

A Case of Basaloid Degeneration of Nevus Sebaceous during Childhood: Should Nevus Sebaceous Be Excised or Followed Up?

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Facial Plast Surg 2016;32:576–577.

Nevus sebaceous of Jadassohn (NSJ) is a complex hamartoma located on the face and scalp. Its incidence ranges between 0.05 and 1%. At birth, it is a well-circumscribed solitary yellowish, verrucous patch. At puberty, it becomes larger, more elevated, and cerebriform. Numerous secondary neoplasms may occur in NSJ with an estimated rate from 10 to 30% increasing with age. The most common evolution is toward benign or malignant basaloid neoplasms. However, malignant basaloid degeneration is uncommon before puberty with only 10 documented cases of transformation into basal cell carcinoma (BCC).

An 11-year-old girl followed up since birth for an NSJ extended across the whole nasal dorsum developed a 5 × 4 mm nodule in a few months, in the middle part of the lesion, without telangiectasia or ulceration. A biopsy was made and histological analysis was suggestive of degeneration into BCC.

The lesion was totally excised with a 5-mm surgical margin. Definitive histological examination showed focal basaloid hyperplasia with no tumor residue, in a typical NSJ. Clinical follow-up showed a satisfying aspect of the scar 6 months after surgery (→ Figs. 1–3).

Historically, malignant transformation into BCC arose in 6.8 to 17.2% of cases of NSJ. It was designated as a risk factor



Fig. 1 Preoperative image. Unsightly nevus sebaceous of Jadassohn extended across the whole nasal dorsum.

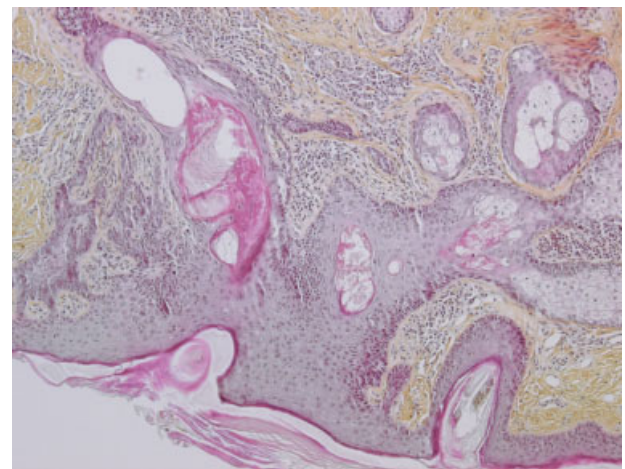


Fig. 2 Definitive histology showed focal basaloid hyperplasia with abundant fibroblastic stroma, no tumoral residue (hematoxylin-eosin stain; ×10).

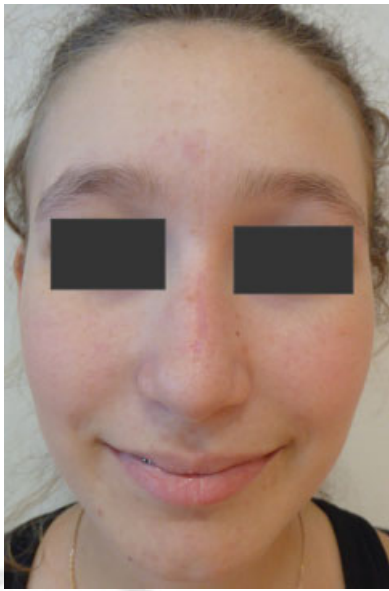


Fig. 3 Postoperative image. Satisfying aspect of the scar 6 months after surgery.

for developing malignancy and was treated by prophylactic surgery.

In 1993, Ackerman defined the word trichoblastoma (TB) as a generic term for all adnexal neoplasms of follicular germinative cells. After this, almost all basaloid neoplasms developing in NSJ were considered as TBs and not BCC. In 2000, Kaddu confirmed this definition in a larger study and refined microscopic criteria to better characterize basaloid neoplasms in NSJ. On the basis of these criteria, most neoplasms arising in NSJ are benign, and BCC occurs in only 0.8 to 1.1% of the cases. In studies with a large patient population no malignant transformation was observed during childhood. In

light of these data, clinical follow-up was proposed for the management of NSJ before puberty.

Nevertheless, some basaloid neoplasms and especially nodular lesions occurring in NSJ combine features of TB and of BCC and pose a real problem for anatomical pathologists. Sellheyer in 2013 demonstrated that immunohistochemistry staining using the novel stem cell marker pleckstrin homology-like domain, family A, member 1 (PHLDA1) to differentiate between TB and BCC challenges the reliability of histological macro- or microscopic criteria.

Despite this, good practice recommends clinical follow-up rather than systematic prophylactic excision. Immunohistochemical staining using PHLDA1 calls into question the reliability of conventional histopathologic criteria. At present, any anatomopathologic diagnosis can be sure, but still influences decisions between excision and follow-up. However, for cosmetic indications most facial NSJ are removed during childhood or puberty. Full-thickness excision, or possibly Mohs micrographic surgery, is the best way to treat NSJ.

Surgical excision presents both a cosmetic benefit because of the unsightly aspect of NSJ and a medical interest, avoiding the risk of degeneration into BCC or histopathologic misdiagnosis. We therefore think that surgical removal of NSJ must be made as often as possible.

Informed Patient's Consent

Informed consent was obtained from all individual participants included in the study. Additional informed consent was obtained from all individual participants for whom identifying information is included in this article.

Conflict of Interest

None declared.