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Case Report

Asymptomatic Mesenchymal Hamartoma of the Chest Wall in Child With Fluorodeoxyglucose Uptake on PET/CT—Report of a Case

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We had experience with a case of mesenchymal hamartoma of the chest wall (MHCW) with fluorodeoxyglucose (FDG) uptake on positron emission tomography/computed tomography (PET/CT). We reported the first case of asymptomatic MHCW in a child with preoperative PET/CT. Mesenchymal hamartoma of the chest wall is a rare benign tumor that usually presents as a visible chest wall mass or respiratory problems secondary to compression of the lung in early infancy. It is often reported that malignant transformation is extraordinarily rare. Positron emission tomography/CT is useful for diagnosis of malignancy. There is no report of MHCW in a child with preoperative PET/CT before. We examined an asymptomatic 1-year-old girl with an incidental finding on a chest x-ray. Scans of CT and PET/CT were performed before surgical resection. After surgery, the resected tumor was examined histologically. Chest x-ray and CT scan of the chest confirmed a 25- × 20-mm round shaped intrapleural mass containing calcification and destructing the rib, arising from the third rib. Scan of PET/CT demonstrated the mass with light FDG accumulation. Histologically, the mass was homogenous, with thick funicular of hyaline cartilage interdigitating with scattered fiber. There were no malignant cells. No malignant MHCW was demonstrated in the

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mass, with light FDG accumulation by PET/CT. PET/CT might be a useful tool to distinguish malignant MHCW in children.

Key words: Mesenchymal hamartoma – Child – PET/CT

Mesenchymal hamartoma of the chest wall (MHCW) is a rare lesion of uncertain pathogenesis that arises from one or more ribs, nearly always in neonates or in early infancy.[1–4] The common presentation is in the form of a visible chest wall mass with or without respiratory distress.[2–5] Mesenchymal hamartoma has usually a benign course and may rarely have severe, and sometimes fatal, respiratory compromise.[1–12] However, some have reported that malignant transformation is extraordinarily rare.[13,14] We herein report a case of benign MHCW with fluorodeoxyglucose (FDG) uptake on preoperative positron emission tomography/computed tomography (PET/CT), with a literature review of this rare entity.

Case Report

An asymptomatic 1-year-old girl was admitted to our hospital with an incidental finding on a chest radiograph. A radiograph was performed when she got influenza. Physical examination did not reveal any abnormality. Cardiovascular and respiratory system examination was normal. Ultrasound scan confirmed the presence of a solid lesion. Skeletal survey showed no other abnormalities and blood parameters were normal.

Chest x-ray demonstrated a 25- × 20-mm round-shaped lesion derived at a left rib (Fig. 1A). Computed tomography scan of the chest confirmed a 25- × 20-mm round-shaped intrapleural mass containing calcification and destructing the rib, arising from the third rib; but there was no evidence of invasion (Fig. 1B, 1C).

Preoperative PET/CT was performed, because we considered that the mass may be malignant. Scans of PET/CT demonstrated a mass with light FDG accumulation. There was no FDG accumulation in the lymph node (Fig. 1D). We suspected the mass was borderline malignant because FDG accumulation was light. Surgical resection was performed.

At surgery, the mass was found to be intrapleural, arising from the left third rib. The underlying lung appeared grossly normal and uninvolved. The mass was firm, calcified, and brittle, and could not be removed intact (Fig. 2A). The mass was excised together with a part of the third and fourth ribs, because of potential for malignancy.

Subsequently the patient's postoperative course was unremarkable. She was discharged home 3 weeks after surgery. After 5 years of follow-up, she has no evidence of recurrent disease.

Pathological Findings

The resection specimen was 25 × 17 × 20 mm in size. It was encapsulated by pleura with smooth surface (Fig. 2B). On bisection, the mass was solid and white uniformly with areas of hemorrhage. The mass was on the ribs, but ribs were intact without destruction and invasion.

Histologically, the mass consist of mature hyaline cartilage and loose fibrous tissue. There are immature fibroblast-like cells (Fig. 2C). In the ribs, there was unnatural mature hyaline cartilage. It was considered related to the mass (Fig. 2D).

