Atypical Presentation of Mesenchymal Hamartoma of the Chest Wall in a One-Month-Old Infant

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A one-month-old infant had an unusual presentation of Mesenchymal Hamartoma of the Chest Wall (MHCW). The patient presented with excessive crying. Chest X-ray revealed a large chest mass. The CT and MRI scans revealed heterogeneous mass originating from the posterior aspect of the fifth rib. The patient developed severe respiratory distress and required resection of the mass. The histologic study on the specimen confirmed the diagnosis of mesenchymal hamartoma of the chest wall.

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Mesenchymal hamartoma of the chest wall (MHCW) is a rare entity[1-5]. MHCW is a very rare benign extrapleural tumor that arises mainly from the central part of the ribs[1,6-9]. There are approximately 80 reported cases in the literature[1]. The tumor is composed of maturing skeletal components. Most of the cases present with a chest wall deformity[1,10-12]. It may result in respiratory symptoms, such as recurrent asthma-like symptoms or pneumonia. It may, in some cases, progress to severe respiratory failure and death[13]. Nevertheless, if the condition is asymptomatic or causing tolerable symptoms, the tumor could be managed conservatively, and usually, it regresses[1,5,9,14]. Surgical resection is required in certain cases, especially if there is a significant respiratory compromise or in the case of massive hemorrhage[13,14].

The aim of this report is to present a case of a rare tumor in a one-month-old infant who had a MHCW and required resection of the mass due to respiratory distress.

THE CASE

A one-month-old Indian infant presented with excessive crying. The infant was irritable, but his vital signs were stable and the physical examination was normal. A chest X-ray revealed a large expansile lytic lesion involving the left ribs with significant right-sided mediastinal shift, see figure 1 (A). CBC, liver function test and kidney function were within normal limit. CT chest revealed a large well-defined expansile lytic lesion measuring 5.5x5x5 cm projecting within the left hemithorax and originating most probably from the fifth rib posteriorly with a significant mediastinal shift, see figure 1 (B and C). MRI confirmed the previous findings and revealed the heterogeneity of the mass, see figure 1 (D). Within two days, the infant became very tachypneic and had labored breathing with severe intercostal retractions; as a consequence of the severe respiratory distress, thoracotomy and mass resection were performed. The mass was originating from the posterior aspect of the fifth rib which was excised during the procedure. The excised mass measured 4x4.5x2 cm and weighed 34g. On gross section, the mass had cartilage-like tissue and bone. Microscopically, the mass contained large foci of mature hyaline cartilage with foci of endochondral ossification admixed with spindled areas resembling aneurysmal bone cyst. There were blood-filled spaces of variable sizes that were linked by fibroblasts, separated by cellular fibrous septa and containing osteoclastic giant cells, reactive woven bone and calcifying fibromyxoid tissue. The features were compatible with mesenchymal hamartoma of the chest wall. The patient had a stable postoperative period and was discharged after ten days.

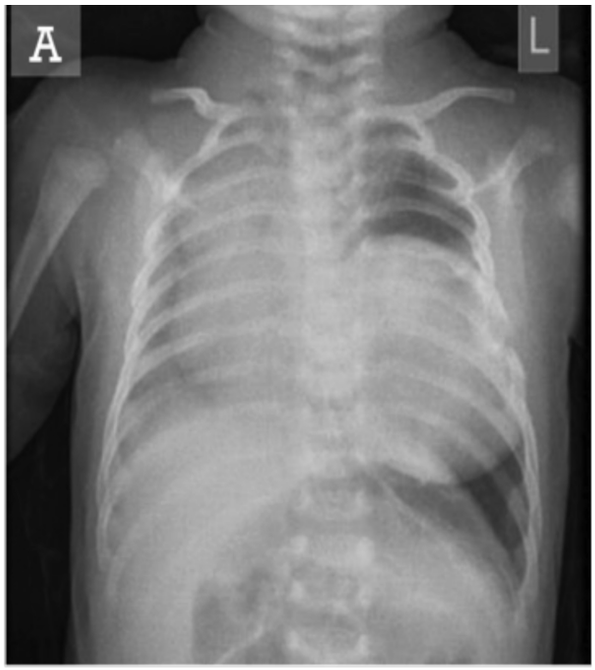

Figure 1 (A)

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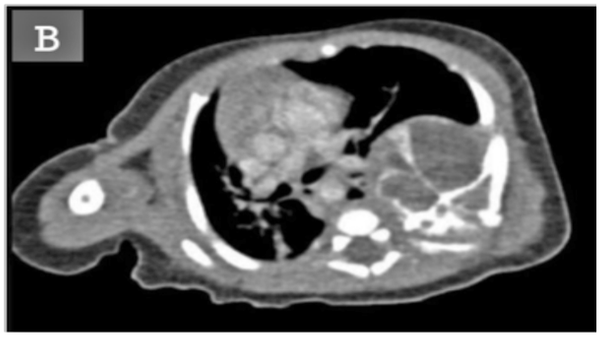

