Case Reports

Non-immune foetal hydrops: a case report

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Summary

Foetal hydrops occurs when a certain amount of interstitial fluid, produced by capillary ultrafiltration, overcomes the amount of interstitial fluid that returns to the blood circulation through the lymphatic system. Hydrops is classified as immune (IH) due to the presence of circulating maternal antibodies against the foetal red blood cell's antigens, and non-immune (NIH) that includes all the other causes of hydrops. This classification is still valid, but only under a clinical point of view because they differ in aetiology and management.

In this article the management of a case of non-immune foetal hydrops is described, in which, unlike most other cases of non-immune foetal hydrops, the foetus survived.

Key words: Foetal hydrops; Immune; Non-immune.

Introduction

Foetal hydrops must not be considered a disease but a morphological description that implies the presence of a problem.

Dr. Edith Potter in 1943 made the first distinction between an immune and non-immune type of the disease. Today, due to progress in the prevention and treatment of Rh-isoimmunization disease, about 90% of foetal hydrops are of the non-immune type.

Hydrops is defined as an excessive accumulation of extracellular fluids in the foetus. The echographical diagnosis is not difficult, although there still are no proper definitive criteria; one of the most used and restrictive definitions is the following: "the presence of excessive extracellular fluid in two or more parts of the foetus associated with cutaneous oedema > 5 mm, placental thickness > 6 cm and polyhydramnios".

Hydrops is classified as immune (IH) due to the presence of circulating maternal antibodies against the foetal red blood cell antigens, and non-immune (NIH) which includes all the other causes of hydrops. This classification is still valid, but only from a clinical point of view because they differ in aetiology and management.

Since the utilization of anti-Rh immunoprophylaxis and intrauterine therapy, the frequency of immune types is very low, although they have not completely disappeared.

Hydrops is easily diagnosed, but if for the immune types the aetiology is clear (Rh isoimmunization), for the non-immune types the causes do not always justify a clear physiopathological mechanism. Therefore, it is often difficult to identify the pathogenetic mechanism and consequently the foetuses that will benefit from prenatal therapy. The perinatal mortality associated with this type of hydrops varies between 58% and 98% and has not changed much in the last few years, even after recent improvements in diagnosis and therapy (Figure 1). Moreover, morbidity and mortality are strictly related to the aetiology of hydrops, especially in the immune type.

Foetal hydrops occurs when a certain amount of interstitial fluid, produced by capillary ultrafiltration, overcomes the amount of interstitial fluid that returns to the blood circulation through the lymphatic system. Therefore hydrops is determined by those conditions that increase transudation from the blood vessels and obstruct the return of the lymph to the blood circulation.

Immune foetal hydrops is related to the presence of maternal antibodies against the foetal red blood cell antigens and today the most frequent causes of Rh isoimmunization are:

- antepartum isoimmunization (asymptomatic): 38%;
- administration of an insufficient dose of immunoglobulins: 14%;
- prophylaxis not performed: 9%.

The non-immune type of hydrops is postulated when there is no evidence of incompatibility of blood groups between mother and foetus. NIH can be a complication of a malformation due to a genetic or infectious alteration: there are more than 120 foetal abnormalities responsible for NIH.

The diagnosis of foetal hydrops is easy during an ultrasound scan, but brings us to a series of questions that do not always have an easy answer: Are we looking at hydrops or at something else? What significance must we give to this picture? What is its cause? What prognosis does this pregnancy and the next ones have? What should we tell the parents?

Today, due to the utilization of anti-Rh immunoprophylaxis, the frequency of IH is very low, although it has not completely disappeared.

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Immunoprophylaxis, if correctly carried out, can reduce sensibilisation in 99% of the cases; Therefore the clinical consequences become very rare. Due to progress in prevention and therapy, cases of IH are rapidly decreasing. In contrast, cases of NIH are increasing and therefore, today, this is the predominant type of hydrops to treat.

Unfortunately the cases that can benefit from treatment in the uterus are few because of the seriousness of this pathology.

In our study we illustrated a case of non-immune foetal hydrops in which, unlike most other cases of non-immune foetal hydrops, the foetus survived. This case was observed when analyzing cases of non-immune foetal hydrops at the Department of Gynaecological Sciences, Perinatology and Puericulture of the University of Rome "La Sapienza" between January 2001 and June 2005. This foetus was affected with congenital chylothorax.