Discussion

Mesenchymal hamartoma of the chest wall (MHCW) in infants is a rare abnormality that is usually a benign condition. Other names have included benign mesenchymoma, infantile osteochondroma, osteochondrosarcoma, and infantile cartilaginous hamartoma.[2–6] The currently accepted name of mesenchymal or chest wall hamartoma, as initially proposed by McLeod and Dahlin in 1979,[1] is now considered the most appropriate, as it best reflects the benign nature and multiple histologic components of this lesion.[1] Its incidence is estimated to be 1 in 3000 among primary bone tumors, and <1 in million in the general population.[16,17] Mesenchymal hamartomas are not considered true neoplasms and are composed of maturing, proliferating normal skeletal elements, with no propensity for invasion or metastasis.[5] Approximately 70 cases have been described to date.[6,7]

The incidence is the ratio with a male predominance of 2:1 to 4:1. Mesenchymal hamartoma of the chest wall accounts for 7 to 14% of all solitary lung nodules.[13] The most common modes of clinical presentation of MHCW are as a chest wall mass in

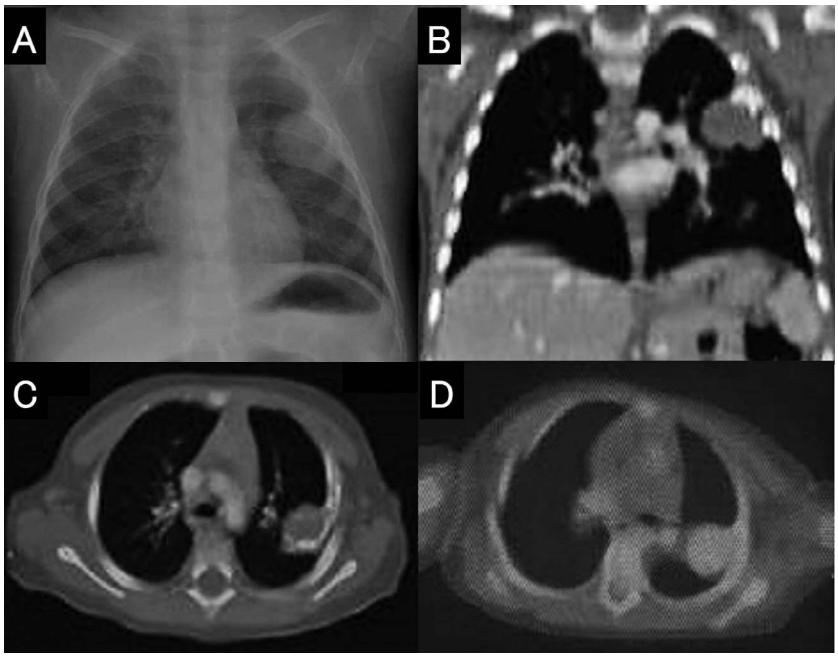

Fig. 1 (A) Chest x-ray demonstrated a round-shaped mass derived at a left rib. (B) Coronal reformatted image also demonstrated a round-shaped mass derived at a left rib. (C) CT scan of the chest showed an intrapleural mass destructing the rib. Within it, there was calcification. (D) PET/CT showed the mass with light FDG accumulation.

infancy, with or without respiratory distress. Less commonly, MHCW might be an incidental finding on a chest x-ray done for other reasons, such as respiratory tract infection.

Clinically, it usually presents as a unique lesion involving more frequently the right hemithorax, although bilateral and multiple ipsilateral cases have been described previously.[4,6–8] Typically, these lesions arise from the central portions of one or several ribs, and can range in size from several centimeters to very large expansile lesions involving most of the thoracic cage. Radiological findings on chest x-ray, CT, and MRI are usually quite typical.[7] Cross-sectional imaging (CT and MRI) clearly depicted the rib origin, osseous expansion, and associated extrapleural soft-tissue masses. The intrinsic characteristics corresponded with the pathologic composition of the lesion. Computed

