

Microcystic Adnexal Carcinoma Originating in a Nevus Sebaceous: A Case Report of a 16-year-old Boy

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Nevus sebaceous (NS) is a congenital hamartoma, comprised of abnormal epidermal and dermal components including both the adnexal (follicular pilosebaceous and glandular) and epithelial parts in different proportions (1). NS affects mainly the scalp and the estimated prevalence in newborn infants is 0.3% (2, 3). Normally the hamartoma remains benign. However, NS transforms into different types of neoplastic tumors in about 10–20% of cases (4). The majority of these secondary neoplasms is benign and occurs in adults older than 40 years of age (3). The rate of malignant transformation of NS has been reported to be up to 3% (3, 5) and the most common tumor is basal cell carcinoma (BCC) (3, 5). Malignant transformation in children and adolescents is rare. In an extensive literature review by Moody et al. (4), they reviewed 4,923 cases of NS and found that only 24 of 204 cases of malignancies such as BCC, squamous cell carcinoma and keratoacanthoma had occurred in individuals younger than 18 years old. Microcystic adnexal carcinoma (MAC) is a rare cutaneous neoplasm of the skin adnexa, which very rarely originates within a NS, although it has been reported in adults (5–8). To the best of our knowledge MAC originating in a NS in a child or an adolescent has not previously been reported.

CASE REPORT

A 16-year-old boy, born with a yellow-grey NS on the parietal area of the scalp, attended the primary health care center in May 2016 due to increasing itching in the lesion. Apart from the itching, which commenced 12 months prior to the visit he had no other symptoms, was generally healthy and taking no medications. There was no history of skin cancer in the family. The boy had skin type 1 as graded by Fitzpatrick. He had not been exposed to radiation in the head and neck region. Clinical and dermatoscopic examination revealed a 20 × 35 mm NS of classical appearance and the lesion was thus deemed to be benign. However, the patient noticed that the lesion was growing slightly and that the itching intensified. He therefore re-attended 6 months later and was examined again by a general practitioner who on ocular inspection did not find any abnormal changes and decided that excision of the lesion was not necessary. Because of the symptoms, however, the patient was referred to the Department of Dermatology at the Linköping University Hospital for a second opinion. The dermatologist found no abnormal clinical or dermatoscopic changes within the lesion (Fig. 1). Due to continued itching, however, the lesion was excised two months later. The histopathological examination showed a NS with an area containing keratinous cysts, solid nests and strands of basaloid cells and duct-like structures in a sclerotic stroma, consistent with MAC. Focal perineural invasion was present.



Fig. 1. The lesion located on the scalp 2 months before excision.

Immunohistochemistry showed low proliferative activity (Ki67, Dako Denmark, dilution 1:200) and sparse nuclear p53 expression with low to moderate intensity (Dako, dilution 1:200), consistent with MAC (9). The lesion was very close to the side margin (Fig. 2). The initial surgical excision was done with 2 mm margins, and hence re-excision was needed. The wide local excision (WLE) was done with approximately 2 cm margin to ensure cancer clearance. No residual NS or MAC was present in the WLE specimen. Cervical lymph nodes were not palpable. The patient is doing well, with no signs of recurrence 2 years after surgery. Magnetic resonance imaging showed no remaining lesion.

DISCUSSION

MAC is a rare locally aggressive cancer of the skin adnexa. It was first described by Goldstein et al. in 1982 to originate from a pluripotent adnexal keratinocyte (10). According to a review article by Gordon et al. (11) and a study by Yu et al. (12) MAC occurs most often in middle-aged to older Caucasian individuals and has an incidence of between 1.6 and 6.5 per 10,000,000 individuals (12). Only 4 cases of MAC arising from NS have been reported and all were identified in adults aged 75, 26, 62 and 65 years, respectively (5–8). To our knowledge this is the first reported case of MAC originating in a NS in an adolescent.

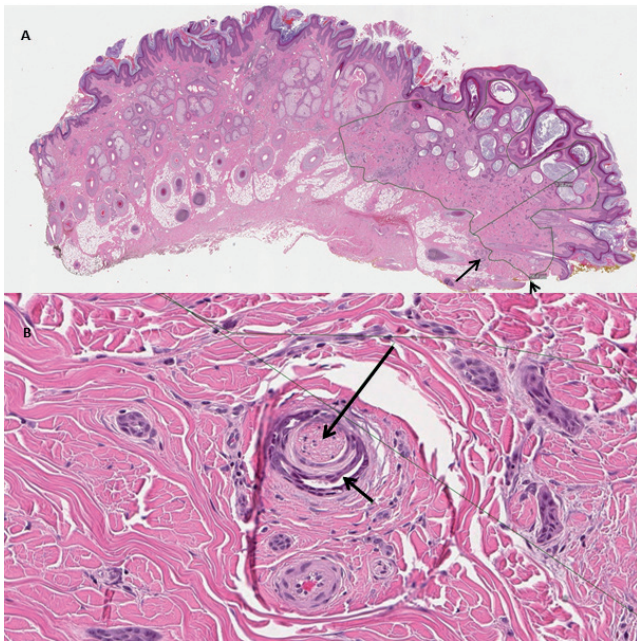


Fig. 2. A. Excised nevus sebaceous (NS) with microcystic adnexal carcinoma (MAC). The epidermis is papillomatous, sebaceous glands are multilobular, enlarged and focal apocrine metaplasia, typical for NS, is indicated by long arrow. The MAC area, encircled with the line, shows characteristic keratinous cysts in the superficial part and small islands and strands of basaloid cells and duct-like structures in sclerotic stroma in the deeper part. The short arrow points to the MAC 0.06 mm from the side margin (Hematoxylin-eosin stain; original magnification: $\times 2.4$). B. Perineural invasion. The long arrow points to the deep dermal nerve, the short arrow points to the duct-like structure in the surrounding MAC (Hematoxylin-eosin stain; original magnification: $\times 40$).

MAC commonly affects the head and neck region and often has a clinically benign appearance (11). The surface is usually smooth and non-ulcerated in contrast to many other malignant tumors (13). This may explain why the tumor was deemed benign in our patient and that the excision was delayed. Furthermore, Rosen et al. found no obvious clinical features or gross morphological changes in 5 children with BCC within NS (14). Together with other studies with similar findings they therefore concluded that an NS lesion cannot be deemed to be a truly benign lesion based on its uniform appearance and young age.

It is not clear when MAC arose in our patient. The slow progression and innocuous clinical appearance of MAC can result in a delay of several years from onset to diagnosis (8). MAC is usually asymptomatic but symptoms such as pain, burning, or paresthesia may occur secondary to the perineural invasion, which is commonly seen in MAC (8). The increasing itching in our case might be explained by perineural invasion. A symptom like itching in an otherwise benign looking tumor could have alerted the clinician to consider an excision.

This case report shows the diverse neoplastic potential of NS and the ability of this hamartoma to transform into a malignant tumor. There is an ongoing debate whether it is advisable to remove NS in childhood. To avoid a less complicated procedure Moody et al. propose a prophylactic excision in all children before the expansion of NS occurs and when the lesion is still small (4). However, most NS remain benign and excision may not be necessary in cases where the NS is small and not otherwise disturbing. In this case an excision when the boy was prepubertal would have been beneficial since the excision once MAC was diagnosed had to be done with wide margins requiring skin transplantation.

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