

# Linear Nevus Sebaceous Syndrome in a Neonate Conceived by Intracytoplasmic Sperm Injection

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## ABSTRACT

Linear nevus sebaceous syndrome is a multisystem disorder including nevus sebaceous, which is the hallmark of this syndrome, together with central nervous system, ocular and skeletal anomalies. We report a case of extensive skin lesions, CNS and eye anomalies in a full term infant who was conceived by intracytoplasmic sperm injection.

**Key words:** Epidermal nevus syndrome, intracytoplasmic sperm injection, Jadassohn nevus phakomatosis, linear nevus sebaceous syndrome, organoid nevus phakomatosis

## INTRODUCTION

In 1895 Jadassohn was the first to describe congenital cutaneous lesions in a linear distribution, for which he coined the term of 'Naevi sebacei'.<sup>[1]</sup> Epidermal nevus syndrome (ENS) has been used to describe the association of an epidermal nevus with systemic features (potentially affecting multiple organs including neurologic, ocular, skeletal, and in rare cases, cardiac and renal anomalies).<sup>[1]</sup>

We report a case of epidermal nevus syndrome with extensive skin lesions, CNS and eye anomalies in a full term infant who was conceived by intracytoplasmic sperm injection (ICSI).

## CASE REPORT

Full-term male neonate, conceived by ICSI, was born to a 21-year-old mother (gravida 1, para 1) at 39 weeks of gestation by caesarean delivery and resuscitation was assigned as uneventful. A prenatal sonogram at 32 weeks gestation revealed polyhydramnios as well as a well-defined hyperechoic mass 4.5×3 cm in the left lateral side of the face and neck with central cystic area.

The initial physical examination showed normal growth parameters with an evident cutaneous abnormality of the face consisted of linear, well-demarcated, raised, yellowish, fleshy and hairless plaques with velvety surface located on the left side of the forehead, the parietal region of the scalp, the face, chin, lower lip, and left ear with a smaller lesion on right side of face [Figure 1]. A similar, less well-formed lesion was present over the midline of the back at the lumbar and the sacral regions and scrotum. Also, a black nodule was noted over the medial canthus of the right eye. There were no other cutaneous manifestations, such as

cafe-au-lait spots or hypo- or hyper-pigmentation lesions. The infant appeared healthy otherwise with no apparent neurologic deficit on physical examination.

Magnetic resonance imaging (MRI) of his brain revealed an extra-axial CSF signal intensity lesion in the left side of the posterior fossa and the cistern magna suggesting arachnoid cyst with mild left cerebellar hypoplasia (Dandy Walker variant) [Figure 2]. Ophthalmologic examination showed bilateral posterior polar cataract. Abdominal sonogram, Doppler of renal vessels, bone survey and echocardiography revealed normal results. Also, serum calcium, phosphorus and urinary calcium were normal.

Histopathology showed thick hyperplastic epidermis with underlying large immature sebaceous lobules and free cut margins with no evidence of cellular atypia.

During hospitalization of the patient, no seizures occurred and no medication was prescribed. A chromosome study of peripheral blood showed a normal male karyotype.

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**Figure 1:** Well-demarcated, raised, velvety plaques on left side of face and ear with other linear ones in midline of face and chin

Written informed consent was obtained from the parents of our patient for publication of this case report and any accompanying images. A copy of the written consent signed by the father of our patient is available for review by the Editor-in-Chief of this journal.

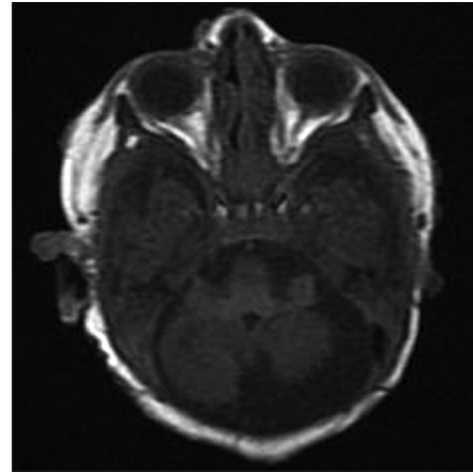
## DISCUSSION

Linear nevus sebaceous syndrome (LNSS) includes sebaceous nevus and defects of the brain, eyes, or bones. Estimated incidence of epidermal nevi is about 1-3 per 1000 live births, with no sexual predilection.<sup>[1]</sup> One third of the patients with epidermal nevi have the nevus sebaceous type.<sup>[2]</sup>