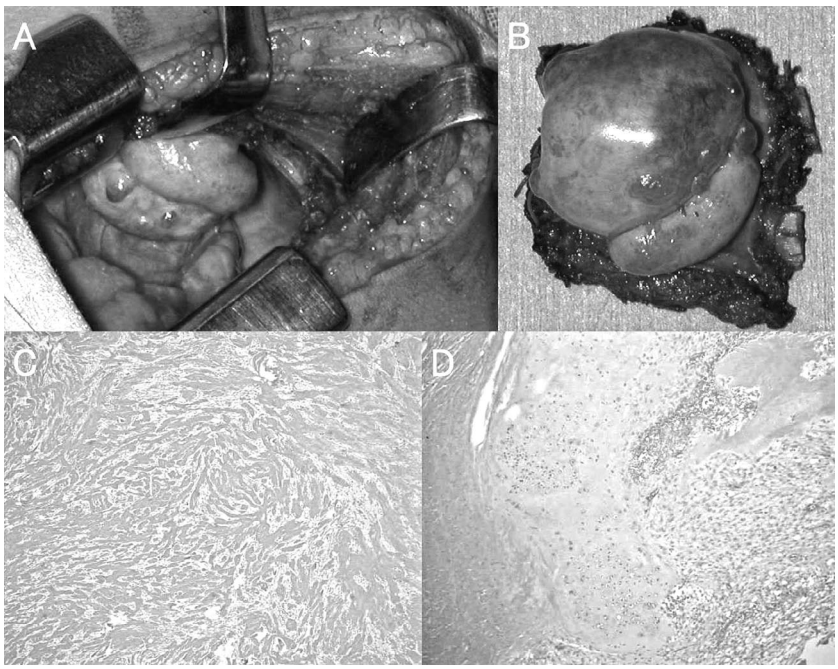

Fig. 2 (A) Operative findings: the mass was found to be intrapleural, arising from the left third rib. It was firm, calcific, and brittle. (B) Gross surgical specimen showed the resected tumor. (C) The mass was homogenous, with thick funicular of hyaline cartilage interdigitating with scattered fiber. (D) In ribs, there was unnatural mature hyaline cartilage; it was suggested in relation to the mass.

tomography best demonstrated the matrix mineralization, seen in 100% of lesions, with the mineralization appearing chondroid alone in 79%.[7] Hemorrhagic cystic regions (secondary aneurismal bone cyst areas) also were seen with CT (fluid levels in 64% of lesions), although T2-weighted MR images were superior in demonstrating this feature in 80% of cases.[7]

Histologically, the lesion is composed of cartilage, smooth muscle, and respiratory epithelium forming a disorderly mass. The stroma consists of oval or spindle mesenchymal cells with no atypia or abnormal mitotic activity. Osteoclast-like giant cells in the vicinity of blood-filled cysts resemble aneurysmal bone cysts.

In this case, chest x-ray, CT, and MRI all are useful in diagnosis, with CT being particularly useful because of the calcification and bony origin.[4] Mesenchymal hamartoma of the chest wall is usually considered a benign expansive lesion with a favorable outcome. However, for approximately 70 cases of mesenchymal hamartoma reported, to our knowledge there are 4 reports of malignant transformation.[9–12] And it may be considered radiographically suspicious of malignancy due to the presence of rib destruction and rapid growth. We suspected malignant MHCW of our case, so PET/CT was done. A percutaneous needle biopsy tends to cause massive bleeding, thus making it difficult to achieve a definitive diagnosis.[18,19] Considering the possibility of malignancy at the time of the operation, we believed the partial resection of the ribs was acceptable.

It is known that PET/CT imaging can facilitate diagnosis in adults with malignant diseases by enabling the differentiation between benign and malignant tumors. Positron electron tomography/CT may be useful for pediatric neoplastic lesions, the nature of which is often difficult to determine preoperatively.

Furthermore, glucose transporter-1 protein (Glut-1) is the major glucose transporter protein in tumor cells. Fluorodexyglucose is now widely used to evaluate regional glucose metabolism in a variety of tumors, and high correlations were found between FDG uptake and Glut-1 expression in most malignant tumors.[20–22] Uptake of FDG includes many complicated factors, and it would be difficult to make a differential diagnosis in mesenchymal tumors using FDG uptake alone. Zhao *et al*[23] reported that the benign tumor patients who had high FDG uptake had negative scores in Glut-1. Even in our case, Glut-1 might be useful for preoperative diagnosis.[23]

In summary, MHCW is a non-neoplastic benign lesion in the majority of cases. However, PET/CT might be a useful tool to distinguish malignant MHCW in children. Its early and complete excision is the adequate therapy to avoid lethal respiratory complications and more aggressive treatments, although there is recommendation for conservative management in asymptomatic cases.

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