

## Nevus sebaceous on the scalp: answer

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### Answers

1. Nevus sebaceous (NS).
2. The lesion should be examined by a dermatologist to confirm the diagnosis of NS. Routine neurological and ophthalmological evaluations should be performed by a pediatrician/neonatologist to exclude other lesions. A healthy child with a solitary NS can be followed up in regular consultations until adulthood. The risk of malignant transformation during childhood is insignificant.
3. When the child presents an NS associated with other organ system disorders, an epidermal nevus syndrome (ENS) should be suspected. The

most common organ involvement in all types of ENS is the nervous system (mental retardation and seizures are the most common diagnoses). In such cases, a neuropsychiatrist should be involved.

Ocular findings of coloboma or choristomas, which are easily diagnosed, should prompt an extensive neurologic examination because their presence increases the risk of neurologic disorders. These children should also be evaluated by an ophthalmologist. Hypophosphatemic rickets and muscle weakness should increase suspicion of two specific ENS: cutaneous skeletal hypophosphatemia and phacomatosis pigmentokeratolica. In these cases, the child should be evaluated by a metabolism specialist. Changes in the shape or color of NS should raise suspicions of NS transformation. In such cases, the child should be observed by a dermatologist.

## Introduction

NS, the most common adnexal malformation during pediatric age, is observed in 0.3% of newborns [1]. It is typically present at birth, preferentially on the face or scalp and appears as a salmon, yellow-orange or tan, hairless plaque [1, 2]. Its diagnosis is clinical, and a histological study should be reserved for doubtful lesions. In most cases, NS is a benign solitary lesion. Rarely,

it can be the cutaneous manifestation of an ENS, which typically involves the cerebral, ocular and/or skeletal systems [3]. It is known that both ENS and NS have the same genetic cause: a postzygotic mutation in intracellular RAS pathway proteins [4]. Other factors beyond specific gene defects determine the phenotype (e.g., timing of mutation during fetal development and the levels of the defected gene expressed within specific cell types) [2, 4]. The potential malignant transformation of NS is insignificant during childhood and low during puberty [5, 6]. The most frequent malignant transformation is basal cell carcinoma, the probability of which at an age younger than 18 years is 0.1% [3, 5].

## Clinical course

We describe the case of a 1-month-old, full-term male infant with an NS (localized, well-delimited, waxy alopecic area on the scalp) (**Fig. 1**) that has been present since birth. He was re-evaluated 3 months after the first observation; apart from the lesion on his scalp, the patient was physically normal. Neurological and ophthalmologic evaluations were unremarkable, and the infant exhibited normal neuromotor development. The child is now 1 year old, the NS keeps the same characteristics and dimensions, and the general growth and development are within the normality.



**Figure 1.** A well-delimited plate-like, waxy, skin-colored lesion free of hair follicles and localized on the scalp.

## Discussion

A solitary NS lesion in a healthy infant is just a benign hamartoma of the skin. The natural evolution of the NS consists of a proportional enlargement of the lesion with age. At puberty, via the action of hormones, the lesion typically becomes more thick, yellowish and cerebriform [3]. In some cases, secondary tumors arise within the NS. The most common benign transformation is trichoblastoma and syringocystadenoma papilliferum [5, 6]. The most frequent malignant transformation is basal cell carcinoma; in adulthood the rate of basal cell carcinoma can approach 2% [5, 6]. In childhood, the risk of malignant neoplasm is very low – the rate of malignancy in some series is zero [5, 6]. Considering this fact, the surgical excision of an NS is not potentially mandatory until adulthood. Factors such as its size, location and the patient's desire to take it off will help with the decision on what to do.

In rare cases, an ENS (i.e., a cutaneous skeletal hypophosphatemia or phacomatosis pigmento-keratolica) may be present. Such a condition is typically associated with more extensive NS, with a linear shape that is often distributed along Blaschko lines (a classic pattern of cutaneous mosaicism – two or more genetically different populations of cells existing side by side – occurring in a wide variety of congenital and acquired skin conditions,

they differ from dermatomes, and appear as single or multiple lines, whorls/swirls and wave-like shapes in the skin) [3]. Even so, recent studies have shown that there is not a linear association between the size of the lesion and the presence of neurologic abnormalities [2]. Primary neurologic abnormalities include mental retardation and seizures [2, 3]. The most common ocular findings include colobomas and choristomas [2, 3].

## Declaration of interest

The Authors declare that there are no conflicts of interest and that they did not receive any financial support.

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