EXTRALOBAR PULMONARY SEQUESTRATION REVEALED BY PRENATAL HYDROTHORAX

Mouna chelli Bouaziz*, Sonia Sghaier*, Dalenda Chelli**, Mohamed Fethi Ladeb*, Hela Chelli**

** Service de radiologie. Institut Kassab d'orthopédie. Ksar Said Tunisie*
*** Serivce de Maternité A. Centre de Maternité et de néonatalogie. Tunis. Tunisie*

M.Chelli Bouaziz, S.Sghaier, D.Chelli, M.F.Ladeb, H.Chelli

SÉQUESTRATION PULMONAIRE EXTRA-LOBAIRE RÉVÉLÉE PAR UN HYDROTHORAX PRÉNATAL

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RÉSUMÉ

Prérequis : La sequestration pulmonaire est une anomalie congénitale rare qui peut être diagnostiquée in utero.
But : Raportez une nouvelle observation
Observation : Les auteurs rapportent un cas de séquestration pulmonaire extralobaire révélée par un hydrothorax et décrivent l'aspect prénatal et post-natal de cette affection selon différentes techniques d'imagerie. (échographie-Doppler obstétricale, imagerie par résonance magnétique foetale, angioscanner multicoupe psot-natal)

MOTS-CLÉS

Photosensibilité - Metformine - Antidiabétiques oraux - Effet indésirable

SUMMARY

Background : Pulmonary sequestration is a rare congenital pulmonary anomaly that can be diagnosed in utero.
Aim : Report a New case
Case report : In this case report of extralobar pulmonary sequestration, the authors report a case revealed by hydrothorax and describe this disease appearance in different imaging technique (Doppler ultrasonography, magnetic resonance imaging and post-natal multislice CT angiography).

KEY-WORDS

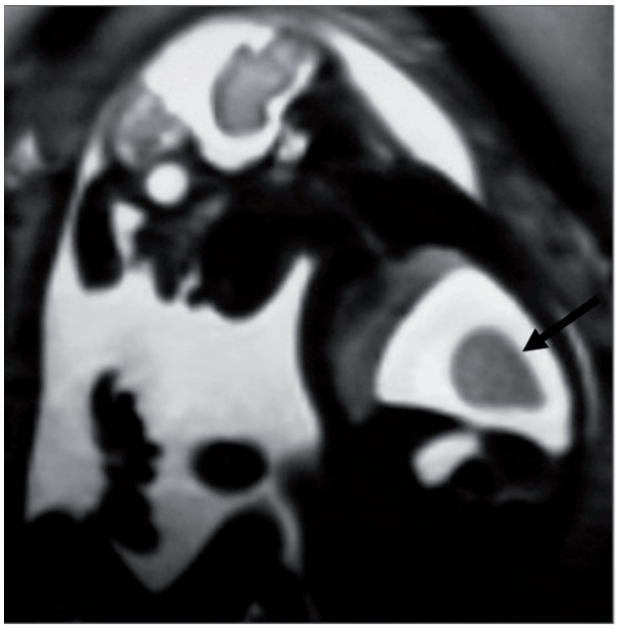
Photosensitivity - Metformin - Oral antihypoglycemic drugs - adverse drug Effect

Pulmonary sequestration is a rare congenital abnormality in the spectrum of congenital broncho-pulmonary malformation. Prenatal diagnosis can be made either by ultrasound (US) or magnetic resonance imaging (MRI). The imaging features of pulmonary sequestration have to be known by obstetricians as well as radiologists to allow screening and sometimes positive diagnosis of this anomaly. The aim of this article is to report one case of extra-lobar pulmonary sequestration, which was diagnosed in utero by US and MRI with focus on the imaging characteristics of this rare anomaly.

CASE REPORT

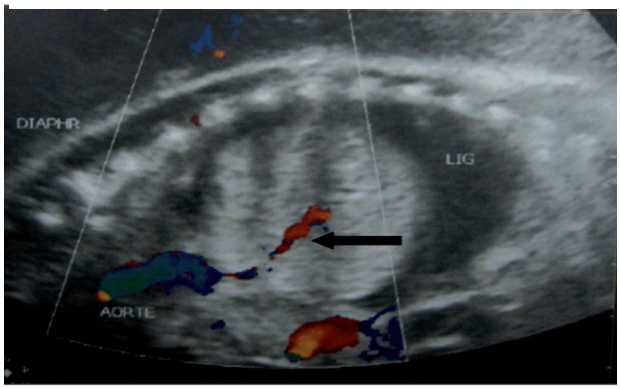
A 26 year-old primipara has been referred by her obstetician to the radiology department to undergo fetal MRI because left pleural effusion had been detected on an ultrasound examination performed at 31 weeks of amenorrhea. MRI was performed using a 1 Tesla magnetic field, with multiplanar T2 single-shot turbo spin echo sequences (SSTSE) (HASTE: Siemens Medical Systems). It revealed a well defined triangular left lower lobe thoracic mass in shape showing a high T2 signal intensity compared with normal homolateral and contolateral lung but a lower signal than surrounding pleural fluid (Fig.1).

