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Ectopia Cordis Thoraco-Abdominal Diagnosed with Antenatal Ultrasound

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ABSTRACT

In this article we report a case of ectopia cordis, which is a rare congenital malformation characterized by an extra-thoracic position of the heart. This is the thoracic-abdominal form, of antenatal discovery on a pregnancy of 35 weeks and 03 days, during an obstetric ultrasound monitoring pregnancy during the third trimester of pregnancy, made at the University Hospital Professor Zafisaona Gabriel, Mahajanga Madagascar. The objective of this work is to report the antenatal ultrasound diagnostic elements of this rare malformation. Ultrasound plays an important role in antenatal diagnosis and the search for other related malformations, especially in low-income countries.

INTRODUCTION

Ectopia cordis or cardiac ectopy is an extremely rare congenital cardiac malformation, characterized by a partially or completely extra-thoracic topography of the heart, whose prevalence is estimated at 5.5 to 7.9 cases per million live births (Hornberger L K. *et al.* 1996). Depending on the topography, it can be cervical, thoracic, thoraco-abdominal or abdominal. The thoracic-abdominal form is generally associated with the pentalogy of Cantrell or one of its variants which includes a bifid sternum, a defect of the diaphragm, the anterior abdominal wall and an intracardiac malformation (Apte AV. 2008). We report a case of isolated thoracic-abdominal cordis ectopia, diagnosed with antenatal ultrasound at 35 weeks and 3 days. To our knowledge, this is the first case documented and reported in Madagascar. The objective of this work is to report the antenatal ultrasound diagnostic elements of this rare malformation. Ultrasound plays an important role in antenatal diagnosis and the search for other related malformations, especially in low-income countries.

METHODOLOGY

This is a 37-year-old woman from Marovoay, Mahajanga, in the west of Madagascar, without any notion of inbreeding marriage, already having five living children, the first of whom is 17 years old and the last of 3 years old. She's being referred by a city medical office for an obstetric ultrasound. This is her sixth move, without a history of spontaneous or induced miscarriage. No history of birth defects was reported in siblings and family. The pregnancy was followed by three series of prenatal consultations in health facilities in the countryside, which had not objectified any particular abnormalities. She had received two ultrasound examinassions during the first and second trimesters, the results of which are normal. There was no medication or abnormal symptomatology during pregnancy. Clinically, it is in good general condition, without any particular functional sign. There is no exaggeration of uterine height. There was also no abdominal pain, water loss, or genital bleeding. The abdominal ultrasound showed a pregnant uterus, with a live fetus in cerebral presentation, of 35 SA and 3 days depending on the measurements performed. Amniotic fluid is normal volume with a PHELAN index measured at 13 cm. There is a 65 mm long anterior thoracic abdominal parietal defect in median sagittal section with totally extra-thoracic heart chambers (Figures 1 and 2), with regular activity at 148 beats per minute. There is also a strip of part of the hepatic parenchyma up to the level of the umbilical region next to the abdominal parietal defence. In Mode B ultrasound there were four visible heart chambers and no other malformative abnormalities visible at the head pole, abdomen, upper and lower limbs. There were also no placental and umbilical cord abnormalities. Doppler scanning could not be performed due to a lack of suitable equipment. In total, it is a progressive monofetal pregnancy of 35 SA and 03 days of age ultrasound with malformation type abnormality of closure of the anterior chest-abdominal wall associated with ectopia cordis thoraco-abdominal, without any other associated visible malformation. Given the complexity of the management of these malformations and the insufficient technical plateau in the hospitals of Madagascar and the lack of social coverage for such management. An in utero transfer to a well-equipped hospital was planned by pleading aid to international

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humanitarian associations, but the parturiente was lost sight of it.

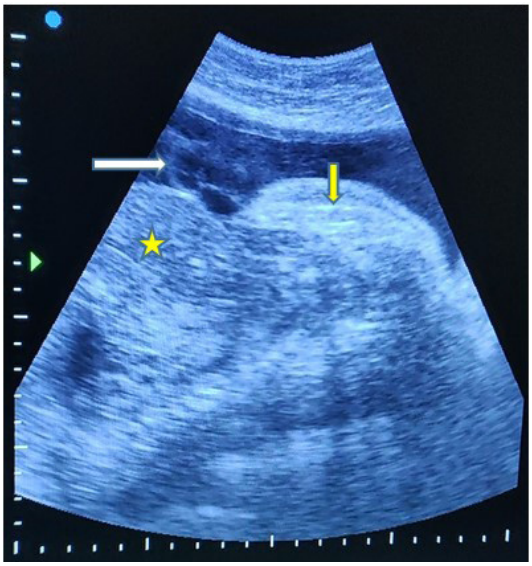

Figure 1: sagittal ultrasound through the fetal sternum (yellow arrow), showing a posterior thoraco-abdominal parietal defect with extra-thoracic localization of the heart cavities (white arrow) without pericardial sheath and exposing part of the liver (yellow star).

