

## Mesenchymal Hamartoma of the Chest Wall: A Case Report of Prenatal Detection and Review of the Literature

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### Abstract

Mesenchymal Hamartoma of the Chest Wall (MHCW) is a rare benign lesion almost exclusively found in early infancy and childhood. To date, just over 100 cases have been reported and only in 15 cases, including ours, the mass was detected prenatally, although a certain diagnosis was possible only after birth with biopsy. The optimal treatment of MHCW remains controversial. We report a case of congenital MHCW, detected in the prenatal age and followed up to three years of life. We reviewed the literature, analyzing only the reported cases with fetal detection of the lesion, with emphasis on Fetal Magnetic Resonance Imaging (MRI) diagnosis, to provide the correct prenatal and postnatal management.

### Key Points

1. An accurate prenatal diagnosis of MHCW is crucial to allow a correct prenatal and postnatal management of the lesion; moreover, it helps in prenatal counseling and avoids aggressive, unnecessary and harmful treatments, such as chemotherapy, radiation and/or debulking surgery. A prenatal diagnosis of a thoracic lesion often requires diagnostic insight with fetal MRI, which allows to identify the exact origin of the lesion, to estimate lung maturation and to plan the delivery and the best post-natal care path. The detection of a mass involving the chest wall, which tends to grow and to change its nature during the pregnancy, with evidence of aneurysmal and cystic remodeling and secondary calcification, is highly suggestive of MHCW.
2. After birth, all patients should be studied with cross-sectional imaging including a CT scan or MRI in order to obtain a differential diagnosis with the others benign and malign conditions. Although definitive diagnosis would require histological examination, there is now sufficient experience in order to obtain a diagnosis relying only on the radiological findings associated with physical examination; the problem is only to keep in mind the MHCW in the differential diagnosis of fetal or neonatal thoracic lesion.

3. Observation with close follow-up consultation is possible in cases of asymptomatic lesion, whereas a surgical resection has been considered the treatment of choice in symptomatic cases. Surgery should be conservative whilst trying to achieve margins free of lesion, if it is possible.

**Keywords:** Chest Wall Tumor; Fetal MRI; Fetal Ultrasound; Mesenchymal Hamartoma; Neonatal CT; Pediatric Surgery; Prenatal Diagnosis; Skeletal Hamartoma

### Case Report

A primigravid, 34 years-old woman with no past medical history was referred to our hospital at 35+5 weeks of gestation for evaluation of an intrathoracic mass of her only fetus. Second level Ultrasound (US) showed an intrathoracic mass in the left hemithorax suggesting the diagnosis of left diaphragmatic hernia (Figure 1). The subsequent MRI performed at 36+5 weeks of gestation evidenced an expansive lesion arising from the left chest wall, involving ribs, with intrathoracic development and right shift of mediastinum, heart and left lung parenchyma (Figure 2). The lesion showed dimensions of 48x56x47 mm and did not involve lung parenchyma nor mediastinum. The mass showed a high signal in T2weighted sequences, and lobulated and regular

margins, surrounded by T2\* hypointensity due to hemorrhage and/or calcification.



**Figure 1:** Prenatal ultrasound, performed at 35+5 weeks of gestation, shows a heterogeneous mass occupying fetal left hemithorax (white arrow), suggesting a left diaphragmatic hernia.



**Figure 2:** Fetal MRI, Single Shot Turbo Spin Echo (SSh, TSE) T2weighted imaging, performed at 36+5 weeks of gestation. The exam displays a heterogeneous lesion occupying the left hemithorax of the fetus, characterized by solid and cystic compounds with associated pleural effusion. The left lung is more hyperintense than the other one.

