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Case Report

Perivellousa disease massive fibrin deposition, association with Down syndrome: case report and literature review

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ABSTRACT

The disease perivellousa massive fibrin deposition (MPFD), is a condition characterized by uncontrolled mainly fibrin deposition intervillous space. The incidence worldwide is 0.028% per 1000 live births, there is only one case report where this condition is associated with trisomy 21, in our country there are no reports of this disease. The MPFD has high morbidity, obstetric mortality, recurrence, as well as neurodevelopmental significance of newborns. The etiology until the moment is unknown, difficult diagnosis and management for the obstetrician. The aim is to report MPFD association with trisomy 21 (T21) and a review of the medical literature regarding this condition.

Keywords: Fetal growth restriction, Fibrin deposition, Globular placenta, Maternal floor infarction, Therapy

INTRODUCTION

Disease massive fibrin deposition in the intervillous space, some authors also described as maternal floor infarction (MFI), incidence reports in the international literature range from 0.028% to 0.5% per 1000 live births in January.¹

It is a disease with high morbidity, mortality and obstetric likelihood of recurrence; In relation to the infant also increases neonatal morbidity and mortality in later stages have been documented neurodevelopmental disorders. The etiology is unknown so far, most theories agree on an immunological component responsible for the pathogenesis. It is a diagnostic and management challenge for the obstetrician.

Because MPFD, is a disease of difficult prenatal diagnosis with important implications for both mother and fetus, association with T21 it is rare, so we consider necessary to make the following presentation of the case, in order to contribute in efforts that exist on the diagnosis and management of this disease in prenatal stages.

CASE REPORT

Patient 22 years old with no medical history of relevance, who attends the obstetrics service to present suspected pregnancy and vaginal bleeding, 10.4 weeks gestation / threatened abortion pregnancy was diagnosed. Urinary infectious process was discarded, the ultrasound shows subchorionic hematoma of 15% (Figure 1). Is managed as outpatients based rectal suppositories rest and indomethacin 100 mg per three doses, folic acid 4 mg daily was started.

From the date the patient comes to service multiple times obstetric referring colic pain and persist with vaginal bleeding, receiving the same treatment as described above. In the prenatal exams of the first trimester: uroculture and vaginal exudate without development, serological test for syphilis: negative, serological test for infection by human immunodeficiency virus: negative.

In one of the visits to the obstetrics they will perform obstetrical ultrasound control reporting: pregnancy of 12.1 weeks, subchorionic hematoma of 50% (Figure 2). Again with no evidence of urinary or vaginal infection, TORCH negative, thyroid function in normal parameters, being necessary hospitalization. She was conservatively managed on the basis of resting and indomethacin suppositories. The hospital stay was promulgated since the patient referred to obstetric pain, being discharged from hospital at 15 weeks of gestation

Triple biochemical marker was performed at week 16 as aneuploidy screening high risk throwing for trisomy 21, amniocentesis is offered to the patient to confirm the diagnosis, it rejects because of their religious affiliation. Morphological ultrasound at week 24: without sonographic markers suggestive of trisomy 21, the placenta with hyperechoic areas, quantitatively normal amniotic fluid (Figure 3).

The patient continued with the same clinical behavior, manifesting obstetric pain, without cervical changes. Receiving the same treatment as in previous occasions. At the 28th week of gestation, the fetus presents a decrease in the growth rate, being this more accentuated at week 32, reason why a scheme of fetal pulmonary maturation begins.

At week 34, type III growth restriction, severe oligohydramnios, non-tranquilizing fetal status was diagnosed, so it was decided to perform cesarean section.

Newborn female is obtained with the following somatometry: weight: 1275 grams, size: 39.5 cm, capurro: 36 weeks, apgar: 8/9, silverman: 1, clinically suggestive of down syndrome, which was confirmed by karyotype. Others surgical findings were: anhydramnios, umbilical cord 50 cm with a vein and two arteries, small placenta of fibrotic appearance.

The placenta is sent to pathology. Macroscopically: 15 cm long axis, by the maternal face 50% of it with fibrotic appearance of brownish yellow undefined cotyledons (Figure 4).

Microscopically-hyaline fibrotic component around the chorionic villi forming extensive areas hyalinisation even seen. There is the presence of residual trophoblast focal (Figure 4 and 5).

Finally, the patient has undergone the puerperium without complications, the hematologist has ruled out

antiphospholipid syndrome and thrombophilia, as well as providing genetic counseling for her next pregnancies. In relation to the neonate, she attends an extended stay in the intensive care unit due to complications associated with prematurity



Figure 1: Transvaginal ultrasound, at week 10 of gestation, subchorionic hematoma of 15% is seen at the posterior wall level in its upper portion.



Figure 2: Obstetric ultrasound, at week 12 of gestation subchorionic hematoma of 50% is seen, in its middle portion of heterogeneous aspect probably in resolution phase.

