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Prenatal Ultrasound Diagnosis Of Spina Bifida

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Abstract: *Spina bifida or spinal dysraphism is a neural tube closure anomaly that affects 1/1500 of the population, a congenital malformation that occurs during the first four weeks of embryogenesis. Ultrasound is of great interest in this pathology, allowing most often an antenatal diagnosis, this diagnosis is based on the search for direct signs of this pathology often oriented by the visualization of certain indirect signs that constitute alarm signs. The management of this disease is primarily based on the search for other associated malformations, allowing to establish a prognosis and to propose an adapted management. In this work we are interested in the antenatal ultrasound diagnosis of this pathology***.**

Keywords: spina bifida; spinal dysraphism; antennal ultrasound diagnosis.

1. INTRODUCTION

Spina bifida or spinal dysraphism is an anomaly of neural tube closure that affects 1/1500 of the population [1], a congenital malformation that occurs during the first four weeks of embryogenesis. It consists of a defect of closure of the posterior vertebral arches. Several risk factors have been identified [2].

There are several forms of varying severity. This malformation is accessible to a prenatal diagnosis which is based on a detailed morphological ultrasound. Two main clinical forms are defined, the occulta and cystica forms. isolated or associated with malformations, notably cerebral, which will allow the antenatal diagnosis to be well oriented.

The prognosis depends on the type of dysraphism and the importance of the malformations, and the treatment of certain forms is currently possible and can be done antenatally.

2 Clinical case:

Mrs. A.Y. has no particular history, notably no notion of consanguineous marriage, no use of antiepileptic drugs. G2P1. The first pregnancy was normal with vaginal delivery of a male child with good psychomotor development. Who consulted for follow-up of her current pregnancy estimated at 26 weeks of amenorrhea. The course of the current pregnancy was without particularity with the notion of taking folic acid in pre-conception and prolonged during the whole first trimester, in preventive dose. The ultrasound of the first trimester was performed without any morphological anomaly detected and with a fine nuchal translucency. During the current consultation, the patient underwent a detailed morphological ultrasound which showed the presence of a cystic mass, with fine heterogeneous content, attached to the lumbosacral spine (figure 1), opposite which we find on an axial section opening behind the bony part of the medullary canal (figure 2). In view of this aspect, the diagnosis of myelomeningocele was evoked. Other signs were then sought: an Arnold Chiari II anomaly (figure 4), colpocephaly and club feet were found (figure5).

The patient was informed about the risk of this pathology and the prognosis, with regular follow-up until spontaneous delivery at 36 weeks in our maternity hospital, of a male child with myelomeningocele, referred to pediatric surgery for further management (figure 6).

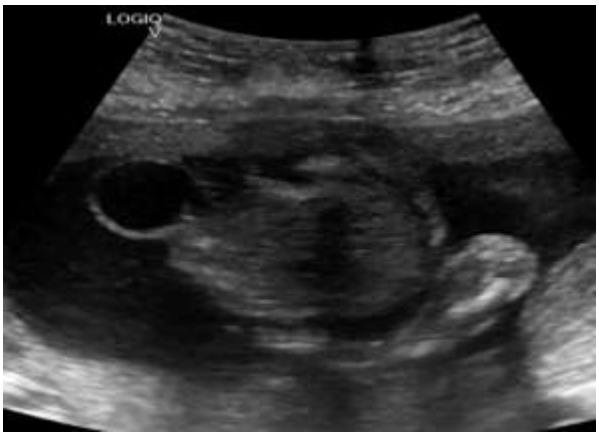

Figure 1: lumbosacral cystic mass with fine echogenic content, associated in the lower part of the image with a club foot.

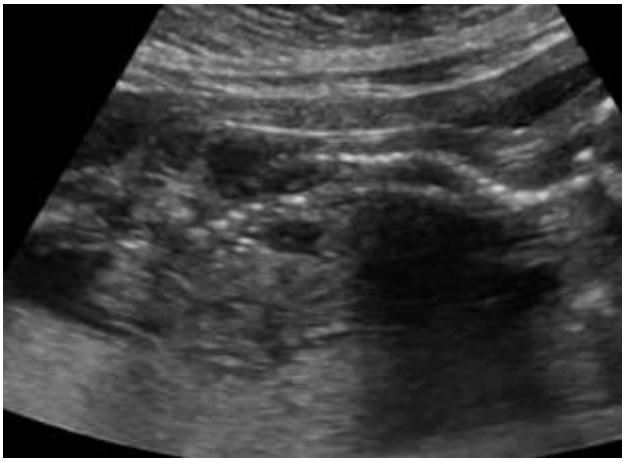

Figure 2: spinal deformity with loss of continuity at the lumbosacral level.