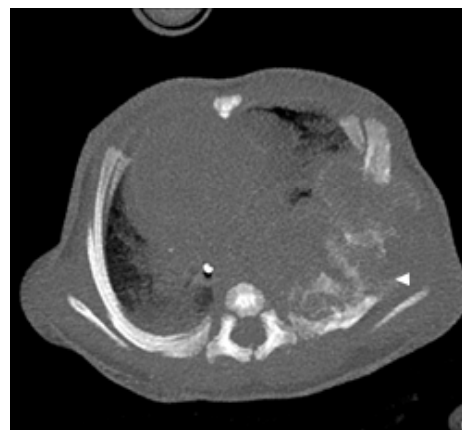
These findings excluded the diagnosis of left diaphragmatic hernia and suggested a Chest Wall Tumor (probably a MHCW). Thanks to Fetal MRI, it was possible for the mother to deliver an asymptomatic 2800g male baby, with left hemithorax deformity, through a caesarean birth at the 37<sup>th</sup> week of gestation. The caesarean birth was due to pre-existing conditions for the mother

and was not necessary for the child's condition, as also reported in the literature; it was done in a specialized center, to avoid risk of respiratory distress. After a few hours he showed respiratory distress and he was immediately transferred to the neonatal intensive care unit. The first radiogram one day after birth (Figure 3) showed a deformity of the left hemithorax ribs and an opacity occupying the left hemithorax.

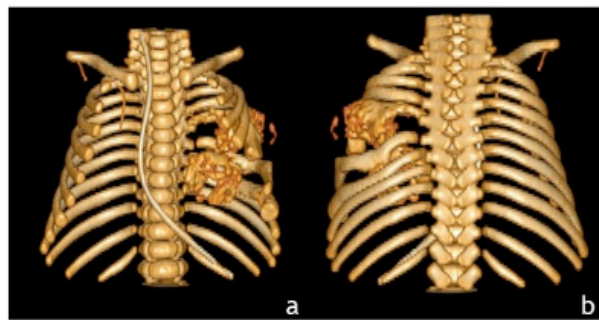


**Figure 3:** Chest X-ray, performed at birth, displays a heterogeneous mass of the left hemithorax, associated with malformations of the 4<sup>th</sup>, the 6<sup>th</sup> and the 7<sup>th</sup> ribs and the disruption of the 5<sup>th</sup> rib. It shows a rightward tracheal deviation.

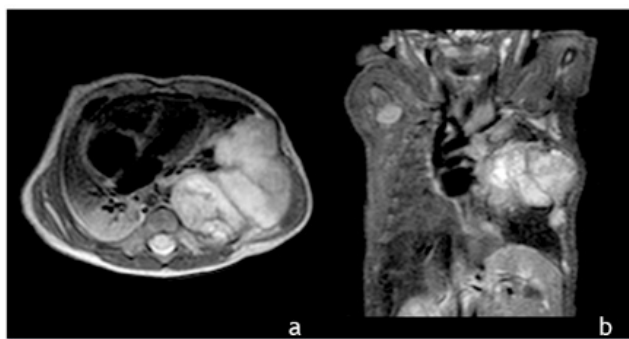
Two days after birth, chest CT was performed and evidenced a mass growth of the 5<sup>th</sup> left costal arch, extended to the underlying ribs (Figure 4 and 5); the chest MRI (Figure 6 and 7), performed in the same day, confirmed the suspect of MHCW. A US-guided fine needle aspiration (FNA) biopsy with 18G Biomol® was performed on the 8<sup>th</sup> day of life and revealed typical features of MHCW.



**Figure 4:** A bone window of the CT scan, performed two days after birth, shows the calcifications inside the lesion and the disruption of the 5<sup>th</sup> left rib (white arrow).



**Figure 5:** Volume rendering 3D CT, performed two days after birth, shows an anterior (a) and a posterior (b) view of the MHCW. It highlights rib involvement and deformation of the left chest wall due to the heterogeneous lesion.

