surgical resection has been the mainstay treatment. Recently, however, close observation has been recommended in asymptomatic neonate cases, as spontaneous regression has been noted [16], with MHCW reportedly decreasing in size after the first 2 years of life. In addition, Jozaghi et al. suggested that the relative tumor size peaks at birth and starts to involute thereafter [3]. However, in older cases (although there were only two adult cases with information about the tumor volume change with the time course), the tumor volume increased after the initial recognition of the tumor in both cases [5, 14]. This suggests that spontaneous regression should not be expected in older patients. Therefore, when dealing with MHCW in older cases, if symptoms are noted, surgical resection is recommended, as observation alone may not improve the symptoms. At surgical resection, tumors must be completely resected, as recurrence has been reported with incomplete resection [10]. Furthermore, the surgery should be as minimal as possible, as this is intrinsically a benign tumor. Scoliosis is a common complication after unnecessarily invasive surgery and might adversely affect the patient's quality of life [16].

After resection, the specimen must always undergo a pathological analysis, as a definitive diagnosis for MHCW can be made only by a pathologic examination [14]. The pathologic features include a macroscopically grey-to-white solid tumor with cartilage, calcification, and cystic lesions filled with blood; microscopically, solid areas consisting primarily of mature hyaline cartilage; the proliferation of immature fibroblastic cells (without atypia, which differentiates MHCW from malignant lesions) and hemorrhagic cysts with osteoclast-like giant cells in the cyst wall with ossification (secondary aneurysmal bone cyst) [17].

In conclusion, it is desirable to consider potential malignancy when treating primary chest wall tumors, since the majority of them are malignant [1]. If sufficient information for a diagnosis cannot be obtained from imaging studies, we should perform a biopsy to deny malignancy before conducting radical surgery to avoid unnecessary extended surgery. However, because a biopsy can cause bleeding from the biopsied site with some blood-rich tumors, such as MHCW, a thoracoscopy-assisted biopsy is a good option, especially when the tumors are well enhanced on imaging studies.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

References

1. Yamamoto H, Soh J, Ichimura K, Konishi Y, Toyooka S, Nojima T, et al. Adult mesenchymal hamartoma of the chest wall: report of a case. Ann Thorac Cardiovasc Surg. 2014;20(Suppl):663–5.
2. Shamberger RC, Grier HE, Weinstein HJ, Perez-Atayde AR, Tarbell NJ. Chest wall tumors in infancy and childhood. Cancer. 1989;63(4):774–85.
3. Jozaghi Y, Emil S, Albuquerque P, Klam S, Blumenkrantz M. Prenatal and postnatal features of mesenchymal hamartoma of the chest wall: case report and literature review. Pediatr Surg Int. 2013;29(7):735–40.
4. Ayala AG, Ro JY, Bolio-Solis A, Hernandez-Batres F, Eftekhari F, Edeiken J. Mesenchymal hamartoma of the chest wall in infants and children: a clinicopathological study of five patients. Skelet Radiol. 1993;22(8):569–76.
5. Altaner S, Yoruk Y, Bilgi S, Puyan FO, Doganay L, Kutlu K. Multifocal mesenchymal hamartoma of the chest wall. Respirology. 2006;11(3):334–8.
6. Sodhi KS, Aiyappan SK, Menon P, Dey P, Khandelwal N. Unilateral multifocal mesenchymal hamartoma of the chest wall: a case report and review of literature. J Pediatr Surg. 2009;44(2):464–7.
7. Marulli G, Duranti L, Cardillo G, Luzzi L, Carbone L, Gotti G, et al. Primary chest wall chondrosarcomas: results of surgical resection and analysis of prognostic factors. Eur J Cardiothorac Surg Off J Eur Assoc Cardiothorac Surg. 2014;45(6):e194–201.
8. Lisle DA, Ault DJ, Earwaker JW. Mesenchymal hamartoma of the chest wall in infants: report of three cases and literature review. Australas Radiol. 2003;47(1):78–82.
9. Anton CG, Towbin JT, Koch BL, Rubio EI, Podberesky DJ, Manaster BJ, et al. EXPERTddx: Pediatrics. 2009;2-85.
10. Groom KR, Murphey MD, Howard LM, Lonergan GJ, Rosado-De-Christenson ML, Torop AH. Mesenchymal hamartoma of the chest wall: radiologic manifestations with emphasis on cross-sectional imaging and histopathologic comparison. Radiology. 2002;222(1):205–11.
11. Wie J, Kim JY, Kwon JY, Ko HS, Shin JC, Park IY. Mesenchymal hamartoma of the chest wall: prenatal sonographic manifestations. J Obstet Gynaecol Res. 2013;39(6):1217–21.
12. Okamoto K, Tani Y, Yamaguchi T, Ogino K, Tsuchioka T, Nakajima M, et al. Asymptomatic mesenchymal hamartoma of the chest wall in child with fluorodeoxyglucose uptake on PET/CT-report of a case. Int Surg. 2015;100(5):915–9.
13. Conlan AA, Hurwitz SS, Gritzman MC. Giant chondromatous hamartoma of the first rib. A case report. S Afr Med J Suid-Afrikaanse tydskrif vir geneeskunde. 1982;62(19):703–4.
14. Ahmadinejad M, Pour AA, Hosseini PK, Hashemian AM, Ahmadi K. A rare case of hamartoma chest wall following trauma in a 42-year-old man. Med Arch. 2016;70(5):398–400.
15. Tsuji Y, Maeda K, Tazuke Y, Ono S, Yanagisawa S. Mesenchymal hamartoma of the bilateral chest wall in neonates. Pediatr Surg Int. 2012;28(9):939–42.
16. Freeburn AM, McAloon J. Infantile chest hamartoma—case outcome aged 11. Arch Dis Child. 2001;85(3):244–5.
17. Fletcher CDM, Unni KK, Mertens F. WHO classification of tumours of soft tissue and bone. 2002;348.