



fig. 2 Characteristic 'double lip' appearance.

Upper lip and eyelid oedema usually appear at about the same time, and in more than 80% of cases the condition manifests itself before the age of 20.<sup>6</sup> Even though euthyroid or non-toxic goiter is considered an important clinical finding of the syndrome, it is thought to be present in less than 10% of cases and may appear many years after eyelid and lip involvement.<sup>2,7,8</sup>

Eventually the oedema results in eyelid laxity, which explains the name of the condition, and occasionally causes cosmetic and visual impairment.<sup>5</sup> Progression of the disease can result in prolapse of the periorbital fat and lacrimal glands, as well as blepharoptosis. In some cases the alteration to the lips can become a cosmetic handicap.<sup>3,9</sup> The treatment of choice is simple or extensive blepharoplasty for correction of the blepharochalasis and transverse elliptical excision of both labial ends to correct the duplication of the labial folds in the severe forms of the disease.<sup>3,9,10</sup>

Recognition of the characteristic features of this syndrome will prevent several misdiagnoses, including hereditary angioedema, early dermatochalasis, acquired cutis laxa and variants of granulomatous cheilitis.<sup>4</sup> Equally, early diagnosis of this unusual entity will prevent unnecessary tests and delays, and allow discussion of the condition with patients and relatives, and prompt scheduling of the appropriate surgical treatment if required.

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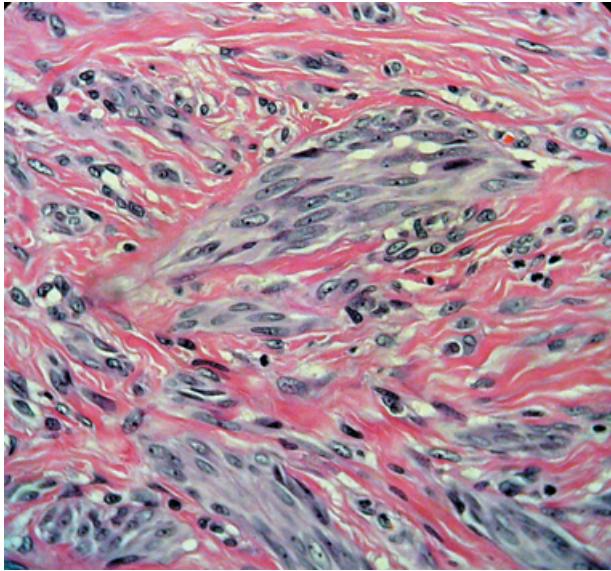
## Epithelioid combined nevus in a Caucasian boy with no evidence of Carney complex

### Editor

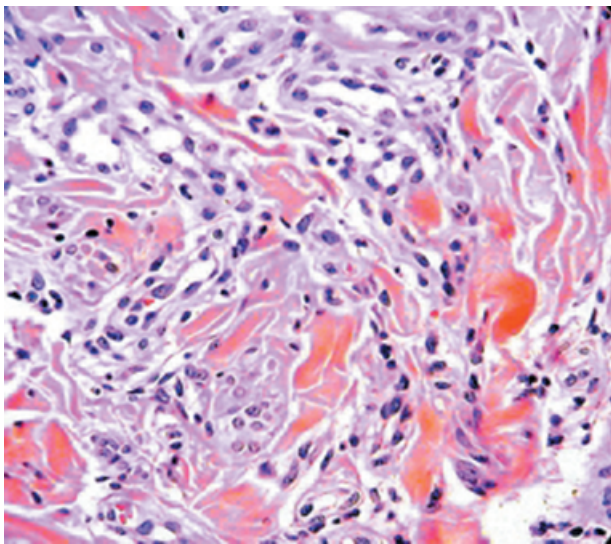
We present the case of a 13-year-old boy, who attended our unit to have a slightly brown papule on his right knee evaluated. The patient was uncertain how long the lesion had been present. His parents were worried about a single bleeding episode that had occurred during the previous 2 weeks after a minor trauma.

Physical examination showed a 0.6-cm, darkly pigmented papule on his right knee. Dermoscopically the lesion revealed a well-circumscribed papule with regular edges, symmetrical pattern, and dark brown colour with focal silver-grey patches. No signs of malignancy were observed.

The lesion was excised, with differential diagnoses of dermatofibroma vs. Spitz nevus. Microscopically, a dermal lesion could be observed displaying two types



**fig. 1** Large, spindle-shaped epithelioid cells embedded in a collagenous stroma.



**fig. 2** Vascular ectasia in the deep portion of the dermis.

of melanocytes: one intensely pigmented, globular and fusiform, and the other lightly pigmented, and polygonal or spindle shaped (fig. 1). Rare mitotic figures were seen. Vascular ectasia was also seen in the deep portion without neurotropism (fig. 2). Therefore, Blitz nevus was diagnosed.