Nevus sebaceous, which is a hallmark of LNSS, is a hamartoma of the epidermis, hair follicles and sebaceous and apocrine glands.<sup>[1,3]</sup> It usually presents at birth as a yellow-orange to pink, finely papillomatous, alopecic plaque that is often oval or linear.<sup>[4]</sup> The lesions vary in size from a few millimeters to several centimeters in length, but unusually large and exophytic lesions had been reported.<sup>[5]</sup> There is a predilection for the scalp and the face, but they may also occur on the neck and trunk.<sup>[3]</sup> During puberty, they have a tendency to become more raised, verrucous, and greasy, at least in part because of androgen stimulation of sebaceous glands.<sup>[6]</sup> The development of secondary malignant neoplasms is rare.<sup>[7]</sup>

The most frequent extra-cutaneous manifestation of nevus sebaceous syndrome is CNS involvement, including seizures, developmental delay, and structural brain abnormalities. The structural brain abnormalities may include hemimegalencephaly, cerebral heterotopias, agenesis of corpus callosum, and dysplasia of brain vessels or Dandy Walker malformation.<sup>[2,6]</sup>

Ocular abnormalities occur in about half of patients with LNSS. Indeed, it has been suggested that the syndrome



**Figure 2:** MRI brain T1-weighted with axial plane showed extra-axial CSF signal intensity lesion in the left side of the posterior fossa and the cistern magna suggesting arachnoid cyst with mild left cerebellar hypoplasia (Dandy Walker variant)

could be considered an oculo-neuro-cutaneous syndrome. The ocular lesions include choristoma of conjunctiva, limbal dermoids, colobomas of the eyelid, iris, and retina, cataracts, strabismus, ptosis, microphthalmos, macrophthalmos, absence of the ganglion cell layer, nystagmus and optic nerve hypoplasia.<sup>[1]</sup> Our patient has extensive nevus sebaceous with a linear configuration that involved the left side of the forehead, scalp, face, back and genital area, left cerebellar hypoplasia (Dandy Walker variant), and bilateral posterior polar cataract. These findings are consistent with the diagnosis of LNSS and further support the possibility of this syndrome to be part of oculo-neuro-cutaneous syndromes.

A range of musculoskeletal, cardiovascular, and urogenital manifestations have been described with LNSS.<sup>[1]</sup>

Nearly all cases are sporadic, but occasionally, nevus sebaceous may affect several members of a family. Paradominant inheritance has been proposed to explain this paradox. When post fertilization loss of heterozygosity occurs in a somatic cell, a homozygous or hemizygous cell clone would originate and give rise to a mosaic patch. It has been suggested that the earlier the mutation event occurs in embryogenesis, the more deleterious consequences will be manifesting as nevus sebaceous syndrome.<sup>[6]</sup> Also, HPV infection of a pluripotent stem cell at an early stage of embryogenesis could play a role in the pathogenesis of nevus sebaceous syndrome.<sup>[6]</sup>

Hansen and colleagues found that infants conceived with use of intracytoplasmic sperm injection or *in vitro* fertilization have twice as high a risk of a major birth defect as naturally conceived infants.<sup>[8]</sup> The ICSI procedure itself or using sperm from sub-fertile men may increase

the potential to transmit genetic or chromosomal, either sex or autosomal, aberrations to offspring created from ICSI technology.<sup>[9]</sup> Reported malformations with ICSI include; cardiovascular, urogenital, chromosomal, and musculoskeletal defects.<sup>[9]</sup> Although skin nevi were previously reported with ICSI,<sup>[10]</sup> to the best of our knowledge this is the first case to describe the association between LNSS and ICSI.

Although, surgical excision is the treatment of choice, a multidisciplinary approach is mandatory. The pediatrician, neurologist, ophthalmologist, dermatologist, and geneticist should give advice.<sup>[6]</sup>

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