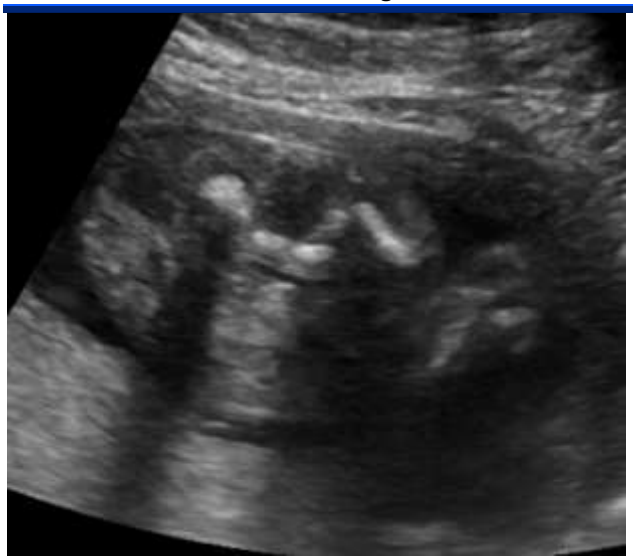

Figure 3: Loss of continuity of the posterior bony arch, a sort of open V is seen in the back.

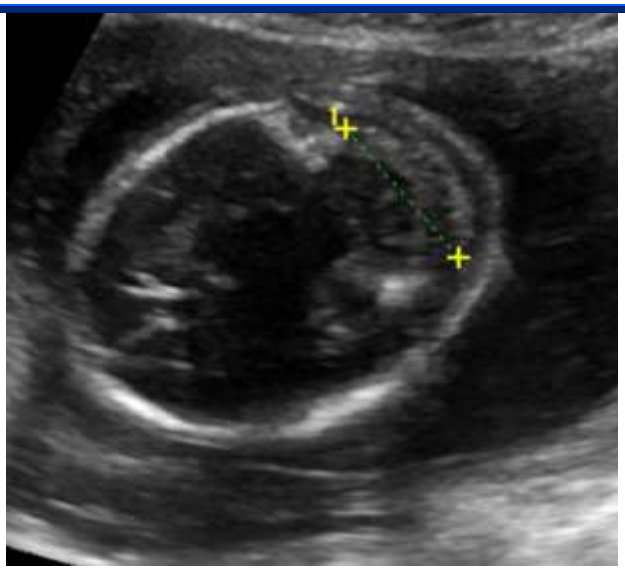

Figure 4: Arnold Chiari II anomaly. The cerebellum takes the shape of a banana palmed against the occipital bone with disappearance of the large cistern.

3 Discussion

Spina bifida is one of the most frequent congenital anomalies compatible with long-term survival. Secondary to a neural tube closure defect often at the lumbosacral level, it forms a spectrum of pathology ranging from occult forms with no apparent lesion to forms with a totally open spine or rachischisis with severe motor and sensory deficits through cystic forms of varying severity [5].

There are two main classes:

Spina bifida occulta: often in the lumbosacral region, the meningeal and nervous structures are in their usual location, the visualization of a dimple, a tuft of hair or a deviation of the gluteal sulcus are often present. This form is often asymptomatic and of fortuitous discovery

Spina bifida cystica or cystic spina bifida presents as a cystic mass often attached to the lumbosacral spine and sometimes covered by the skin, the contents of which will define the meningocele, where there is a herniation of the meninges without nerve tissue, or myelomeningocele when the meninges and nerve tissue are present in the sac. This form is always symptomatic and depends essentially on the level of the spinal cord defect.

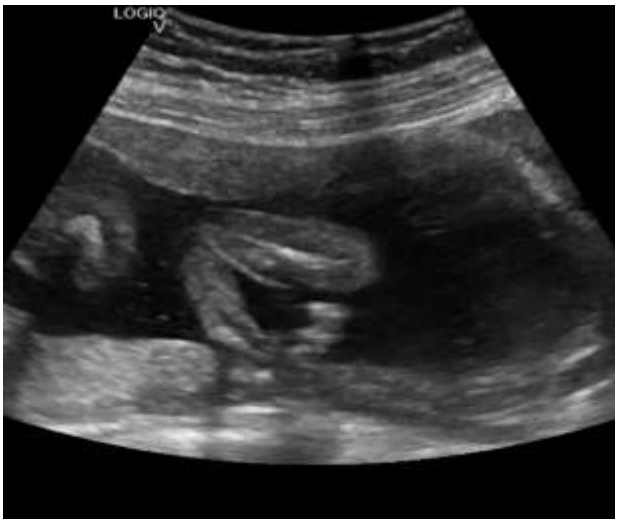

Figure 5: clubfoot malformation of the lower limb.

