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Antenatal diagnosis of a giant intra-abdominal cystic lymphangioma (Case report)

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Abstract: *Cystic lymphangioma is a benign congenital malformation rare of the lymphatic system, the reported incidence is 1/16,000 birth. Typically, cystic lymphangioma is located in tissues soft of the head and neck on the other hand the intra-abdominal location is very unusual with a percentage of 2 to 3% whose location mesenteric remains the most common followed by the omentum then the mesocolon and finally the retroperitoneum. The diagnosis of mesenteric cystic lymphangioma is the most often mentioned during the first years of life with a clinical picture polymorphic ranging from simple abdominal pain to a symptomatic occlusion with peritonitis, while there are few cases diagnosed in prenatal described in the literature. On the occasion of an illustrative clinical observation, we report the case of a giant intraperitoneal cystic lymphangioma diagnosed in antenatal by an obstetric ultrasound performed on a 2nd parent 33 weeks of amenorrhea. We discuss the antenatal signs which should raise suspicion of diagnosis and lead to magnetic resonance imaging (MRI), as well as the care to be adopted pre- and postnatally.*

Keywords: Cystic lymphangioma, antenatal diagnosis, prognosis, differential diagnosis

1- INTRODUCTION

Lymphangiomas are benign tumors of the lymphatic vessels that have the potential to infiltrate surrounding structures.(1) .
They can be classified into three groups: simple lymphangiomas, made up of capillary-sized channels; cavernous lymphangiomas, made up of dilated lymphatic channels, often covered with a fibrous adventitial covering; and cystic lymphangiomas or hygromas, composed of multiple cysts of variable size lined with endothelial cells.
They can affect almost any area of the body where lymphatics are present. They are located in the neck (cystic hygroma), head and the armpits, in 95% of cases. Among lymphangiomas located below the diaphragm, 63% are left-sided.
Prenatal diagnosis is important because it makes it possible to plan the birth in a level 3 structure provided with rapid postnatal resuscitation is possible, in order to improve the prognosis.
It allows the choice to terminate the pregnancy if a poor outcome is anticipated.
We report the case of a voluminous Intraperitoneal cystic lymphangioma diagnosed by ultrasound at 33 weeks of gestation.

2- CLINICAL CASE

A 34-year-old parturient, first degree consanguineous marriage, 2nd procedure, with a living child gives birth by vaginal delivery for macrocrania, currently with a ventriculoperitoneal shunt, who presented for prenatal care at 30 weeks gestation.

A detailed ultrasound performed at 33 weeks of gestation + 1 day according to a precise last period date revealed an abdominopelvic mass lateralized intraperitoneal on the right side associated with bi-ventricular hydrocephalus (Figure 1).

The mass was large, multilocular, multiseptate and hypo-echoic, suggesting a lymphangioma, measuring 8x9 cm. She was responsible from a repression of the hepatic parenchyma backwards and the agglutinated grelic loops, it comes into contact with the portal trunk and encompasses the vein

of arantus, in contact with the right kidney, also represses the right pulmonary hemi-field upwards (figure 2).

The patient gave birth vaginally at 36 weeks, of a newborn male weighing 2100g with a large fluctuating mass taking all the abdomen.

A head circumference of 21 mm with bulging fontanels. A fetal blood sample with a fetal karyotype was 46, XY.

He died on D2 of life due to respiratory distress. Histology of the mass revealed large vascular spaces lined by endothelium containing lymphocytic aggregates.

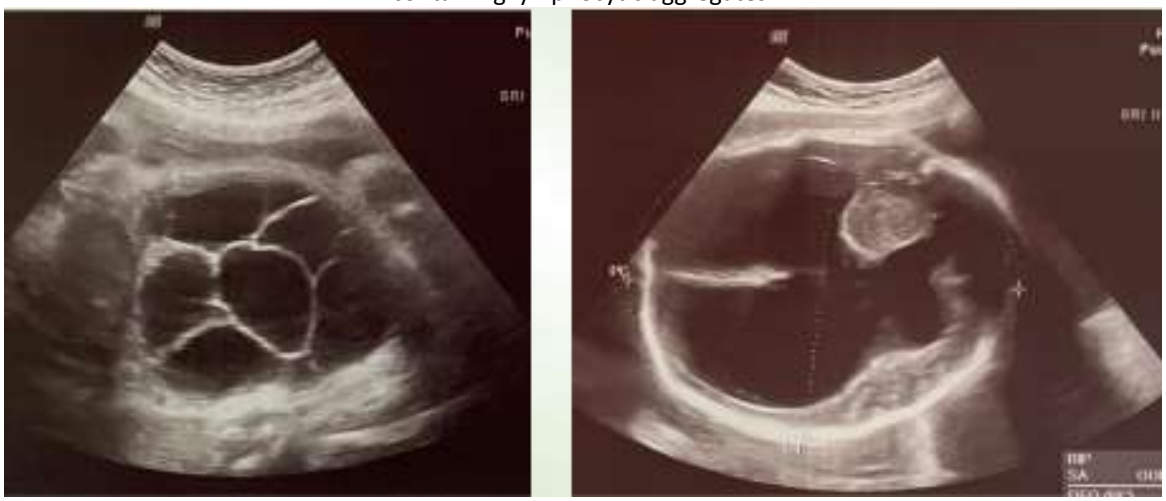

Figure 1 : ultrasound image showing a cystic image on the right multilocular intraabdominal of 9 cm and on the left a bihydrocephalus significant ventricular lamination of the cerebral parenchyma facing

