

Neonatal Lupus: 5 Pediatric Cases

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ABSTRACT

Neonatal lupus erythematosus is a rare condition that manifests itself in different types of damage, whether cutaneous, hepatic, hematological, neurological or even cardiac. We report here five cases of neonatal lupus occurring in newborns, two of them from completely asymptomatic mothers and revealed by cardiac involvement and one case revealed by skin involvement.

Key words: Neonatal lupus erythematosus; congenital heart block; newborn; antinuclear antibodies

INTRODUCTION

Neonatal lupus erythematosus is manifested by cutaneous, hematological, hepatic or cardiac involvement including possible congenital atrioventricular block (BAVc) occurring in a heart free from malformative heart disease, dilated cardiomyopathy and endocardial fibroelastosis. This rare syndrome is consistently associated with the presence in the mother of anti-SSA / Ro and / or anti-SSB / La antibodies. Skin involvement has been rarely reported in association with the isolated anti-RNP antibody.

Cutaneous and systemic damage is transient, unlike cBAV which is definitive and is associated with morbidity and mortality which makes this syndrome so serious.

We report five observations of LN occurring in newborns (NB) including two from completely asymptomatic mothers and one newborn from a mother followed for Gougerot-Sjögren syndrome.

OBSERVATIONS

Observation 1

A female newborn, from a 37-year-old mother with no pathological history. Poorly monitored pregnancy conducted at 34 weeks. Cesarean delivery for extreme fetal bradycardias. Having presented at birth with neonatal respiratory distress associated with bradycardia at 65b / min, a complete assessment was carried out, a complete blood count objectifying anemia at 11.1g / dl, without leukopenia or thrombocytopenia. A correct hepatic and renal workup and positive anti-nuclear antibodies, positive anti-SSA (Ro52 and Ro60) and positive anti-RNP. A chest x-ray objectified an aspect in favor of an early neonatal infection with pulmonary localization. An ECG objectified a 3rd degree AVB. A cardiac ultrasound showed a muscle VIC of 2 mm, an aneurysmal oval foramen. The evolution was marked by the appearance of cutaneous involvement at the age of 1 and a half made of a butterfly wing erythema of the face. The mother has a stable antibody level with no associated clinical manifestations.

Observation 2

A male newborn, from a 42-year-old mother, with the notion of 1st degree consanguinity, having a miscarriage as a history. Resulting from a well-followed pregnancy and carried to term and delivery by cesarean section for suspected heart disease due to fetal bradycardia. The newborn presented at birth a bradycardia at 50 b / min associated with respiratory distress on the 2/10 side according to the silverman score for which he was hospitalized in neonatal intensive care. A complete workup was carried out with a normal blood count, a renal and hepatic workup without abnormality and anti-nuclear antibodies positive to 1/320 of the speckled type, anti-SSA (Ro52 and Ro60) positive. An ECG objectified a 3rd degree AVB. A chest x-ray objectified cardiomegaly with an ICT of 0.64. A cardiac ultrasound showed a very altered global and segmental contractility, a foramen ovale versus CIA and a membrane on the aortic arch suggesting a coarctation of the aorta. The course was marked by the worsening of his bradycardia and his respiratory distress. The newborn died after a four-day stay in intensive care. The mother was diagnosed at this age and currently followed with stable immunological and clinical status.

Observation 3

A male newborn, from a normal pregnancy carried to term, and a mother followed for suspected lupus in front of rheumatoid purpura + dry syndrome + positive ANA with anti-SSA and anti-SSB positive. Hospitalized in neonatal intensive care at day 6 of life for a postnatal infection before: thrombocytopenia and a positive CRP. An infectious assessment was performed showing a urinary tract infection with *Escherichia Coli*. Put on antibiotic treatment for 10 days. The course was marked by the persistence of thrombocytopenia and the appearance of rounded erythematous papular lesions, covered by fine scales (figure 1). The count showed thrombocytopenia at 58,000 with anemia of 11.5 normochromium, normocytic without leukopenia. ECG and ETT did not show any abnormalities. NAAs are 1/640 positive with positive anti-SSA and anti-SSB. The evolution was towards the regression of symptoms in 4 months. The mother is watched, the hindsight is one year.

Observation 4

It is the child Nour Mallak, aged 4, asymptomatic mother. She presented for a cardiology consultation for bradycardia. On examination, the child presented with bradycardia at 59b / min without other associated signs, in particular no skin involvement. A lupus assessment was requested which objectified positive anti-nuclear antibodies. The indication of a pacemaker was asked but for lack of means not achieved. The child is stable and bradycardia is tolerated.



Figure 1: Skin involvement made up of rounded erythematous papular lesions, covered with fine scales

Observation 5

A female newborn, from a mother followed for lupus. He was hospitalized in neonatal intensive care for fetal bradycardias + IUGR. An ECG objectified a grade 3 AVB. The AANs were positive at 1/640 of speckled type, anti-SSA positive (Ro52 + Ro60) at 100IU / ml and anti-SSB positive at 100IU / ml. The biological assessment, in particular the CBC and the hepatic assessment, were normal. He presented a nosocomial infection following his hospitalization in intensive care.

