Multifocal mesenchymal hamartoma of the chest wall in a newborn

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Mesenchymal hamartoma of the chest wall is an extremely rare, benign chondro-osseous chest wall tumor which originates from one or more ribs, in newborns or infants. It usually appears as a solitary lesion but more rarely can be multifocal and bilateral. Sometimes it may mimic chest wall malignant tumors because of its destructive radiological nature. Herein, we aim to present the imaging characteristics of a 4-day-old boy with multifocal mesenchymal hamartoma of the chest wall.

Key words: mesenchymal hamartoma, chest wall tumor, neonate, computed tomography, magnetic resonance imaging.

Mesenchymal hamartoma of the chest wall (MHCW) is quite a rare, benign chondroosseous tumor of the bone which originates from ribs. The tumor typically appears like a solitary chest wall mass with or without respiratory symptoms in newborns or infants. However, even more rarely multifocal and bilateral lesions have been shown in some Imaging findings may resemble malignant lesions. It is usually seen as a heterogeneous, solid to partially cystic, erosive-destructive and expansile mass arising from chest wall bones.1-4 Herein, we present the X-ray, computed tomography (CT) and magnetic resonance imaging (MRI) features of a newborn with multifocal mesenchymal hamartoma of the chest wall which became larger during 10 months.

Case Report

A 4-day-old boy was presented with a mass on his back without respiratory distress. The baby was born via spontaneous vaginal delivery following an unremarkable prenatal course to a 20-year-old gravida 2 mother.

Birth weight was 2.3 kg. Fetal ultrasound examination performed at 19th week was unremarkable. Blood parameters were normal. Chest radiograph was performed due to the swelling on the back of infant. Plain chest radiograph revealed multiple rib abnormalities as dystrophic, destructive and expansile changes at several levels in posterior ribs (Fig. 1). Computed tomography (CT) of the chest was performed and showed a complex, intraosseous, expansile and extensively calcific solid mass, measuring 40x27x25 mm, arising from the posterior-lateral aspect of the left chest, involving posterior elements of 4-5th ribs (Fig. 2). Other lesions of similar nature but of smaller sizes were also observed in 7th posterior rib and in the inferior of the sternum at the 4-5th costochondral junction in the anterior chest wall. The lesion which was in inferior location to the sternum showed chondroid mineralization. In addition, another paravertebral ossified measuring 35x17x13 mm, was noticed which also destructed the transverse processes and laminae of the vertebrae between the 9-10th posterior ribs in the left chest wall. The masses

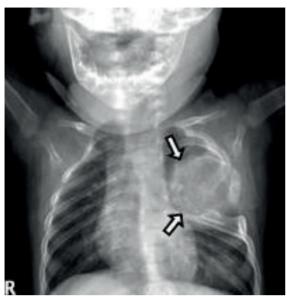


Fig. 1. Plain chest radiograph shows large heterogeneous mass (arrows) with multiple rib destruction, thinning, distortion and calcifications in the left hemitorax.



Fig. 2. Contrast enhanced axial computerized tomography (CT) scan reveals left-sided calcified, lytic, heterogeneous mass which origins from left 4-5th posterior ribs and causes expansion of the posterior parts of vertebrae (arrows).

extended through the posterior chest wall with associated thickening of the posterior ribs and extension into subcutaneous tissue. There was no evidence of metastatic disease in this study. Cardiac, head and abdominal ultrasound studies did not show significant abnormalities. Mesenchymal hamartoma and osteochondromatosis were considered initially in the differential diagnosis based

on these radiological findings. MRI was performed after 3 months. All of these lytic, destructive-expansile, heterogeneous solid lesions which were previously described in the CT scan, showed significant increase in size. The lesions were observed extending to the neural foramen and spinal canal which were at 4-5th and 9-10th posterior costa. In addition, a new lesion with a similar nature, measuring 13x10 mm, was observed in the right chest wall at the 8th posterior rib. MR images showed predominantly intermediate and heterogeneous signal intensity on T1weighted images demonstrating areas of focal high signal intensity, reflecting hemorrhagic regions (Fig. 3a). On T2-weighted images, all lesions were heterogeneous, with two lesions having predominantly intermediate signal intensity, whereas the other four lesions had primarily high signal intensity. Fluid levels consistent with hemorrhagic cavities were seen in lesions on T2-weighted images (Fig. 3b). After intravenous administration of contrast agent, all four lesions demonstrated mild to moderate diffuse enhancement of the solid components and lack of enhancement of hemorrhagic and cystic areas (Fig. 3c). The baby was evaluated in our institutional tumor board. As a result, biopsy was postponed due to the baby's age. Thus, close follow-up of the patient was decided. The baby didn't receive any treatment. In the follow-up period the masses showed progressive dimensional changes. It was decided to take a biopsy in the second tumor board. An open biopsy was performed on the month of life 5, revealing an admixture of benign bone, cartilage and mesenchymal tissues. No malignant cellular features were detected in the excised paravertebral lesion. He was discharged home 2 weeks after surgery. Thorax CT scan at 3 months postoperatively showed two of the lesions were stable but the other ones increased in size. He will undergo an operation after follow-up until the suitable age for surgery.

