

Urgent resection of bleeding congenital mesenchymal chest wall hamartoma in an infant

Dringliche Resektion eines blutenden mesenchymalen Thoraxwand-Hamartoms beim Säugling

Abstract

We report a case with prenatally diagnosed large cystic-solid mesenchymal chest wall hamartoma. An attempt of conservative management was made however repeated intralesional hemorrhage led to enlargement and severe anemia which required urgent resection at the age of 8 weeks. The infant had an unimpaired development over a follow-up of 4 years.

Keywords: mesenchymal hamartoma, congenital chest wall tumor, surgery, intralesional bleeding

Zusammenfassung

Wir berichten über ein Neugeborenes mit einem bereits pränatal diagnostizierten, ausgedehnten Thoraxwand-Hamartom von zystisch-solider Beschaffenheit. Aufgrund der Gutartigkeit der Läsion und der Erkenntnislage strebten wir zunächst eine watch-and-wait-Strategie an. Allerdings stellten sich im Alter von 8 Wochen eine Zustandsverschlechterung mit Anaemie sowie Größenzunahme des Tumors infolge Einblutung ein. Es erfolgte die Resektion unter Einbeziehung der betroffenen Rippen. Zur Augmentation der Thoraxwand verwendeten wir eine Dura-Patch. Der postoperative Verlauf war unspektakulär und das Mädchen entwickelte sich über eine Nachbeobachtung über 4 Jahre körperlich und geistig unbeeinträchtigt.

Schlüsselwörter: mesenchymales Hamartom, angeborener Tumor der Thoraxwand, Thoraxwand-Resektion, Tumorblutung

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Introduction

Congenital mesenchymal hamartoma of the chest wall (MH), synonymous and partly incorrectly named as congenital mesenchymoma [1], is a rare benign lesion which usually presents antenatally or during early infancy. The majority of patients reported in the literature has been treated by surgical resection. However large thoracic wall resections in the growing child subsequently result in deformations of the thoracic cage and spine. To avoid these long term sequels conservative management of patients has been suggested. However this option may be of limited value in infants with extended cystic and enlarging MH.

Case report

After an uncomplicated pregnancy antenatal routine obstetrical ultrasonography of the fetus at 39 weeks of age had revealed an extensive chest mass. Intrauterine magnetic resonance imaging (MRI) depicted the presence of a large heterogeneous mass of 7 x 5.3 cm occupying the fetus' right thorax. The female baby was the 2nd child of young healthy parents. The baby was delivered by Caesarean section with a weight of 3170 g. APGAR scores were 5/7/8 and postnatal oxygen supplementation was applied. The baby was in a stable condition, however, a protrusion of the right chest wall was obvious (Figure 1).



Figure 1: Clinical appearance of the thoracic cage. Protrusion of the lower right thoracic wall.

Chest X-ray showed a large cloudy calcificated mass extending within the right hemithorax shifting the mediastinum to the left. The lesion destructed parts of the 6th to 9th rib and displaced the right lung, vessels and the mediastinum to the contralateral side. An open biopsy was performed at the first day of life. Histopathology revealed cartilage with expression of protein S100 and a low proliferation rate (MiB-1). Cystic areas within the mass were filled with a hemorrhagic fluid of brownish color. Furthermore, proliferating spindle cells, osteoclast-like giant cells, newly generated fibrous ossifications as well as ectatic vascular spaces were found (Figure 2).

Postnatal MRI showed a multicystic mass of 6.7 x 5.8 x 5.6 cm arising from the thoracic wall and consisting of large fluid contained cavities. Histopathology results and imaging both were consistent with the diagnosis of a congenital mesenchymal chest wall hamartoma.

As the baby was in a fair condition a conservative treatment was favored. At three weeks of age the infant was sent home in a good condition.

In the course of the follow up examination at 5 weeks of age we noticed an anemia with the necessity of a transfusion (Hemoglobin 6.8 g/dl). The condition of the child worsened dramatically. The MRI disclosed an increase in size to 9.3 x 8.1 x 7 cm due to hemorrhage. This enlargement was accompanied by shifting the large vessels to the left side and compressing the inferior vena cava. Sedimentation levels within the large cystic spaces of the hamartoma indicated intralesional bleeding (Figure 3). Under these circumstances surgical resection was performed at the age of 8 weeks. An extended resection of the thoracic wall encompassing the ribs 7 through 9 were nearly completely as well as parts of the 6th and 10th rib. Chest wall reconstruction was facilitated by preservation

of the outer musculature and the use of a bovine pericardium patch.

The girl recovered quickly and is now doing well without any further respiratory problems. She showed a slight deformity of the right thoracic wall due to a bony defects of the ribs without concomitant deformation of the spine. Her neurodevelopmental outcome is unimpaired. MRI controls at 12 and 36 months of age excluded recurrence of the MH.

