Neonatal Lupus Associated with Dyslipidemia

Neonatal lupus syndrome (NLS) is a rare disease characterized by an autoimmune process associated with the presence of auto antibodies in maternal fetal circulation against proteins SSA/Ro and SSB/La.

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A male patient, three months old, born at term gestation without complications, was taken by his mother for medical evaluation due to a two-month evolution, large, bilateral, erythematous, fine-scaling mask-like plaque on the face, which was worsened by sun exposure (Figs. 1,2). After long evaluation, the possibility of Neonatal Lupus Syndrome was suspected and ANA of the patient was collected. Serum sample was initially rejected due to its unexpectedly heavy hyperlipidemic nature, which persisted in subsequent samples. Hemogram and Lipidogram tests were collected, presenting the following results: eight percent eosinophilia, 70 percent lymphocytosis, and triglyceride levels of 976mg/dL. ANA was ultimately tested and demonstrated positive in 1:640. Skin biopsy confirmed the diagnosis of Lupus. Electrocardiogram has shown no impairment. Parallel mother evaluation demonstrated speckled-patterned ANA anti-Hep 1:2560 and anti-Ro 1:3200, despite lack of rheumatologic disease diagnosis and symptoms.

No nephrologic impairment was revealed during evaluation of the child. Skin lesions remitted after four months of treatment. Triglyceride levels reached normality after eight months. The patient was subjected to evaluation every three months during his first year of age and semiannually until four years old and remained asymptomatic during monitoring.

Discussion

The neonatal lupus syndrome (NLS) is a rare disease characterized by an autoimmune process associated with the presence of auto antibodies in maternal fetal circulation against proteins SSA/Ro and SSB/La. Its clinical manifestations include cutaneous, hematologic and hepatobiliary disorders and presence of isolated congenital heart block (ICHB). Recent studies have also demonstrated the existence of a "lupus pattern" of dyslipoproteinemia characterized by very high levels of VLDL cholesterol and triglycerides (TG) and low HDL cholesterol levels. This case approaches NLS lipid abnormalities not explained by use of drugs like corticosteroids, anticonvulsants, antihypertensives, estrogen and/or progesterone, known to impair lipid profile, nor by condi-
tions like uremia, nephrotic syndrome, diabetes mellitus and thyroid diseases. The case supports the hypothesis that lipid abnormalities in patients with lupus may be part of the plethora of events derived from Lupus itself via several immunological and inflammatory changes occurring in these patients. Recent discoveries of the involvement of cytokines such as IL-1 and especially TNF inducing down-regulation of LPL activity (an essential enzyme whose low activity results in the accumulation of chylomicrons and VLDL, leading to high triglyceride levels and low HDL levels) demonstrate the accomplishments in the disease study.

In 2007, Sawant et al.\(^4\) reported a case of NLS presenting with appearance of typical lesions of NLE at classical sites. Recently, in November 2009, del Boz et al.\(^5\) described a case of NLE and cutis marmorata telangiectatica congenita-like lesions. More recently, in January 2010, Lynn Cheng et al.\(^6\) reported a case of congenital lupus erythematosus presenting at birth with widespread erosions, pancytopenia, and subsequent hepatobiliary disease.

Our case is unique in the appearance of a very specific finding that is a periorbital “owl-eye” rash (Fig. 2) associated with an intriguing systemic finding: high triglyceride levels in a patient with NLE.

Much of the pathogenesis of SLE remains unclear, and the case presented may shed some light on the understanding of lipid abnormalities in lupus.