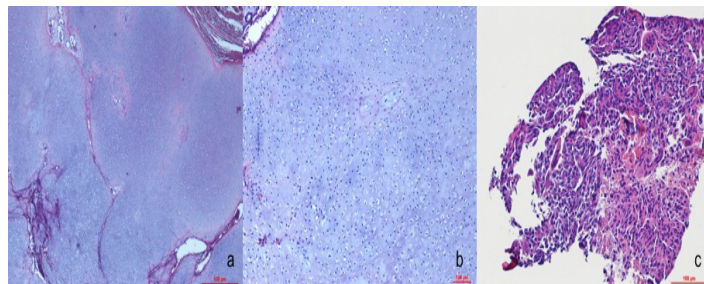


**Figure 6:** An axial view Turbo Spin Echo (TSE) T2weighted (a) and a coronal T2weighted STIR (b) of the MRI performed two days after birth, shows the complex of Aneurysmal Bone Cyst (ABC) surrounded by inhomogeneous tissue. It is evident the right shift of mediastinum.



**Figure 7:** A coronal view of the MRI performed two days after birth (fast field echo). T2\* weighted sequence shows the mass arising from chest wall and involving ribs, occupying the left hemithorax. It is important to note the dishomogeneous signal of the upper part of the mass, due to hemorrhage inside ABC and calcification of the cartilage matrix.

The first decision was to establish a conservative follow up of the lesion. Though, two weeks later, the baby presented an important worsening of respiratory status; for this reason, it was decided to perform a debulking of the lesion. The baby underwent a left posterolateral thoracotomy over the 6<sup>th</sup> intercostal space; the mass involved the 5<sup>th</sup> and the 6<sup>th</sup> ribs and consisted of an intrathoracic and extrathoracic component. The intrathoracic part was surgically removed until the costal plane and the left lung was totally rehabilitated. The postoperative period was uneventful and the baby was extubated on the 10<sup>th</sup> postoperative day. The histological examination (Figure 8) confirmed the initial diagnosis of MHCW. Microscopic examination revealed a neoplasm composed of mature hyaline cartilage with a nodular/lobular pattern of growth. No atypical or binucleated cells were present. Mitotic activity was absent. In addition, there were areas of Aneurysmal Bone Cyst (ABC), formed of hemorrhagic dilated cystic spaces and containing reactive bone and osteoclast-like giant cells.



**Figure 8:** a) and b) Microphotograph of MHCW shows solid areas composed primarily of mature hyaline cartilage and disposed in a nodular/lobular fashion. No atypia, mitosis or binucleated cells were present. H&E, original magnification 4x (a) and 10x (b).

c) Microphotograph of MHCW shows the typical features of ABC-like areas composed of stromal cells and multinucleated giant cells. H&E, original magnification 20x.

After one month, the baby showed a sudden deterioration of his ventilation with respiratory distress. A new CT scan was performed with evidence of an increase in dimension of the lesion, probably due to hemorrhage of aneurysmal cyst within the residual mass. For this reason, it was decided to re-operate the baby.

A new left posterolateral thoracotomy over the 4<sup>th</sup> intercostal space, using the previous skin incision, was made; the mass showed adhesion with the pleura, but the lung was free from tumor. The lesion was completely excised with a resection of the 4<sup>th</sup>, the 5<sup>th</sup> and the 6<sup>th</sup> ribs and of the pleural component; an extemporaneous examination confirmed the nature of the lesion. The defect in the chest wall was repaired with a Gore-Tex patch. The post-operative course was regular and the child was discharged after 25 days. The patient was subsequently followed-up with regular medical examination, that showed parameters of growth and respiratory

function within the limits; a thoracic MRI was performed every six months for the first year and every year for the next two years. The first MRI reported a residual mass arising from the 7<sup>th</sup> rib; the left lung volume and dimension were superimposable to the one of right lung. The subsequent MRIs revealed spontaneous reduction of the mass. (Figure 9 and 10).



**Figure 9.** Postoperative MRI scan obtained two years after surgery. The residual lesion is heterogeneous and it shows two large ABCs with decrease in lung volume. The atelectasis is related to general anesthesia. Sagittal, Coronal and Axial T2 weighted imaging.



**Figure 10.** Postoperative MRI scan obtained three years after surgery. It is evident a spontaneous reduction of the lesion with an increased amount of ventilated lung. There is no scoliosis while only a minimal chest wall deformity is evident. Sagittal, Coronal and Axial T2 weighted imaging.

