



## A solitary nodule of the right cheek of a 25-year-old man

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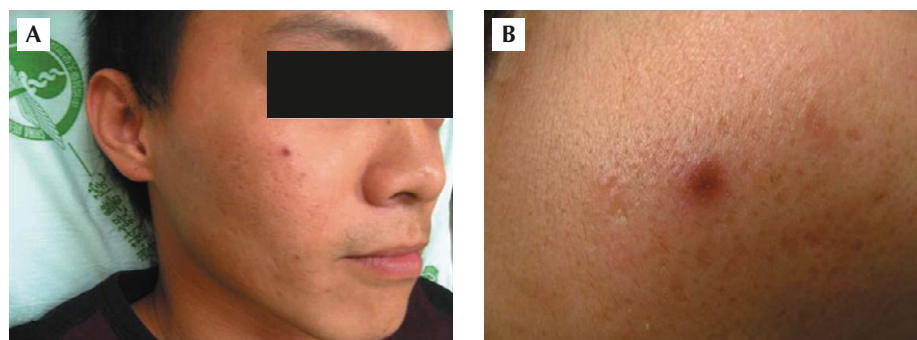
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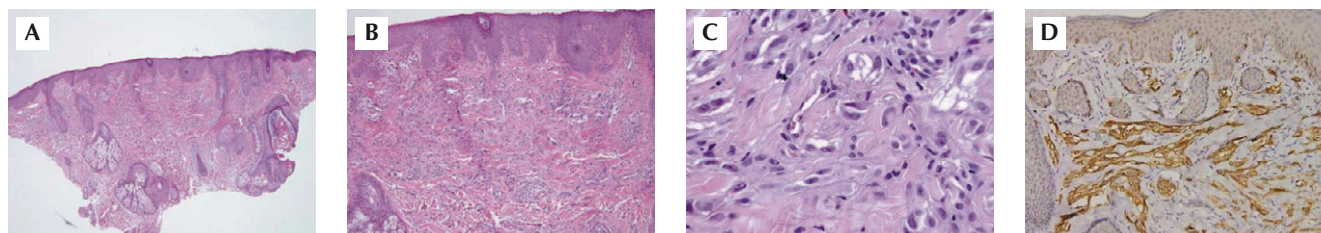
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### Case report

A 25-year-old man presented with a 1-year history of a small nodule on the right cheek. The nodule presented as an ill-defined, reddish lesion, with a diameter of 5 mm (Figure 1). The nodule was excised and subjected to pathological examination, which revealed pleomorphic spindle and epithelioid cells infiltrating dermal collagen bundles. These cells showed discernible downward maturation (Figure 2). Microscopic examination under a high-power field showed nuclear enlargement, pleomorphism, and indistinct mitotic figures. Foci of Kamino bodies and scattered melanin deposition were detected in the papillary dermis. Immunohistochemical analysis showed that the tumor cells had increased cytoplasmic expression of anti-melan-A, HMB-45, and CD68, and that these cells were negative for the expression of CD34 and epithelial membrane antigen (Figure 2). Immunoreactivity to Ki-67 antibody was scored as less than 2% positive cells. Total excision of the lesion was then performed, and no recurrence was noted during follow-ups at our clinic approximately half a year later.



**Figure 1** (A,B) A dome-shaped reddish nodule approximately 0.5 cm in diameter over the right cheek of a 25-year-old male.



**Figure 2** (A) A small, ill-defined lesion confined to the dermis (H&E, 40 $\times$ ). (B) There are multiple pleomorphic spindle and epithelioid cells infiltrating the fibrotic dermal collagen. Note that the size of the cells at the bottom is smaller than that of at the top (maturation) (H&E, 100 $\times$ ). (C) The spindles cells reveal pleomorphism and indistinct mitotic figures (H&E, 400 $\times$ ). (D) Immunohistochemical features: most large epithelioid or spindle cells express anti-melanin A (200 $\times$ ).

