

Desmoplastic Melanocytic Naevus in a Paediatric Patient: Dermoscopic and Histopathological Findings of a Rare Entity: An Overlooked Diagnosis or a Misnomer?

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A 15-year-old female patient with an erythematous nodule in the periocular area for 6 years was referred to our dermatology outpatient clinic. On dermatological examination, a pinkish erythematous nodule on the right eyebrow was revealed (Fig. 1a). Dermoscopic examination showed linear and glomerular vessels arranged focally on the pinkish erythematous background (Fig. 1b). The lesion was totally excised by the Department of Plastic and Reconstructive Surgery. Histopathological examination revealed scattered nevoid cells in the collagenized desmoplastic dermis (Fig. 2a). Nevoid cells

were positive for SOX10 (Fig. 2b) and Melan A (Fig. 2c), whereas staining for HMB45 and p16 was negative (Fig. 2d and e).

What is your diagnosis?

1. Spitz naevus
2. Spitzoid melanoma
3. Desmoplastic melanocytic naevus
4. Intradermal naevus

See next page for answer.

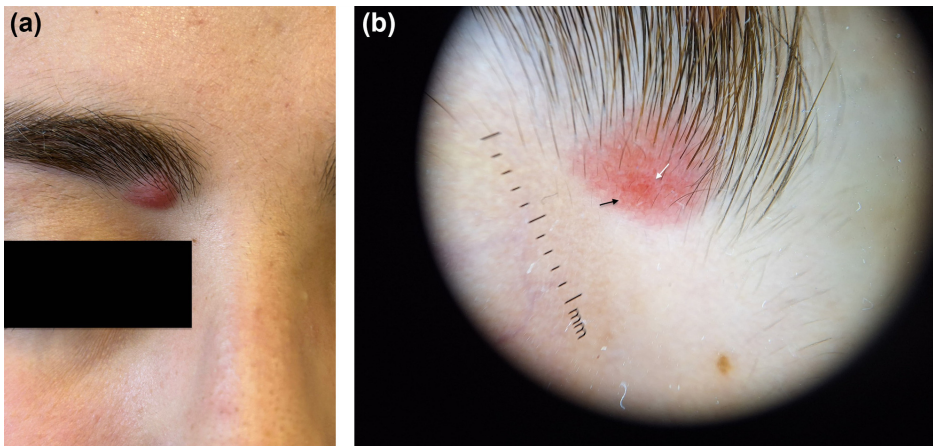


Fig. 1. (a) Pinkish erythematous nodule on the right eyebrow. (b) Focally distributed linear (black arrow) and glomerular vessels (white arrow) on the pinkish erythematous background.