## Discussion

MHCW is a rare benign lesion of the chest wall, arising from one or more ribs, usually from the central part [1, 2]. Its incidence is less than 1 in million in general population, although it may be underestimated due to misdiagnosis; it is more frequent in male than female (ratio 2:1 to 4:1) [3-5]. It usually occurs prenatally or within six months, with only 2 cases reported in adulthood at age 39 and 60 [2-4, 6, 7]. To date, just over 100 cases have been reported and only in 15 cases, including ours, the mass was detected prenatally [1, 2, 8-21]. Although a biopsy is required in all cases, the diagnosis may be achieved solely with radiological examinations [1]. The correct diagnosis is crucial to establish management [22]. It should not be considered a true neoplasm, since it consists of maturing, proliferating normal skeletal elements with no propensity for invasion or metastasis [4, 22]. We review literature cases of prenatal detection of MHCW and reported the major findings in the table (Table 1).

|                             | GA at detection (weeks) | Prenatal exams | Diagnostic features (presumptive diagnosis)                                                                                                                                                  | Sex | Side  | N° of lesions | Change of size                               | Prenatal complications | Perinatal complications                                                                                                 | Postnatal investigations                                                                                                               | Treatment after delivery                                                                 | Follow-up/Complications                                     |
|-----------------------------|-------------------------|----------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----|-------|---------------|----------------------------------------------|------------------------|-------------------------------------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------|-------------------------------------------------------------|
| Brar et al. [8]<br>1988     | 35                      | US             | Mass of heterogeneous echogenicity (parts similar to bones), displacement of the right lung and fetal heart, involvement of the CW<br><br>DS: mediastinal neuroblastoma or thoracic teratoma | M   | Right | 1             | Ns                                           | PH<br>PE               | C-section at 37Ws due to fetal distress<br><br>Respiratory distress at birth (intubation)<br><br>Deformity of the chest | X-ray and CT scan: mass involving right ribs (T1-T10), left shift of mediastinum, secondary scoliosis<br><br>DS: MHCW                  | Needle biopsy confirmed MHCW<br><br>Debulking at 1 month of life                         | No PO complications<br><br>Follow-up: 15 days after surgery |
| D'ercole et al. [9]<br>1994 | 29                      | US             | Heterogeneous mass with hyperechogenic areas, displacement of fetal heart                                                                                                                    | M   | Left  | 1             | 4cm (prenatal)<br><br>9x5x5.5cm (IO finding) | PE                     | C-section due to abnormal fetal heart rate at the beginning of labor<br><br>Deformity of the chest                      | X-ray and CT scan: partially calcified mass involving CW but not the lung, partial destruction of the ribs, right shift of mediastinum | En-bloc resection (including 3 ribs) at 12 days of life<br><br>Histology documented MHCW | No PO complications<br><br>Follow-up: Ns                    |
| Jung et al. [10]<br>1994    | 35                      | US             | Ns                                                                                                                                                                                           | M   | Right | 1             | Ns                                           | PH                     | C-section at 37Ws due to preterm labor and fetal distress<br><br>Respiratory distress at birth                          | Ns                                                                                                                                     | Debulking at 1 and 9 months of life<br><br>Histology documented MHCW                     | Ns                                                          |