Cardiological, endocrinological and neurological examinations were carried out in order to rule out Carney complex, including cranial magnetic resonance imaging, electroencephalogram, echocardiogram and sexual, adrenal, thyroid and growth hormone profiles. No abnormalities were found.

Groben *et al.*<sup>1</sup> classified epithelioid combined nevi into three well-characterized phenotypes: (a) the classic or Carney complex pattern, (b) those that showed overlap with deep penetrating nevus, and (c) those that have many dermal Spitz-nevus features, blue + Spitz nevus, also called Blitz nevus, as in the present case report.

Carney complex is an autosomal dominant, clinically heterogeneous syndrome of multiple neoplasia. Cardiac myxomas, spotty pigmentation (including lentigines, ephelids and blue nevi), endocrine over-activity, and psammomatous melanotic schwannomas define this syndrome.<sup>2</sup> Epithelioid blue nevus had been considered important because of its strong association with Carney complex, and the risk of developing cardiac myxoma. Nevertheless, many authors have communicated sporadic cases of epithelioid combined nevi with no evidence of Carney complex, including in paediatric patients.<sup>3</sup> We wonder whether this kind of nevi is under-diagnosed or misdiagnosed. Is it obligatory to dismiss Carney complex whenever we diagnose this entity? Are there any dermo-pathological features that can help us to avoid excessive complementary radiological and laboratory examinations? No consensus responses have been found in the medical literature. In our opinion, follow-up of the patient and the clinical course are, once again, essential to answer all these questions.<sup>4</sup>

Clinical diagnoses included malignant blue nevus, atypical nevus, melanoma, congenital nevus, and dermatofibroma. Recognition of amelanotic epithelioid combined nevus is important because the lack of pigmentation may result in clinical and pathological diagnostic difficulties.

Our pathological case shows the typical features of blitz nevus: epithelioid and spindle cells, marked fibrosis and prominent vascular ectasia on the deep portion of the dermis. Therefore, histopathological differential diagnoses include those of benign and malignant pigmented dermal melanocytic proliferations composed of epithelioid and/or spindle cells, particularly with desmoplastic and collagenous nevi.<sup>5</sup>

In summary, epithelioid combined nevus is an entirely benign melanocytic lesion, and simple excision is curative. Pathologists and dermatologists should consider this entity, despite its unusual and uncommon presentation, in the differential diagnosis of non-pigmented melanocytic skin lesions in adult and paediatric patients.

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## West Nile virus rash on the palms and soles of the feet

### Editor

We read with great interest the case study and review of West Nile virus exanthem by Anderson *et al*.<sup>1</sup> In their review, case 3 was a 66-year-old man with no significant past medical history, who was admitted to hospital in July 2003 with a 2-day history of fever, chills, malaise, and a non-blanching erythematous, macular rash on his extremities, specifically the palms and soles, after a recent visit to a forested area in Dallas, Texas.

In the same publication, Anderson *et al*. displayed four photographs of the cutaneous manifestations of West Nile virus infection. We have additional photographs that show this rash on the palms and on the soles of the feet. There were numerous 1–3-mm erythematous macules and petechiae on the sole of the right foot (fig. 1). We believe the picture in this letter represents the first published photograph illustrating a West Nile virus rash on the sole of the foot.

West Nile virus, a member of the Flaviviridae family, first appeared in North America in 1999.<sup>2</sup> Since then the virus has spread steadily westward across the country.<sup>3</sup> As of March 2004, there were 9377 human cases of West Nile virus infection and 244 deaths for the year 2003 ([http://www.cdc.gov/ncidod/dvbid/westnile/surv&controlCaseCount03\\_detailed.htm](http://www.cdc.gov/ncidod/dvbid/westnile/surv&controlCaseCount03_detailed.htm)).

About 20 to 50% of patients have a maculopapular or morbilliform rash involving the face, neck, trunk, arms, or legs, which can last for up to a week.<sup>3,4</sup> It is of interest that



**fig. 1** Rash associated with West Nile virus infection. Numerous 1–3-mm erythematous macules and petechiae on the sole of the right foot.

the resolution of this patient's rash coincided with the onset of neurological symptoms. Currently, there is no description of a West Nile virus rash in relation to the onset of neurological symptoms.

This clinical presentation of fever, macular lesions on the palms and soles spreading centripetally, and neurological abnormalities, should also lead to consideration of tick borne-related diseases (i.e. Rocky Mountain spotted fever, Ehrlichiosis), spirochete infections (i.e. secondary syphilis), and possibly other viral exanthems.

We believe physicians should consider West Nile virus infection when evaluating a patient who has a fever, an erythematous macular rash on their palms and soles that spreads centripetally, and neurological abnormalities, especially during late spring through to early autumn, or throughout the year in warm climates.

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