Figure 1: Sagittal prenatal MR image (sequence HASTE): left-sided triangular pulmonary mass of increased signal intensity surrounded by hydrothorax.



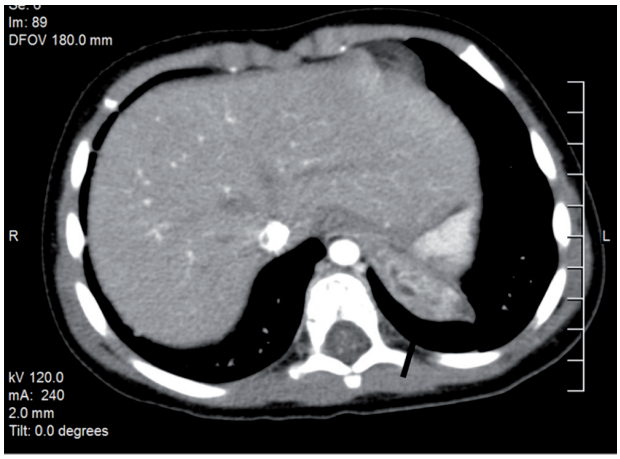
Moreover, both left pulmonary lobes and mediastinum were displaced by the pleural effusion. MRI did not show any diaphragm or other thoracic abnormality. The diagnostic of pulmonary sequestration was suspected on MRI and a complementary exploration by Doppler US was indicated.
This US examination was first made in bidimensional B mode, followed by a color Doppler US using a 3 to 5 Mhz abdominal probe. The inferior lobe left thoracic mass appeared as a well defined triangular hyperechoic homogeneous mass surrounded by pleural effusion. Color Doppler showed the arterial supply of this mass from the abdominal aorta (Fig.2). The diaphragm was of normal appearance on US.

Figure 2: Left sagittal sonogram of fetal chest: echogenic pulmonary mass and visualisation of systemic feeding artery arising from the abdominal aorta.



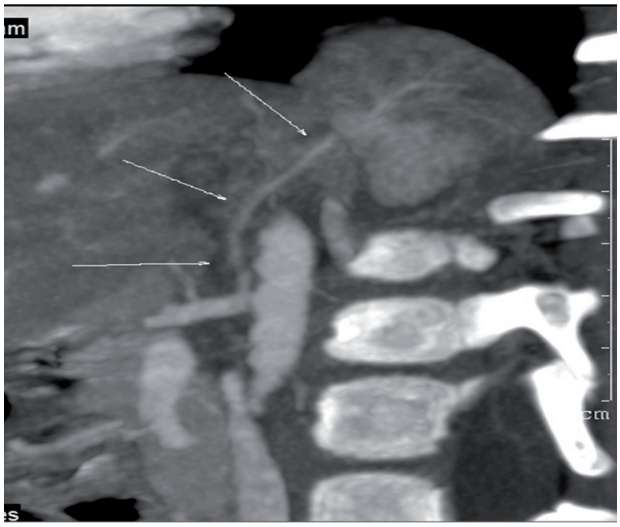
The patient gave birth to a male healthy baby by vaginal delivery and without any perinatal complications. The child was free of repiratory symptoms. However, at the age of 18 months, pediatric surgeons indicated a surgical treatment and a pre-operative multislice angio-CT has been performed. This examination was made by a 6-slice volume acquisition from the pulmonary apex to the symphisis pubis, with a 6x2 mm collimation, 20 ml contrast media intravenous injection, 3 ml/second injection rate and a 7 second injection delay determined « bolus tracking » method.
On angio-CT, the pulmonary anomaly appeared as a well circumscribed fusiform mass of the left lung inferior lobe surrounded by a pleural sheath, which confirmed the diagnosis of extra-lobar pulmonary sequestration.
The spontaneous density of this mass was evaluated at 40 UH with an important enhancement after intravenous contrast administration (120 UH) (Fig.3a). Multiplanar 2D reformations

Figure 3 a: Multislice CT Angiography: Transversal contrast enhanced CT view: well-defined mass of the left pulmonary lower lobe enhanced by intravenous contrast injection



showed the arterial ssupply of this mass originating from the initial portion of the coeliac artery. The venous drainage was made via the hémi-Azygos vein (Fig.3b).