|                              |                           |                      |                                                                                                                                                                                        |   |           |          |                                      |                                       |                                                                                                                                       |                                                                                                                                 |                                                                                                                                   |                                                                                                                 |
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| Masuzaki et al. [11]<br>1996 | 36                        | US<br>CT scan (38Ws) | Internal and external CW mass with bony density arising from the ribs, displacement of the right lung and fetal heart<br><br>DS: MHCW                                                  | M | Right     | 1        | 9cm (prenatal)<br>9x5cm (IO finding) | PH PE (trans-abdominal thoracentesis) | Induced vaginal delivery at 38Ws due to PE<br><br>Deformity of the chest<br><br>7 days after birth, respiratory failure; recurrent PE | Ns                                                                                                                              | En-bloc resection (including the 4 <sup>th</sup> and the 5 <sup>th</sup> ribs) at 13 days of life<br><br>Histology confirmed MHCW | No PO complications<br><br>Follow-up: 30 days after surgery                                                     |
| Rose et al. [12]<br>1996     | 2 <sup>nd</sup> trimester | US<br>Fetal MRI      | Multicystic hyperechoic mass extruding from CW<br><br>DS: atypical gastroschisis, resorbing thoracopagus twin, lymphangioma or hemangioma<br><br>Extracardiac lesion overlying sternum | F | Ns        | 1        | 18x10cm (postnatal)                  | None                                  | C-section at 36Ws due to preterm labor for rupture of membranes<br><br>Cystic mass of the chest composed of pedunculated lesions      | CT scan: lobulated soft tissue mass superficial to the subcutaneous tissue associated with sternal cleft and deviated clavicles | Complete resection of the lesion at 10 hours of life and closure of sternal defect<br><br>Histology documented MHCW               | No PO complications<br><br>Follow-up: Ns                                                                        |
| Lisle et al. [13]<br>2003    | Ns                        | US                   | Fetal thoracic mass                                                                                                                                                                    | F | Bilateral | Multiple | Ns                                   | PH                                    | 37+5Ws at birth<br>Respiratory distress at birth<br>Tracheostomy due to airway obstruction (supraglottic mass)                        | X-ray, CT scan, MRI: bilateral CW masses, arising from the ribs, areas of dense calcification and large blood-filled spaces     | Incisional biopsy at 5 days of life documented MHCW<br>Observation                                                                | Follow up: 6 years<br><br>Reduction in size of chest lesions<br><br>Repeated tracheostomy due to laryngeal mass |

|                               |    |                        |                                                                                                                                  |   |       |          |                                                      |                            |                                                                                                                                                                                               |                                                                                                            |                                                                                                                                                        |                                                                                                                       |
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| Shimotake et al. [14]<br>2005 | 28 | US<br>Fetal MRI (32Ws) | Heterogeneous mass with solid and cystic components, displacement of the right lung and mediastinum, no display of the left lung | F | Left  | 1        | Ns                                                   | PH                         | C-section at 36Ws due to preterm labor<br><br>Respiratory distress at birth with persistent pulmonary hypertension due to pulmonary hypoplasia (intubation and HFO)<br>Deformity of the chest | X-ray: mass involves the left hemithorax including all 12 ribs and thoracic vertebrae (T1 - T10)           | Incisional biopsy at 5 days of life documented MHCW<br><br>Observation                                                                                 | Follow-up: 24 months<br><br>No respiratory symptoms<br><br>Deformity of ribs and vertebrae                            |
| Odaka et al. [15]<br>2005     | 29 | US                     | Heterogeneous mass, displacement of the right lung                                                                               | M | Right | 1        | 3.8x4.5 cm (29Ws)<br>5x4cm (post-natal)              | PE (pleuro-amniotic shunt) | C-section at 33Ws due to fetal distress<br><br>Respiratory distress at birth                                                                                                                  | X-ray and CT scan: a solid and cystic CW mass with calcification, involving posterior portions of the ribs | En-bloc resection (including posterior portion of the 8 <sup>th</sup> and the 9 <sup>th</sup> ribs) at 8 days of life<br><br>Histology documented MHCW | No PO complications<br><br>Follow-up: 51 days after surgery                                                           |
| Braatz et al. [16]<br>2010    | 24 | US                     | Chest mass growing faster than body growth                                                                                       | M | Right | Multiple | 40ml (24Ws)<br>63ml (at birth)<br>106.9ml (6 months) | None                       | Planned C-section at 38+3Ws<br><br>Respiratory symptoms<br><br>Deformity of the chest                                                                                                         | CT scan: multifocal tumor with moulding and erosion of the ribs<br><br>DS: MHCW                            | CT guided biopsy confirmed MHCW<br><br>Observation                                                                                                     | Follow up: 9 months<br><br>Increase in size of chest lesions (slower than body growth)<br><br>No respiratory symptoms |