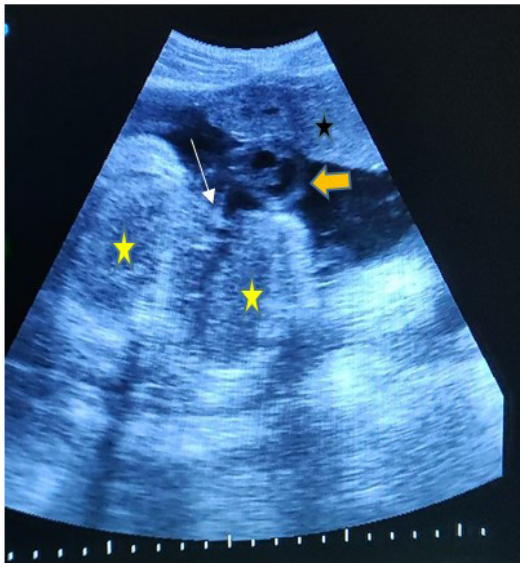

Figure 2: axial ultrasound cross-section through the lower thoracic region of the fetus, showing pulmonary parenchyma (yellow stars), anterior parietal defect (white arrow), extra-thoracic heart chambers bathed in amniotic fluid (yellow arrow), and a placental edge (black star).

DISCUSSION

The ectopia cordis may be complete ; in this case, the naked heart is moved outside the rib cage, without pericardial coverage. One can also have a partial displacement or one can see the heart beating through the skin. This anomaly was first described by Haller in 1706 (Kaplan L C. *et al.* 1985, Chelli D. *et al.* 2008) and the first classifications were attributed to Weese in 1818 (Harrison M R. *et al.* 1982) and Todd in 1836 (Leca F. 1989). The cause is unknown; however, there is a small association noted with trisomy (Twomey E L. *et al* 2005, Diaz JH. 1992) and a few cases have been associated with other abnormalities, such as triploidy and X-related family heredity. The incidence is 5.5 to 7.9 per 1 million live births (Apte AV. 2008). A defect is present in the maturation of the mesodermal midline and a defect of fusion of the cephalic anterior folds can lead to associated abnormalities. The failure of the descent of the heart into the thorax during the 3rd week of gestation may leave it stuck above the upper closure of the sternum, creating a cervical ectopy but more difficult to explain for thoracic and abdominal ectopies (Diaz JH. 1992). It can be classified into cervical (3%), cervicothoracic, thoracic (60%), thoracoabdominal (7%) and abdominal types (30%) (Leca F. 1989, Morrello M.1994). Although the thoracoabdominal types and minor forms of Cantrell pentalogy offer a better prognosis, the cervical type is not compatible with life (Morrello M.1994). In our case, it is the complete thoraco-abdominal form, without any other visible malformation associated with the B-Modeultrasound.

Early ultrasound diagnosis can often be done before birth and is usually noted early in the second trimester (Harrison M R. *et al.* 1982). With the use of ultrasound (3D) and its combination with Doppler power, accurate diagnosis may be possible at an early stage of gestation (Mittermayer C. & Bernaschek G, Lee A. 2002). Liang and colleagues reported one case of CORDIS ectopia diagnosed at 10 weeks gestation using 2D and 3D ultrasound (Liang RI *et al.* 1997). MRI becomes a pillar of prenatal assessment for documenting and planning complications (Liang RI *et al.* 1997). In our case, the antenatal diagnosis is done quite late, at 35 weeks and 3 days gestation, with a portable ultrasound, equipped only with a convex probe, without 3D option or Doppler. Our diagnosis is then limited to the morphological description of the parietal defect and the search for other associated malformations, without being able to specify the existence or no intra-cardiac malformations.

When the diagnosis is made before birth, a caesarean section should be performed because vaginal delivery may result in prolonged cardiac compression, or rupture of an atrial diverticula or omphalocele sac (Sharma VK. *et al.* 2001). If uncorrected intracardiac abnormalities are found, this allows the physician and family to discuss the elective termination option. The complete ectopia cordis presents itself as a neonatal emergency. It is generally considered fatal and death can occur as a result of infection, heart failure or hypoxemia. The associated cardiac abnormalities are interauricular communication (100%), interventricular communication (53%), Fallot tetralogy (20%), and left ventricular diverticle (20%) (Sharma VK. *et al.* 2001). The treatment consists in placing the newborn under a radiant radiator and the defect of the chest wall must be covered with a sterile gauze soaked with warm saline solution. Compression of the extrathoracic heart should be avoided as there may be an overall reduction in cardiac function. When managing

the airway, you should expect difficult intubation secondary to the anterior and cephalic direction of the heart. If a prenatal diagnosis is made, one can electively intubate at the time of delivery by caesarean section with maternal placental support. The surgical strategy depends on the size of the parietal defect, the associated cardiac abnormalities, and the type of ectopia cordis (Mohta A, *et al.* 1993).

In developing countries, whether the diagnosis is antenatal or post-natal, the prognosis is almost always fatal. A study of a series of 10 patients with antenatal diagnosis showed a uniformly fatal outcome without any management (Kumar Basant, 2008). The lack of equipped and functional cardiovascular surgery could explain this poor prognosis. In our case, the only chance to have a better prognosis of survival in front of such an anomaly is to carry out an in utero transfer to foreign hospitals because the technical trays in Madagascar do not y*et al*low an adequate management of these complex malformations.

CONCLUSION

Ectopia cordis is a rare birth defect, often associated with other birth defects. Ultrasound plays an important role in antenatal diagnosis and the search for other related malformations. Primarily surgical management remains a challenge, especially in low-income countries where prognosis is always reserved. When diagnosed early in utero, a high proportion of abortions is expected, hence the interest of regular pregnancy follow-up.

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