A written informed consent was obtained from the parents of the patient for publication.

Discussion

Mesenchymal hamartoma of the chest wall is a very rare benign tumor in childhood. It is not a true neoplasm, at times it have been

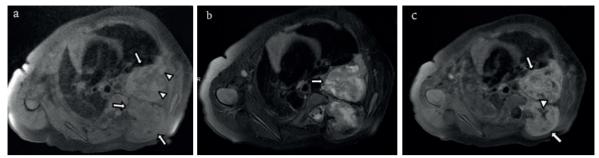


Fig. 3. Magnetic resonance (MR) imaging of the thorax and mediastinum. **(a)** Axial fat-saturated T1-weighted image (TR/TE, 520/10 ms) shows iso-hypointensity consisted with solid components and hyperintensity in the cystic parts of chest wall mass (arrows) that considering hemorrhage (arrowheads). **(b)** Axial fat-saturated T2-weighted (TR/TE, 4780/65 ms) image shows fluid-fluid levels with different signal intensities (arrow). **(c)** Axial post-contrast fat-saturated T1-weighted (TR/TE, 520/9 ms) image reveals diffuse enhancement of the solid components (arrows) and lack of enhancement of hemorrhagic and cystic areas (arrowhead).

classified as a non-neoplastic development anomaly. The term "mesenchymal hamartoma" was used primarily by McLeod and Dahlin⁵ in their 1979 report based on nine cases that they believed non-neoplastic, before that, different names, such as mesenchymoma, infantile cartilaginous hamartoma infantile osteochondroma were used. Most MHCWs are found in neonates and infants, with male dominancy that it has M:F ratio of 2:1 to 4:1. A chest wall mass or deformity with or without respiratory distress are common presentations. Although it is generally defined as a single large mass, some studies reported that it may be seen extremely rarely bilateral and multifocal as in our case.1,2

Radiological features on radiograph, CT, and MRI are absolutely characteristic. Chest radiograph may show an expansile mass involving one or more ribs with cortical irregularity, destruction and osteoid or chondroid mineralization. CT scan clearly reveals the rib origin, osseous expansion, and associated extrapleural soft-tissue mass based on its histopathological nature. CT can also show osteoid, chondroid or mixed type mineralization. In our case, we reported chondroid mineralization in one of the four lesions and osteoid mineralization in two of them. Hemorrhagic cystic regions (aneurysmal bone cyst-like fluid levels) also may be detected with CT and MRI scans, but MRI is generally an ideal technique for demonstrating this feature. MRI shows heterogeneous signal intensities of the mass on both T1- and T2weighted images.3 In our case, MRI was useful

in diagnosis, with detecting hemorrhagic cystic component. Mesenchymal hamartoma may be confused with malignant chest wall tumors such as Ewing sarcoma, rhabdomyosarcoma, osteosarcoma, chondrosarcoma or metastasis of leukemia, neuroblastoma and lymphoma, because of the presence of rib destruction and rapid growth. These tumors are extremely rare in newborns and do not contain fluid-fluid levels.6,7 Fluid-fluid levels are characteristic radiologic features of mesenchymal hamartoma on MRI, for distinguishing from other malignant tumors. Radiologists should differential diagnosis particularly infantile fibrosarcoma with congenital which is a malignant chest wall tumor in neonates. Congenital infantile fibrosarcoma commonly origins from soft tissues. However, mesenchymal hamartoma origins from ribs. Most of the lesions increase in size after the diagnosis, generally stop growing in the first year of life.8 Despite its benign nature, there have been reported many cases mentioning regrowth at the post-operative period, but only one of them presented malign transformation.9

In conclusion, mesenchymal hamartoma of the chest wall may be seen rarely in childhood. It can be misdiagnosed because of bone-destroying radiological findings. Despite its destructive, expansile and rapidly growing appearance, it has a benign histopathology. It usually presents as a solitary lesion but extremely rarely it may be multifocal and bilateral so it should be kept in mind in the differential diagnosis of multifocal chest wall tumors of newborns.

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