Multifocal basal cell carcinoma arising within a linear epidermal nevus

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ABSTRACT

Epidermal nevi are usually benign lesions with a lifelong course if left untreated. In rare cases development of basal cell carcinoma in such nevi has been documented. We describe a 32-year-old man with a multifocal malignant transformation within a congenital epidermal nevus.

Key words: Basal cell carcinoma, epidermal nevus, linear nevus

INTRODUCTION

Linear epidermal nevi are cutaneous hamartomas of ectodermal origin usually present at birth with the estimated incidence of 1:1000 live births.^[1] Manifestation of epidermal nevi later in life is also possible. They vary in size from quite small to very extensive papillomatous or hyperkeratotic skin lesions of segmental or systematized pattern. The major concern for most patients is cosmetic disfigurement of various degrees, since epidermal nevi are usually asymptomatic with the exception of the inflammatory linear verrucous epidermal nevus type. Laser therapy is a preferred method of treatment of epidermal nevi with good long term results, though recurrence is not infrequent.^[1]



A 32-year-old man presented with a linear skin lesion of segmental distribution over his left arm,



Figure 1: Basal cell carcinoma (H and E, ×100)

was not of the common hyperkeratotic pigmented type, but resembled a linear organoid nevus by its softer texture and light color. However, the patient refused additional biopsy and further workup to determine histologic subtype of the nevus. The lesion had been present since birth and was asymptomatic until approximately 1 year ago, when a slowly enlarging ulcer appeared on the upper arm. The ulcer was biopsied and histological investigation revealed basal cell carcinoma (BCC) with deep invasion into the dermis [Figure 1]. The patient was offered

shoulder and upper back. The epidermal nevus



Figure 2: Ulcerated basal cell carcinoma on the arm

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Figure 3: Several foci of basal cell carcinoma within the epidermal nevus on the back

appropriate therapy by an oncologist, but he sought the second opinion and was referred to the Dermatology Department. On clinical examination, it was noted that besides the large crusted ulcer of confirmed BCC on the arm [Figure 2], there were several other loci of evolving tumors on the patient's back [Figures 3 and 4]. Dermoscopy revealed diagnostic features of a BCC. The patient declared no other problems with his health and was advised to follow treatment recommendation of the consultant oncologist.

DISCUSSION

The course of epidermal nevi even without treatment is usually lifelong and benign. It is important to exclude other organ involvement and to rule out epidermal nevus syndrome. In epidermal nevus syndrome, nevus formation of various types may be associated with neurologic, ocular, skeletal, or other systemic abnormalities.

It has been recognized that epidermal nevi, especially of the organoid type, are associated with an increased risk of BCC and occasionally, squamous cell carcinoma. The exact incidence of this tumor type in epidermal nevi is unknown, though some authors state that the risk of developing BCC in a nevus sebaceous, another type of epidermal nevus, is 6.5–50%.^[2] Indeed, most of reported cases of BCC were in concurrence with sebaceous nevi, predominantly located in the head and neck area in postpubertal patients.^[3,4] According to Raid *et al.*, only 13 cases of squamous cell carcinoma arising from verrucous epidermal nevi have been reported so far in the literature, which may not reflect its true incidence.^[5] However, development of BCC in linear verrucous epidermal nevi is well documented.^[2,6] The factors that predispose



Figure 4: Close-up view

to focal malignant transformation are unknown. Clonality was demonstrated in BCC cells arising in the setting of epidermal nevus by means of molecular analysis.^[7] We believe that the present case is of interest due to multifocal malignant transformation.

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