This anomaly is accessible to prenatal screening by fetal morphological ultrasound, usually performed at 22 SA [6]. In the case of our patient, the ultrasound was performed at 26 weeks, as the patient had no particular clinical context with a normal T1 ultrasound. An earlier ultrasound may be requested around 16 to 17 days of age if the alpha-feto protein levels are high.

The ultrasound diagnosis is often attracted by the presence of indirect signs, notably cerebral: a lemon-shaped aspect of the cranium by flattening of the frontal bones, this sign disappears towards the end of T1. An Arnold Chiari II type malformation which consists of a cerebellum flattened against the occipital bone giving it a banana shape with disappearance of the large cistern, and a dilatation of the lateral ventricle visualized on

the occipital horn, called colopocephaly or water drop horn. The presence of clubfoot is sometimes visualized.

In the presence of these indirect signs, a careful scan of the spine in all three planes is necessary.

The sagittal scan of the spine finds an abnormal swelling adjacent to the spine, often with abnormal angulation of the spine. The follow-up of the skin continuity is essential, we can find a discontinuity of the skin covering in the rachischisis, or a prolongation in the hernia sac in the cystic forms, sometimes a simple deformation is visible. On axial section, a V-shaped opening of the posterior arches is visible (figure3).

The analysis of the hernial contents is very important since this analysis is part of the prognosis. A menningocele is seen as a bag with purely liquid anechoic content, in the myelomeningocele the content is heterogeneous with the presence of several echogenic threads corresponding to nerve fibers, the presence of an echogenic image is a sign of a spina lipoma. A check of the level of the terminal cone of the bone marrow is important to look for a low attached marrow.

Finally, the ultrasound must look for malformative associations, in particular urinary obstructive type, conotruncal type cardiopathies [3].

In the case of our patient all the direct and indirect ultrasound signs were highlighted (figures 1 to 5), on the other hand the assessment of associated malformations was negative.

The biological assessment made by the dosage of serum markers of the 2nd trimester: the alpha-feto-protein (AFP) finds all its interest in the screening, a high rate of AFP must have alarmed on the risk of a defect of closure of the neural tube, thus a high rate is associated with 95% of the cases of anencephaly, and up to 80% of the cases spina bifida [4].

The AFP assay is therefore interesting to alarm the sonographer during the T2 ultrasound, its indication after the ultrasound is performed may seem useful to refine the diagnosis [7] [9].

In the case of our patient, the AFP assay was not performed since the ultrasound diagnosis was obvious.

The prognosis of this pathology depends on the type of spina bifida, and the associated malformations. Thus, in the occulta type fruste form the fortuitous discovery is often the case. In the cystic form, the risk is both neonatal, associated with compression of the brain stem in the case of an Arnold Chiari II malformation responsible for respiratory distress, and the risk of infection in the case of absence of epidermalization of the hernial sac [8]. In addition, there is a functional risk due to motor, sensory and sphincter deficits.

Management begins with delivery in a type 3 maternity hospital, and the treatment varies from an indication for IMG to neonatal management. In utero surgery techniques are currently being evaluated which consist of intrauterine skin closure but with a considerable risk of prematurity [10].

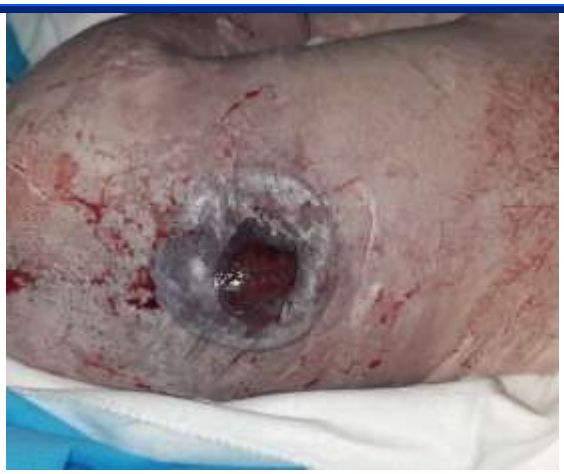

Figure 6: lumbar spina bifida cystica not eperdimized.

4 Conclusion

The spina befida is a frequent pathology, in the diagnosis of the severe forms can be done easily with the ultrasound. A diagnosis which will allow to discuss the neonatal prognosis is to propose a management, and especially to give an enlightened information to the patient and to organize a childbirth in the most optimal conditions.

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