DISCUSSION

Neonatal lupus is a rare syndrome, occurs in 1-2% of babies born to mothers with anti-SSA and anti-SSB antibodies, these women do not need to have clinical manifestations of autoimmune disease.

A large number of studies suggest that neonatal lupus erythematosus is caused by a transplacental passage of maternal autoantibodies at around 12 weeks of gestation, thereby reaching fetal tissue.

Cardiac manifestations

The most notorious complications of neonatal lupus involve the heart. These include first, second and third degree heart block. Neonatal lupus is the most common etiology associated with isolated congenital heart block, accounting for up to 95% of all cases.

Clinically, the fetus probably presents with bradycardia, which varies between 30 and 80 beats / min, as the first indication of a heart complication.

Cardiac involvement is mainly elucidated by congenital atrioventricular block. All three grades of block may be present but 3rd degree atrioventricular block is the most common; and these data join those of our series. Sometimes newborns with atrioventricular block in 5-10% of cases develop cardiomyopathy. [7-12]

It is believed that when maternal antibodies bind to these proteins, a marked inflammatory response ensues, injuring the AV node and causing fibrous changes that render the conduction system incompetent.

It has been reported in rare cases of other cardiac manifestations such as a patent foramen ovale or anomalies of the atrioventricular septum as is the case in our third observation. (Table 1)

Table 1: Cardiac manifestations of neonatal lupus [7-12]
Atrioventricular block Sinus bradycardia Prolonged QT interval Cardiomyopathy and congestive heart failure Structural and valve abnormalities (abnormalities of the atrioventricular septum, permanent foramen ovale, pulmonary stenosis and dysplasia of the pulmonary artery, fusion of the tendon cords of the tricuspid valve). Myocarditis

Non-cardiac manifestations

Affected children can present skin rashes found in 50% of cases, even if the lesions are characteristic, they are sometimes confused with mainly fungal infections or eczema, especially in newborns of asymptomatic mothers.

The lesions are not obvious at birth in 80% of cases and develop after 1 month. The most observed lesions are described as annular macule type. Papules or plaque-like lesions may be

seen. The contours are regular and sometimes raised. The lesions are usually 1cm in diameter and may coalesce to form a large erythematous patch. Mainly located in an area exposed to the sun with a characteristic predilection for the periorbital area. The evolution is marked by a disappearance of the lesions within a period of 7 months in 80% of cases. Sometimes skin atrophy may persist or areas of telangiectasia and hyperpigmentation in 20% of cases.

The rash goes away after the maternal autoantibodies are removed from the baby's circulation, usually without any residue.

Neonatal lupus syndrome can manifest as haematologic involvement in 20% of cases manifesting as anemia, neutropenia and thrombocytopenia, as well as a few cases in the literature which describe pancytopenia. Hepatobiliary damage can be manifested by asymptomatic elevation of transaminases, cholestasis or hepatomegaly in 15 to 25% of newborns with neonatal lupus. Both haematological and hepatic damage are transient and associated with cardiac or skin damage in the majority of cases and can exceptionally be isolated. [4-6]

The data in the literature strongly agree with those of our case, which had skin involvement that appeared after a period of 1 month associated with haematological involvement made up of thrombocytopenia and anemia. The evolution was marked by a disappearance of the lesions after 4 months with good skin condition and without complications.

Gastrointestinal involvement may manifest as increased transaminases, cholestasis or hepatitis. The severity of the disease ranged from a slight elevation in liver enzymes to severe liver failure. The prognosis for hepatic injury is generally excellent, but cases of death following hepatic failure have been reported.

Neurologic involvement may be associated, including hydrocephalus, macrocephaly, hypocalcaemic crises, vasculopathy, spastic diplegia and vasculitis resulting in stroke as well as neuropsychiatric disorders. Transcranial ultrasound should be used to screen for potential neurological problems in infants born to mothers with anti-RO and anti-LA antibodies.

Management consists of close monitoring for cutaneous, hepatobiliary and haematological manifestations. A transfusion in red blood cells and platelets in the event of symptomatic anemia and thrombocytopenia.

Sun protection measures should be applied in the event of skin damage, sometimes the use of topical corticosteroids or the use of hydroxychloroquine is necessary. [7]

In atrioventricular block, a pacemaker is indicated above all in severe bradycardia in newborns and infants. [13] Over 90% of cases have a pacemaker inserted. The use of injectable immunoglobulins in the prevention and treatment of cardiac damage is under study.

CONCLUSION

Our observations report three interesting cases of neonatal lupus, two of which originate from a completely asymptomatic mother and revealed by fetal bradycardia. As well as a case revealed

by a cutaneous and hematological attack whose mother is followed for a syndrome of Gougerot-Sjögren.

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