The difference in diagnosis between chyothorax and pleural effusion is often difficult because the lymph is transparent as it does not contain fat, which is only present after the absorption of milk. In the absence of other co-existing pathologies and when the entity of the compression covers less than 50% of the pulmonary tissue there is a good neonatal prognosis. Chylothorax may be congenital (atresia of the thorax duct, ductal-pleural fistula, birth trauma), post-surgical, post-traumatic or spontaneous (tumours, fistulae, infections). Chylothorax, caused by a congenital malformation of the thorax duct or by an obstacle to the flow of the lymph in the thorax duct due to compression at its entrance into the venal circuit, is manifested in the uterus as hydrothorax. It can be caused by primal lesions of the duct, in which case an isolated pleural effusion on the right or left can be seen, or is associated with symptoms including trisomy 21, monosomy X, lymphatic or vascular abnormalities. The diagnosis is suggested by the milky aspect of the pleural liquid and confirmed by microscopic examination. The condition rarely goes away by itself and is often associated with foetal hydrops. Moreover, the presence of intra-pleural fluid makes it difficult to study the heart of the foetus because of the compression exerted by the effusion, often responsible for a mediastinic shift followed by pulmonary hypoplasia due to the growth of intra-pleural pressure. As a result foetal thoracentesis is indicated, both diagnostic and therapeutic, which permits on the one hand analysis of the effusion and on the other re-expansion of the lung, especially at an early stage. A post-paracentesis recurrence is rare.

Lesions of the thorax have the best prognosis among the disorders responsible for NIH. The survival rate is around 26%.

Case History

A 34-year-old Italian caucasian patient had a pregnancy history of one spontaneous abortion and one caesarean section at 34 weeks of gestation due to preeclampsia. Her blood type was B Rh-negative and her partner's blood type was Rh-positive; the indirect Coombs test was always negative. Routine scans were normal as was amniocentesis: 46 XY. The infectiological pattern showed negative antibodies for Toxoplasmosis, positive antibodies for Cytomegalovirus, and positive antibodies for rubella. An ultrasound scan performed at 31 weeks of gestation revealed hydrothorax, ascites and polyhydramnios (AFI 230). Thus the patient was admitted to hospital. During hospitalisation the following blood tests were carried out: indirect Coombs test which resulted negative; IgM and IgG avidity for toxoplasmosis, rubella, parvovirus B19, syphilis, and HSV, which all resulted negative for any recent infection. An ultrasound scan performed during hospitalisation showed foetal hydrops with particular pleural effusion, slight pericardic effusion, skin oedema, polyhydramnios (max pocket 10 cm), normal heart morphology, normal heart rhythm and frequency and normal foetal fluximetry showing a good oxygenation-metabolic condition. The foetal echocardiogram showed normal AV-VA alignment and absence of right or left stenosis. There was normal foetal cardiac rhythm.

After a double administration of 12 mg of betametasone a caesarean section was performed with the birth of a live female foetus who was immediately incubated. She remained in the incubator for 20 days. The newborn's blood type was AB positive and a direct Coombs test was negative. Immunoprophylaxis with 1250 IU partobulin was carried out on the mother. The newborn underwent the necessary examinations and was found to be affected with congenital chylothorax. All the other tests were normal. After 15 days spontaneous resolution of the congenital atresia of the thorax duct took place. The baby was fed intravenously for 46 days and only with the mother's frozen milk. At present the child is well.

Conclusions

Although the diagnosis of non-immune foetal hydrops should only be made when all the listed diagnostic criteria are obtained, cases of foetal hydrops which do not exhibit all the diagnostic parameters are however frequently found in clinical practice.

Once foetal hydrops has been diagnosed it is important to diagnose the difference between the immune and non immune forms. Whenever a non-immune form is present the mother and the foetus should undergo all the tests recommended by the management schedule so that the "primum movens" is discovered and consequently the couple's questions can be answered, thus reassuring them about the prognosis and possible treatment. In giving information, data from the literature should be evaluated even if there are few studies with a small number of cases and variable data. Unfortunately possible therapies are not applicable to all cases of hydrops and are often correlated to a higher risk of foetal complications.

Improvements have certainly been made, even if foetal hydrops still constitutes a clinical picture with a poor prognosis.

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