Figure 3 b: Multiplanar reformations: the mass receives a blood supply from an aberrant artery originating from the coeliac trunk.



The initially indicated surgical excision of the sequestrated lobe was not performed because the child remained completely asymptomatic. An annual clinical and radiographic follow up was recommended.

DISCUSSION

Pulmonary sequestration is a rare malformation accounting for 0.15 to 6.45% of all pulmonary congenital anomalies [1] but representing 8 [2] to 23% [3] of prenatally diagnosed pulmonary anomalies. This entity is defined by the presence of a non-functioning lung tissue that is separated from the normal trachea-bronchial tree and receives its vascular supply from an aberrant systemic artery.

Two types are distinguished:

- intra-lobar sequestration is the most frequent (75%). It is usually contained within the visceral pleura with the homolateral lung and has a pulmonary venous drainage. Arterial supply originates from the thoracic descending aorta (73%) [4]. Clinical symptoms may appear in young adults with productive cough and recurrent pneumonias [4].
- Extra-lobar sequestration (also called Rokitansky lobe or accessory lung) (25%) is contained in its own pleural sheath and has a systemic venous drainage. This form is usually discovered in infancy, 60% of cases in the 6 first months of life. The child may present with a dyspnea and/or cyanosis or remain totally asymptomatic, as in our case [4].

With the development of prenatal diagnosis, the number of pulmonary sequestrations discovered in utero and reported in the world literature is increasing, and particularly those accompanied with a polymalformative syndrome.

The extra-lobar form représents 25% of pulmonary sequestration [4] and involves girls more frequently than boys [1]. The discovery gestational age ranges between 19 and 31 weeks with a mean of 23 weeks [2].

The sequestrated lobe is usually intra-thoracic, rarely intra-abdominal [4]. The left lung is more frequently involved [1,4]. Bilateral forms have also been described [2].

Extralobar pulmonary sequestration usually appears on US as a round or triangular well circumscribed mass. It may be either hyperechoic and homogeneous or of heterogenous echostructure containig anechoic liquid areas. This latter appearance may correspond either to the sequestration itself or to an associated congenital cystic adenomatoid malformation [5]. A pleural effusion sometimes compressive may also be observed [4, 5] with displacement of the mediastinum and diaphragm due to venous and lymphatic obstruction. US with use of Color Doppler is the method of choice to demonstrate the characteristic association of a hyperechoic thoracic mass and a systemic arterial branch originating from the thoracic or abdominal aorta and directed to the sequestrated lobe [2,4,5]. According to Ruano and al. [6] the use of 3D energy Doppler may facilitate the detection of this arterial branch. US helps to determine the venous drainage type (pulmonary or systemic) [7] and also allows an accurate detection of associated malformations, the most frequently described being congenital diaphragmatic hernia, congenital cystic adenomatoid and bronchogenic cyst [4,5]. US may be limited by an advanced pregnancy age or a maternal obesity. Ultrasonographic distinction between a pulmonary sequestration with a cystic component and a congenital cystic adenomatoid may be rather difficult as well as the detection of the systemic aberrant artery when it is has a little diameter.

Pulmonary sequestration usually appears on fetal MRI as a well circumscribed round or triangular lesion having a low T1 signal intensity and a relatively high T2 signal intensity [5]. The pulmonary anomaly may show either homogenous or heterogenous MRI signal. Cystic areas may be present in relation with coexistent adenomatoid lesions [5]. In all cases, the systemic arterial supply cannot be demonstrated by fetal MRI [8]. However, the main goal of MRI is to diagnose associated abnormalities when they are present.

The follow up during pregnancy is made by repeated US examinations to detect compressive pleural effusion wich may also induce fetal hydrops by cardiac and venous caval compression [5].

Positive diagnosis is now possible with multislice angio-CT which tends to supplant classical lung angiography as angio-CT offers a good spatial resolution and allows multiplanar reformations résolution. Angio-CT confirms the diagnosis and helps to identify the arterial and venous supply and thus the sequestration type. Moreover, vascular assessment is essential for pre-operative planning and aims to reduce per-operative hemorrage.

The spontaneous outcome of pulmonary sequestration does not usually compromise vital prognosis. Some cases of prenatal spontaneous resolution have been described [5]. When a compressive pleural effusion is detected in the fœtus, in utero pleural drainage is possible. In non regressive forms, surgical

excision is usually performed to avoid infectious and haemorragic complications.

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