|                                    |           |                          |                                                                                                                                                                                                                                                       |   |           |          |                                                                                    |                             |                                                                               |                                                                                                                                                       |                                                                                                                                                                            |                                                             |
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| Chu et al. [17]<br>2011            | 24        | US<br>Fetal MRI (34+3Ws) | Heterogeneous mass with echoic capsule Lobulated, well-circumscribed extrapulmonary mass involving a midthoracic rib with multiple fluid- fluid levels and haemorrhage or calcification Displacement of the left lung and mediastinum<br><br>DS: MHCW | F | Bilateral | Multiple | 3.3×3.3 ×3.1cm (34Ws)<br><br>4.6×3.9 ×4.1cm (21 days of life)<br><br>(the largest) | PE                          | Spontaneous vaginal delivery at term<br><br>None                              | X-ray and CT scan: mass with calcified rim, involving ribs with distortion, heterogeneous enhancement<br><br>Multiple lesions                         | Resection of the largest lesion with CW reconstruction at 6 weeks of life<br><br>Histology confirmed MHCW<br><br>Observation of the two smaller lesions                    | No PO complications<br><br>Follow up: Ns                    |
| Martínez-Varea et al. [18]<br>2012 | 23 (Twin) | US<br>Fetal MRI (23+5Ws) | Supradiaphragmatic vascular cystic formation<br><br>Several cysts hyperintensive at T2<br><br>DS: CCAM type 1                                                                                                                                         | M | Right     | 1        | 3.4x2.6cm (23Ws)<br><br>6x5x5cm (postnatal)                                        | Hydrops fetalis and ascites | C-section at 30+5Ws due to preterm labor<br><br>Respiratory distress at birth | X-ray and CT scan: right hemithorax opacity, mass with calcium density involving CW with costal erosion and tracheal deviation<br><br>DS: MHCW        | No treatment due to parent's decision<br><br>Autopsy confirmed MHCW                                                                                                        | Death                                                       |
| Wie et al. [19]<br>2013            | 23+1      | US                       | Heterogeneous mass adjacent to the ribs, above the diaphragm, initially echogenic in whole area, then hypoechoic in the central area and echogenic in the peripheral capsule, indicative of calcification                                             | F | Left      | 1        | 1.4x0.8cm (23Ws)<br><br>3.5x3.2cm (36Ws)<br><br>4.5x3.7cm (postnatal)              | None                        | Vaginal delivery at 38Ws<br><br>Mild CW retraction                            | CT scan: heterogeneous mass with peripheral calcification arising from the 8 <sup>th</sup> rib<br><br>DS: osteochondroma, chondroma or chondrosarcoma | En bloc resection (including the 8 <sup>th</sup> and the 9 <sup>th</sup> ribs) and CW reconstruction with Goretex patch at 6 days of life<br><br>Histology documented MHCW | No PO complications<br><br>Follow up: 10 days after surgery |

|                               |    |                     |                                                                                                                                                                                                 |   |           |          |                                                                                        |      |                                                                                      |                                                                                                              |                                                                                                                                                                                                                                                                                                                     |                                                                                                                                                 |
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| Jozaghi et al.<br>[1]<br>2013 | 28 | US                  | Right mass consisting of solid and cystic components with calcified rim and a hyperechoic core (hemorrhage)<br><br>Left cystic mass with fluid-fluid levels, increasing in size<br><br>DS: CCAM | F | Bilateral | Multiple | Right one: 2.3cm<br><br>Left one: 3.8x3.5 x3.7cm (Prenatal)                            | PE   | Induced vaginal delivery at 39Ws<br><br>Deformity of the chest                       | X-ray, CT scan: bilateral multiple lesions with peripheral bony rims and remodeling of the ribs DS: MHCW     | Thoracoscopic assisted excisional biopsy of the right lesion and CW reconstruction with Goretex patch at 4 days of life                                                                                                                                                                                             | No PO complications<br><br>Follow up: 2 years<br><br>Reduction in size of the left lesion<br><br>No respiratory symptoms, CW or spine deformity |
| Bieda et al.<br>[20]<br>2013  | 39 | US<br><br>Fetal MRI | Large and heterogeneous mass                                                                                                                                                                    | F | Right     | 1        | 7x5.3cm (39Ws)<br><br>6.7x5.8 x5.6cm (postnatal)<br><br>9.3x8.1 x7cm (5 weeks of life) | None | C-section at 39Ws<br><br>Respiratory distress at birth<br><br>Deformity of the chest | X-ray and MRI: multicystic calcified mass arising from the CW, shift of mediastinum, destruction of the ribs | Open biopsy at 1 <sup>st</sup> day of life<br><br>Histology documented MHCW<br><br>Hemorrhage of the lesion with inferior vena cava compression<br><br>En- bloc resection (including from the 6 <sup>th</sup> to the 10 <sup>th</sup> ribs), and CW reconstruction with bovine pericardium patch at 8 weeks of life | No PO complications<br><br>Follow up: 36 months<br><br>No recurrence, respiratory symptoms or spine deformity<br><br>Deformity of the right CW  |