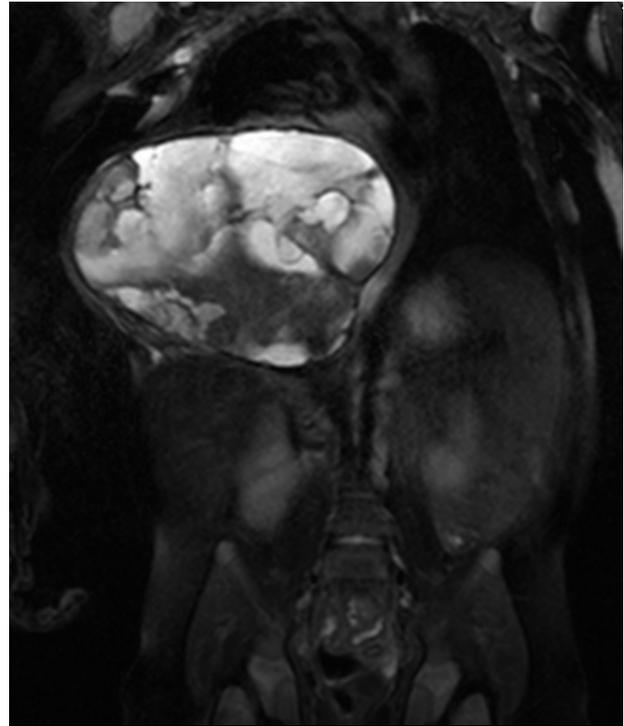


Figure 2: Histopathologic findings of the solid tumor parts: Osteoclast-like giant cells in the vicinity of cysts and ectatic vascular components

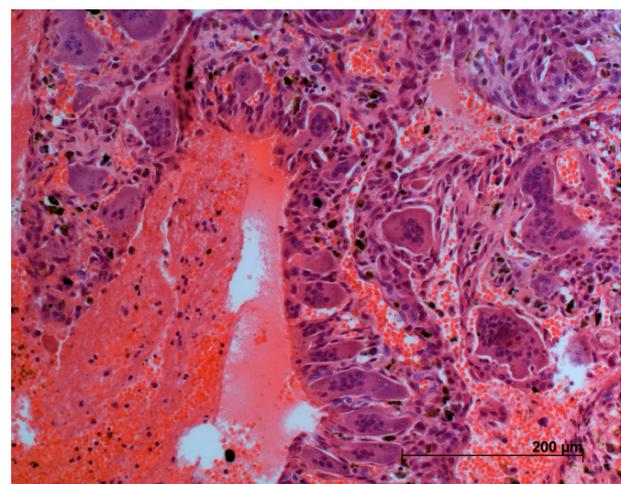


Figure 3: Large intrathoracic extension of the midline crossing tumor

Discussion

MH are rare benign tumor like lesions. In the majority of cases the mass arises from the central part of the ribs [2]. In the recent literature nearly 80 cases have been reported [3] mostly in form of case studies. Usually the mass presents in early infancy although some cases with presentation in adults were observed [4].

Diagnosis is based on imaging (chest X-ray, MRI) and histopathological findings. Typically the lesion is large and well delineated but compresses the surrounding organs and results in a deformity of the thoracic wall.

Following an initial period of rapid growth between the 28th and 36th week of gestation discontinuity of enlargement or spontaneous regression can be expected [5].

Different therapeutic approaches have been reported in the literature: The majority of the hamartomas has been excised by primary intuition or secondary due to respiratory distress, cardiovascular compression or neurological symptoms. Some reported patients underwent incomplete resection.

However a few authors described conservative management and a spontaneous regression supporting the concept to be appropriate for children with minor symptoms [6], [7]. Nevertheless the experience in conservative management is limited to intermediate follow-up. Malignant transformation or recurrence after resection have not been reported.

Lisle et al. presented three cases of MH of the chest wall. In one of the cases the MH was left in place and follow up at the age of 6 years revealed some reduction in size after conservative management [1].

Shimotake et al. [5] described a newborn with a large MH occupying the left hemithorax resulting in severe respiratory insufficiency. A critical period was survived by mechanical ventilation and long term endotracheal intubation. Subsequently the tumor showed a self-limiting growth behavior. As conclusion the authors advocate conservative management after confirmation of the diagnosis [5].

Only Cameron et al. [7] reported a case comparable to our experience. Initially this infant was managed conservatively but in the age of 5 month the tumor increased in size. Intralesional bleeding lead to increasing compression of the lung and excision of the MH.

Based on the suggestions of these case reports we initially favored a conservative management in the presented case with avoiding the risks of a destructive en-bloc resection and the risk of long term complications as impaired thoracic growth and postsurgical scoliosis.

Generally, intralesional bleeding is a characteristic finding in MH. Different signal intensities of the cyst content with fluid-fluid levels are a well known feature of MH in MRI and CT. This bleeding can lead to an enlargement of the tumor size and compression of the lung as reported above [7]. One patient underwent partial tumor resection because of profuse bleeding in the course of open biopsy [2].

In the presented infant the intralesional bleeding resulted in repeated severe anemia with worsening of the patient's condition. After stabilization we performed the resection of this lesion.

In conclusion conservative management of MH requires close follow-up examinations of the patient to recognize potential life threatening complications.

Notes

Competing interests

The authors declare that they have no competing interests.

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