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## Diagnosis

Desmoplastic Spitz nevus.

## Discussion

Desmoplastic Spitz nevus is a subtype of Spitz nevus, which is the benign proliferation of large epithelioid melanocytes; this lesion is sometimes mistaken for a melanoma.<sup>1</sup> Desmoplastic Spitz nevus can be distinguished from other types of Spitz nevus on the basis of histopathological analysis. However, all Spitz nevi present in a similar manner, i.e. they occur as solitary lesions, are usually lightly pigmented, most commonly affect individuals younger than 20 years of age, and show a predilection toward the face or lower limbs.<sup>1</sup> In the absence of histological analysis, desmoplastic Spitz nevus is easily misdiagnosed as a dermatofibroma or an angioma.<sup>1</sup> The diameter of desmoplastic Spitz nevus is often less than 10mm, but this is not always the case. The size of the lesion is commonly used as a criterion in the differential diagnosis between desmoplastic Spitz nevus and melanoma. The clinical course of desmoplastic Spitz nevi appears to be rapid growth in the initial period, followed by a period of stable disease. This is an important feature which distinguishes this lesion from melanoma where the diameter of the lesion seldom remains constant. In our case, a young adult male patient presented with the typical clinical signs of a reddish nodule on the face.

Spitz nevi can be classified into several types according to the histopathological features of the lesions. The World Health Organization classification system classifies Spitz nevi into the following three types on the basis of the dermal layer at which they occur, junctional Spitz nevus, compound Spitz nevus, and intradermal Spitz nevus. Desmoplastic Spitz nevus is classified as the intradermal type because nests of melanocytes are often present between thickened collagen bundles in the dermis. Another method of classifying Spitz nevi was proposed by Ferrara et al:<sup>2</sup> classic or desmoplastic Spitz nevus, pigmented Spitz nevus, and atypical Spitz nevus. DSN is characterized by a relatively well-circumscribed lesion confined to the dermis, as confirmed by microscopic examination in a low-power field, with abundant large epithelioid or spindle cells, mature melanocytes in the dermis, marked dermal fibrosis, and rarely, mitoses.<sup>3</sup> Ancillary features such as Kamino bodies, pagetoid spreading of melanocytes, vascularization, or pigmentation may appear in other variants of Spitz nevus and are not necessarily

present in desmoplastic Spitz nevi. The variants of Spitz nevus can be differentiated despite similarities such as low cellular density, predominance of nesting melanocytes, vascularization, and abundant fibrous stroma. Furthermore, desmoplastic Spitz nevi tend to be small, well-circumscribed, superficial lesions, whereas desmoplastic melanomas are often large, poorly demarcated, and are characterized by deep involvement of the dermis or subcutis. Other differentiating criteria between desmoplastic Spitz nevi and desmoplastic melanomas are that the former are typically composed of large epithelioid cells, while the latter are characterized by hyperchromatic nuclei of pleomorphic spindle cells. Additionally, cells in desmoplastic Spitz nevi are uniformly positive for melan-A, while the vast majority of desmoplastic melanomas are negative in their spindle cell compartments. A high labeling index (>5%) of Ki-67 may indicate the diagnosis of desmoplastic melanoma and distinguish it from desmoplastic Spitz nevi (<2% in our patient).<sup>4</sup> Absence of the expression of CD-34 and epithelial membrane antigen in desmoplastic Spitz nevi can rule out spindle cell tumors originating from fibroblasts or endothelial cells. Complete excision of the lesion with a clear margin is generally considered as the optimal treatment for desmoplastic Spitz nevus. Recurrence resulting from incomplete excision may occur in 7–16% of patients.<sup>5</sup>

In conclusion, Spitz nevus should be considered in the differential diagnosis of a solitary reddish nodule over the face or lower limbs, and skin biopsy should be performed to confirm its nature. The absence of recurrence or metastasis of the lesion after complete excision confirms its benign behavior.

## References

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