|                |      |        |                              |                                                                                                                                                                                                                              |   |      |   |                                                              |    |                                                                             |                                                                                                                                                                                                                                                            |                                                                                                                                                                                                                                                                                                 |                                                                                                                                                                                            |
|----------------|------|--------|------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---|------|---|--------------------------------------------------------------|----|-----------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Present report | 2016 | 35 + 5 | US<br><br>Fetal MRI (36+5Ws) | Mass of heterogeneous echogenicity, displacement of the left lung and fetal heart<br><br>Lesion arising from CW, involving ribs, with heterogeneous solid and cystic compounds and intrathoracic development<br><br>DS: MHCW | M | Left | 1 | 4.8x5.6 x4.7cm (36+5Ws)<br><br>5x4.4 x4.5cm (2 days of life) | PH | Planned C-section at 37Ws<br>Respiratory distress<br>Deformity of the chest | X-ray, CT scan, US:<br>heterogeneous mass arising from 5 <sup>th</sup> rib, occupying left hemithorax with calcifications and right shift of mediastinic structures<br><br>MRI: heterogeneous mass with ABC and soft tissue, hemorrhage and calcifications | US-guided FNA biopsy at 8 days of life and incisional biopsy at 20 days of life confirmed MHCW<br><br>Respiratory distress due to relapse<br><br>En-bloc resection (including from the 4 <sup>th</sup> to the 6 <sup>th</sup> ribs) and CW reconstruction with Goretex patch at 2 months of age | No PO complications<br><br>Follow up: 3 years<br><br>MRI: residual mass arising from the 7 <sup>th</sup> rib<br><br>No respiratory symptoms or spine deformity<br><br>Minimal CW deformity |
|----------------|------|--------|------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---|------|---|--------------------------------------------------------------|----|-----------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|

**Table 1.** GA: gestational age; CW: Chest Wall; DS: Diagnosis of Suspected; M: Male; F: Female; Ns: Not Specified; PH: Polyhydramnios; PE: Pleural Effusion; Ws: Weeks of Gestation; PO: Postoperative; IO: Intraoperative.

The routine prenatal US plays a crucial role in detecting congenital thoracic malformations. In the early fetal age, the MHCW has a homogeneous hyperechogenic signal; later during the pregnancy, the mass increases in size and becomes heterogeneous [19]. Growing up, the lesion may involve costal pleural with lung compression, leading to hypoplasia of the lung, mediastinal shift and pleural effusion which rarely requires a fetal treatment [1, 11, 15, 18]. In addition, the localization of MHCW is high suggestive, usually arising from the posterior chest wall and often affecting multiple continuous ribs [19]. Despite these peculiar features, the definitive prenatal ultrasonographical diagnosis of MHCW is challenging due to the rarity and lack of familiarity with the fetal MHCW and the others chest wall lesions [1]. The fetal MRI is more sensitive than US in revealing the characteristics and origin of the mass [18]. The lesion usually exhibits a heterogeneous pattern; the cystic compounds show high signal intensity in T2weighted images, with fluid-fluid level, and possible artifacts in T2\*weighted imaging for the presence of hemosiderin deposits and calcifications. The solid mesenchymal compounds may display low signal intensity in T1weighted and T2weighted images due to fibrous tissue; high signal intensity on T1weighted images can be related to bleeding and loss of signal in T2weighted images may be suggestive for hemorrhage or calcifications and cartilage compounds. Although a fetal detection of the lesion was made in all the cases mentioned above, a prenatal diagnosis was obtained only in 3 cases, including ours [11, 17]. Differential diagnosis