Figure 1 (B)

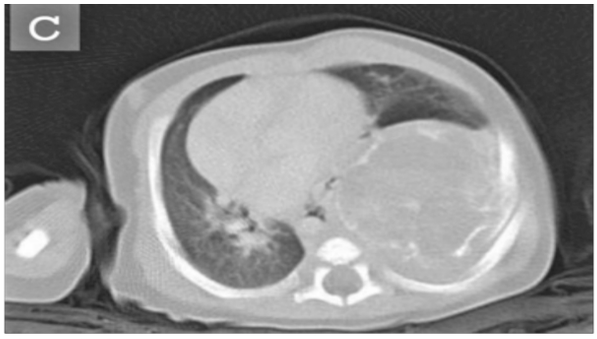

Figure 1 (C)

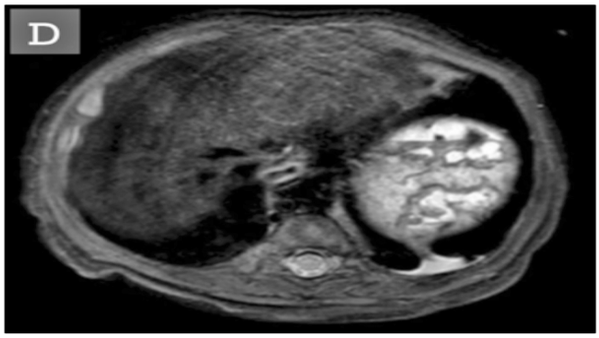

Figure 1 (D)

Figure 1 (A to D): Chest X-Ray Reveals a Large Lytic Left Sided Extra-Pleural Mass (A). CT Reveals a Large, Well-Defined Expansile Lytic Lesion. There is a Significant Mass Effect with a Mediastinal Shift of the Heart and Mediastinal Structures to the Right Side (B and C). MRI Reveals a Heterogeneous Well-Defined Chest Wall Mass Lesion Arising from the Left Fifth Rib Posteriorly (D)

DISCUSSION

Mesenchymal hamartoma of the chest wall is a very rare tumor[1,2,5-9]. The estimated incidence of the tumor is less than one case per million among the general population[1]. Different nomenclatures were used in the literature to describe this entity, such as mesenchymoma, chondromatous hamartoma, osteochondrosarcoma, benign chondroblastoma and osteochondroma[2]. McLeod et al were the first to call it mesenchymal hamartoma of the chest wall, which is currently considered the most appropriate description of the histologic features of the tumor[1,2].

MHCW presents mainly in newborns and infants[3,6,8-10]. They usually present with a palpable chest mass, but they may also present with chest deformity or respiratory distress[10-14]. The mass is composed of several components, hyaline cartilage, large blood spaces (secondary aneurysmal bone cysts) and immature mesenchymal tissue with osteoclastic giant cells and osteoid tissue[9,10].

The mass is well-circumscribed and originates from the posterior aspect of the ribs on plain radiographic film[1,10-14]. Heterogeneous signal intensities on both T1- and T2-weighted images and hemorrhagic cavities are seen within the mass on MRI images[1,3-7,10-13]. CT studies are particularly useful in revealing the rib origin of the mass[1,13,14]. MHCW is benign in nature and is considered non-neoplastic developmental anomaly[1,3,7,13,14]. There are very few reports of malignant MHCW in the literature[1,5,8,10,13]. Therefore, conservative management is considered acceptable and is recommended for asymptomatic patients[1,4,10-14]. Complete resection of the mass is curative and mandatory in some cases to prevent fatal complications[1,13,14]. Nevertheless, the resection of the mass usually results in chest deformity and scoliosis[1,3,5,7,13,14].

Our patient had an unusual presentation that, to our knowledge, is not described in the literature. The patient initially presented with excessive crying and without respiratory distress. The excessive crying and irritability could reflect pain and discomfort, and not surprisingly, within two days, he developed severe respiratory distress that warranted the surgical intervention.

CONCLUSION

Mesenchymal hamartoma of the chest wall is a very rare benign tumor in newborns and infants. It usually presents with a chest wall mass or respiratory distress. Nevertheless, our patient presented with excessive crying. We suggest that this diagnosis should be kept in mind when dealing especially with a newborn who is irritable and excessively crying. A chest X-ray should be considered if no other explanation for his irritability is found.

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Conflict of Interest: None.

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