DISCUSSION

The disease perivellous massive fibrin deposition (MPFD), was first described in 1961 by Benirschke and Driscoll. It is a rare condition, and in 2014 on

classification placenta injuries, it falls under the heading of other placental Processes.² The etiology remains unknown in literature theories reference to the following is done: Cytotoxicity derived from extravillous trophoblast, allograft rejection, infections, abnormal coagulation and fibrinolysis.³



Figure 3: Obstetric ultrasound at week 24 shows an area of greater diffuse and partially delimited echogenicity towards the lower portion of the placenta suggestive of fibrosis zone.

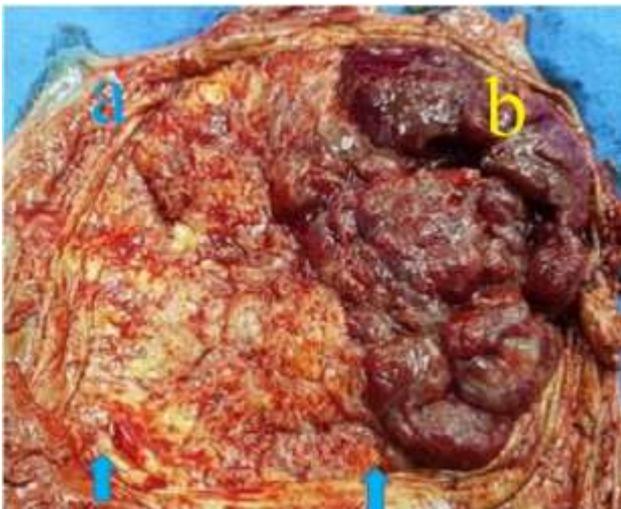


Figure 4: Macroscopic aspect of the maternal placental face, a) Left side with evident fibrosis, b) Right side cotilidenes of normal aspect.

The pathophysiological basis proposed that maternal antibodies attack the syncytiotrophoblast, causing tumor necrosis factor and Hageman factor release, they trigger the coagulation cascade continuously at the same time the ability to fibrinolysis is disturbed leading to progressive deposition and uncontrolled intervillous fibrin in place.⁴

Patients with MPFD confirmatory diagnosis is performed by pathology. At the macroscopic description the placenta is small, white-yellowish, thickened and rigid, the area of

fibrosis is friable, granular, healthy placental tissue on the other hand is soft and spongy.⁴

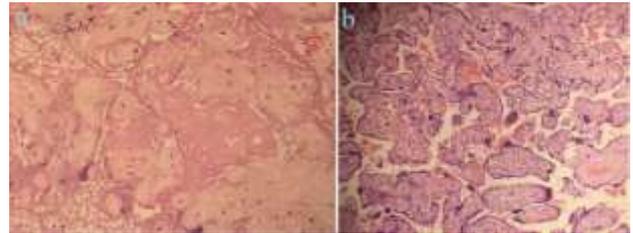


Figure 5: a) Perivillous fibrosis, hyalinization and sclerosis surrounding the villi. b) Comparative. The right side shows tertiary chorion villi.

In an attempt to establish causality, some authors have associated with thrombophilia MPFD, so far the results are controversial, however, if the association has been documented consistently with antiphospholipid antibody syndrome (PAPS).^{5,6}

Unfortunately so far there is not any cabinet or biochemical that can be used to diagnose the disease during pregnancy test. As described above MPFD is a condition that presents recurrence up to 60%, so we must be diligent in obstetric examination during antenatal care.

On the other hand, it should be considered during prenatal consultation that these patients are enrolled in high levels of alpha-fetoprotein, so some screening tests for chromosomopathies may give false positives, especially for neural tube defects.

Uncontrolled fibrin in the intervillous space, from early pregnancy, deposition causes a steady, gradual decrease in functional surface of the placenta and release of free radicals, so that these patients are at increased risk of: restriction on intrauterine growth, fetal death in utero, abortion and preeclampsia.⁷⁻¹¹

Generally these pregnancies are resolved prematurely on average between week 32-34 if it has intervened promptly, the most common cause is intrauterine growth restriction in type IV. So the mortality and morbidity for these infants is high. There are reports that cases of children of mothers who had MPFD have neurodevelopmental disorders. In the medical literature other associations of MPFD have been described, to mention a few: Renal tubular dysgenesis, Coxsackie A16 virus infection, Chronic intervillositis, polymiocytis and down síndrome.¹²⁻¹⁵

Regarding the management of these patients, there are no guidelines to date, it is recommended that patients with a history of MPFD, administer early acetylated salicylic acid to reduce the risk of preeclampsia, Enoxaparin if the pregnant woman suffers from PAPS, Sildenafil if there is restriction in Intrauterine growth, with these therapeutic

measures have been obtained favorable, but not consistent results.¹⁶

Some studies have used pravastatin to reduce oxidative stress and decrease the risk of preeclampsia, without stopping the deposition of fibrin in the intervillous space.¹⁷

CONCLUSION

Massive deposition disease in the intervillous fibrin space is a rare condition, but with high morbidity, maternal and fetal mortality, when associated with trisomy 21, the prognosis for both pregnancy and neonatal development worsens. Finally, before the histopathological report of MPFD, it is recommended to discard the mother, thrombophilia and PAPS mainly.

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