can be not simple, including congenital cystic adenomatoid malformation, pulmonary blastoma, neuroblastoma (especially for the lesions located near to the spine), primitive neuroectodermal tumor and congenital fibrosarcoma [18, 22].

MHCW may develop as an intrathoracic and/or extrathoracic mass, usually as a unique lesion, ranging in size from a few to several centimetres, affecting more frequently the right hemithorax [16, 23]; bilateral and multiple ipsilateral lesions have been described and may be confused with a malignancy [16, 23, 24]. Variable growth patterns of MHCW are described, with generally a rapid initial growth followed by slower growth, arrest, or regression; usually, the MHCW develops in the fetal age, increases quickly between the 28<sup>th</sup> and 36<sup>th</sup> week of gestation and grows variably in the 1<sup>st</sup> and 2<sup>nd</sup> year of life [1, 2]. Postnatally, the patient may present evidence of chest wall deformity or mass associated or not with respiratory distress and/or cardiac compromise due to mass effect [1, 25].

In postnatal age, the typical radiographic appearance of MHCW is a well-circumscribed, extrapleural, heterogeneous lesion, including solid and cystic elements and areas of calcification, arising from the central portion of one or more ribs; the mass may be associated with distortion/destruction of the adjacent ribs and compression of pulmonary parenchyma without invasive behaviour [18, 22, 24].

Cross-Sectional Imaging (CT and MRI) show more accurately

costal origin, associated extrapleural soft-tissue masses, relationship with adjacent structures and composition of the lesion; CT best detects the matrix mineralization and skeletal alteration, whereas MRI best demonstrates secondary ABC areas, fibrous tissue and cartilage [13, 22]. The diagnosis may be supported by histological findings, as in all cases reviewed, including ours. Needle biopsy, incisional biopsy or excisional biopsy are all suitable modalities to obtain a tissue diagnosis. Although the Fine-Needle Aspiration (FNA) is discouraged from many authors due to the difficulty to obtain adequate material for diagnosis, in our case it was useful for diagnosis [2]. The main problem of biopsy is the risk of bleeding, since the mass is highly vascularized and can erode the endothelium lined vascular spaces [1]. Treatments include different options, from conservative management, preferred in asymptomatic cases, to surgical resection and thermal radio-ablation in symptomatic cases [3, 16, 20, 24, 26].

The surgical approaches range from partial or complete surgical mass excision to a wide en-bloc excision of the chest wall, involved ribs, intercostal muscles, pleura and neurovascular bundle [3, 25]. The rationale of surgery is based on possible tumor growth. Moreover, the prognosis after surgery is excellent and any chest wall defect can be repaired with prosthetic meshes or muscle flaps: scoliosis and chest wall deformity are the main long-term postoperative complications in the extended resection of the posterior and lower ribs [3, 16]. Recurrence is rare and reported after incomplete resection, as in our case; in these cases, a more aggressive treatment may be necessary.

The prognosis of MHCW is excellent. In literature, only 3 cases of death have been described, in two cases due to respiratory distress and in one case due to systemic infection secondary to chemotherapy performed for a misdiagnosis [2]. Scoliosis and deformity of the chest are the major long-term complications reported in very large untreated lesions or after surgical resection of a large mass [1, 3, 14, 16, 20]. No other short and/or long-term complications have been described, although a long-term follow-up is missing in most cases.

## Compliance with Ethical Standards

### Conflict of Interest

The authors declare that they have no conflict of interest.

### Informed Consent

“Informed consent was obtained from all individual participants included in the study.”

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