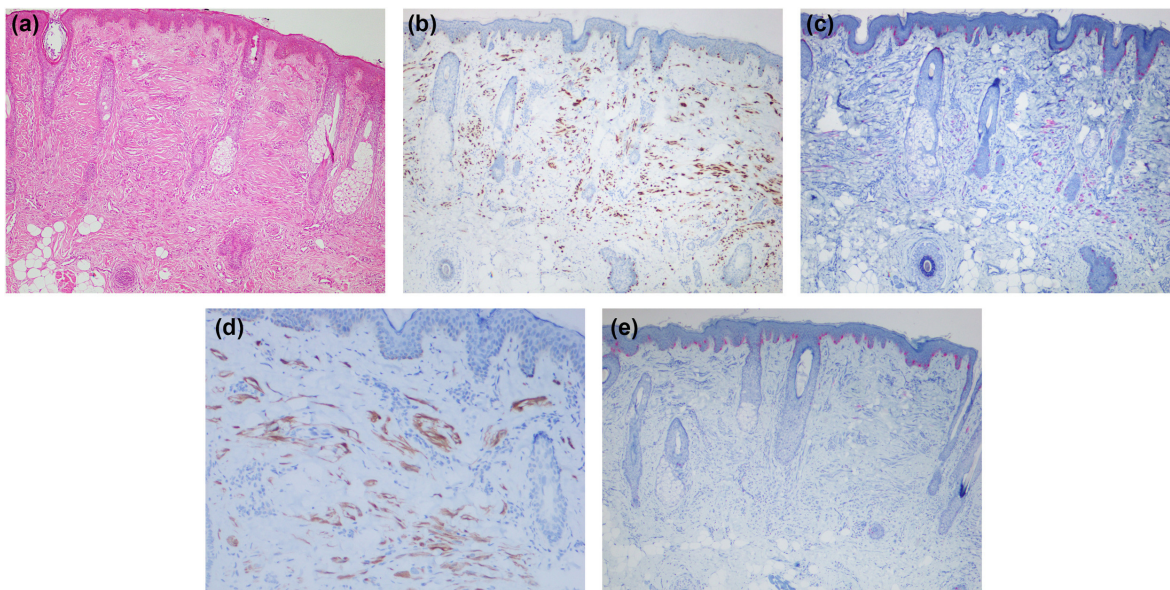


Fig. 2. (a) Nevoid cells scattered in collagenized desmoplastic dermis (H&E, $\times 40$), (b) positive SOX 10 staining ($\times 40$), (c) positive Melan A staining ($\times 40$), (d) p16 negative ($\times 100$) and (e) HMB45 negative ($\times 40$).

ANSWERS TO QUIZ

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Diagnosis: Desmoplastic melanocytic naevus

Based on the clinical and histopathological findings, desmoplastic naevus (DN) was diagnosed. DN is an erythematous, pink-coloured or slightly pigmented papule or nodule and is histopathologically characterized by spindle-shaped cells within a fibrotic stroma (1–3). In 1975, DN was first described and considered as a variant of Spitz naevus (SN) (1, 2). However, Barr *et al.* suggested that DN is not a variant of SN and is a unique histopathological entity (3). In 1992, this idea was also supported by Mackie and Doherty (4).

Scientific reports on DN are generally based on histopathological findings. However, there is only 1 case series of 3 patients with DN in which dermoscopic features are described (5). Herein, we report a 15-year-old girl with DN, and our case represents the second report describing clinical, dermoscopic and histopathological findings.

Histopathologically, DN has an intradermal growth pattern characterized by the presence of compact sclerotic or desmoplastic dermal collagen, composed of large spindle and/or epithelioid cells in a fibrotic stroma (2, 4, 5). Mackie and Doherty (4) also described absent or sparse melanin pigment in DN in their report (4). In our case, although Melan A and SOX 10 were positive, we did not observe any pigment structure dermoscopically or clinically.

There are benign and malignant cutaneous lesions, including desmoplastic Spitz naevus (DSN), DN, dermatofibroma, sclerotic blue naevus and desmoplastic melanoma, that are characterized by dermal proliferation with a desmoplastic component (5). DSN and DN may present similar characteristics such as patient age, location and clinical manifestations. Although it has been suggested that DSN can be easily differentiated from DN based on dermoscopic findings, as we show in our case, the dermoscopic features of paediatric DN may be mistaken for those of DSN. Dermoscopically, SN may present with homogenous pink colour, dotted, hairpin, linear or

comma-like vessels and reticular depigmentation (2, 3, 5, 6). In our case, there were linear irregular and glomerular vessels similar to the vascular features in SN. However, in our case, the vascular structures were focally distributed, whereas in SN, it can be suggested that these vessel structures would be arranged regularly throughout the lesion.

Ferrara *et al.* reported similar dermoscopic findings in all their DN cases (5). However, in their report, all patients were female, and none were paediatric cases (aged 56, 38 and 22 years). Our case is the first reported paediatric case with dermoscopic findings (5). Furthermore, we showed polymorphic vessels, which can be mistaken for those seen in other malignant cutaneous lesions, such as melanoma, and also for the features of SN. In addition, although Ferrara *et al.* (5) reported a light brown network on a pinkish erythematous background in all their cases, we did not observe any pigment network in our case.

DN is a rare condition, and it has been suggested that DN may represent a variant of SN. We support the suggestion that DN is a separate entity from SN, and it is clear that additional reports are needed to describe the dermoscopic features of this naevus type in both children and adults. Dermatologists should be aware that DN may present with different atypical dermoscopic features in children.

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