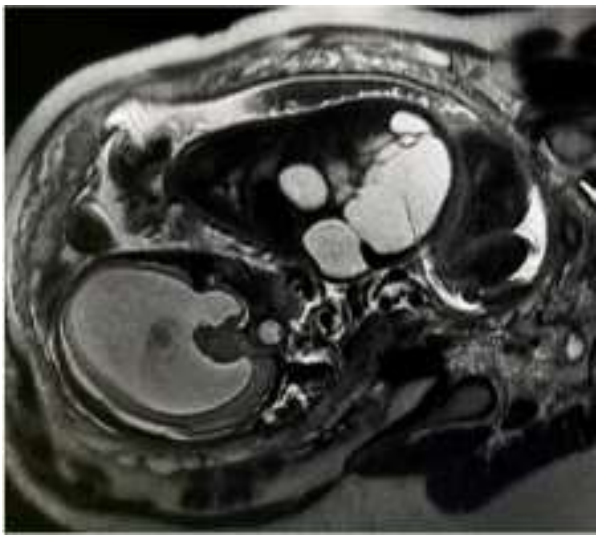

Figure2: Fetal magnetic resonance imaging: large, extensive septate multilocular cystic formation on the intra abdominal wall associated with hydrocephalus

3- DISCUSSION

Cystic lymphatic malformations (CLMs) are rare, slow-flowing, benign congenital malformations composed of dilatations abnormal cysts. The term “lymphangiomas” has been abandoned. (2)

The precise etiology of MLK is poorly understood. They are thought to result from a failure in the development of the lymphatic pathways, which develop usually from the 6th week of gestation, leading to proximal dilatation of the afferent ducts.

Other mechanisms Proposed pathogenic causes include the inability of the embryonic lymph sacs to re-establish communication with the venous system or the aberrant budding of the primordial lymphatic sac. (3)

These lesions can vary in size, from small collections of fluid to huge cysts like the case in our fetus; those who are located in loose connective tissue may become larger than the rest of the fetus with a giant cystic lymphangioma measuring a MRI 9cmx 8cm.

Although these lesions are benign, they can compress adjacent vital organs. In utero, the characteristic ultrasound appearance is that of a hypo-echoic mass with thin walls and multiple compartments.

The prenatal diagnosis of these lesions is rarely reported in the literature (5).

In order to improve the reliability of antenatal diagnosis, it is recommended to combine MRI with ultrasound. It makes it possible to evaluate specifies the locoregional extension, depth and associated malformation thus predicting the prognosis and guiding the proposals therapeutics.

Establishing a definitive ultrasound diagnosis is complex due to the great diversity of diagnoses. differentials (4) such as choledochal cyst, ovarian cyst, renal cyst and dilatation or duplication of the intestine.

At our parturient, Antenatal ultrasound made it possible to precisely identify the lymphangioma and define its anatomical location and extent. She also makes it possible to monitor the growth of this lesion, and this will make it possible to decide on the prognosis which may be poor in the event of rapid growth.

Other abnormalities, including skin edema, fetal hydrops, and polyhydramnios, may be associated with these lesions. In the absence of other associated malformations, LMK is considered as an isolated malformation without karyotype abnormality. That is why it is only in the event of other associated anomalies that a genetic consultation can be offered to the couple and a fetal karyotype can be performed. carried out.(6)

The prognosis is good when the karyotype is normal and hydrops is absent. Lymphangiomas detected in the second and early third trimester have a poor prognosis like the case of our parturient, due to a stronger link with karyotype abnormalities, but detection in the middle or end of the third trimester is associated with a good prognosis.

Postnatal treatment of lymphangioma depends on the site and extent of the lesion. Large but localized lymphangiomas may be completely excised. Surgical treatment of diffuse and multiple lesions is extremely difficult and associated with morbidity and high mortality.

Many adjuvant therapies for the treatment of lymphangiomas have been described with variable results. Of the Positive results have been reported after the use of intralesional bleomycin, sclerotherapy or percutaneous embolization (7).

8- Conclusion:

Cystic lymphatic malformations are slow-flowing congenital vascular malformations. They can be symptomatic from the prenatal period or later in life, most often in childhood. Prenatal diagnosis is fundamental for planning treatment.

Early multidisciplinary charge with collaboration between obstetricians, neonatologists and pediatric surgeons.

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