SPECIAL SELECTION

Slowly Enlarging, Erythematous Macule in a Child

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A 7-YEAR-OLD BOY was brought to the dermatology clinic by his mother for evaluation of a "birthmark." The lesion had appeared on his right cheek at the age of 6 months and had slowly spread to the right submandibular area. According to the mother, a portion of the initial lesion had shown some regression. Over the past year, the advancing border had spread and become raised. The patient reported that the lesions were tender to palpation. He was otherwise in good health.

Examination of the skin revealed a partially blanching, erythematous, macular patch on his right cheek, with extension to the right submandibular area (**Figure 1**). The medial border was raised. A similar-appearing satellite nodule was present on the right side of the chin. An incisional biopsy specimen, which included both the raised and the macular areas, was obtained for histopathologic examination (**Figure 2** and **Figure 3**).

What is your diagnosis?

From the University of Virginia, Charlottesville.

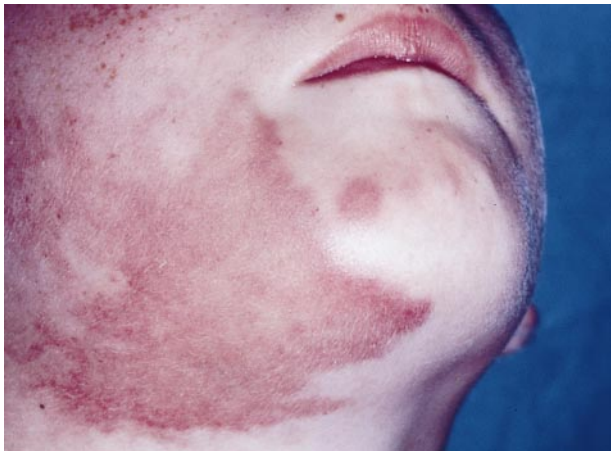

Figure 1.

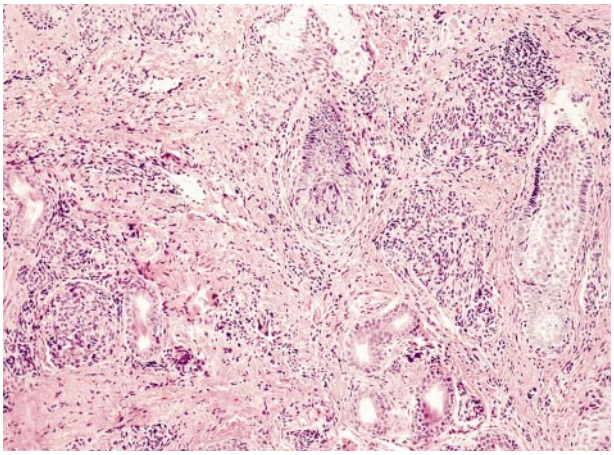

Figure 2.

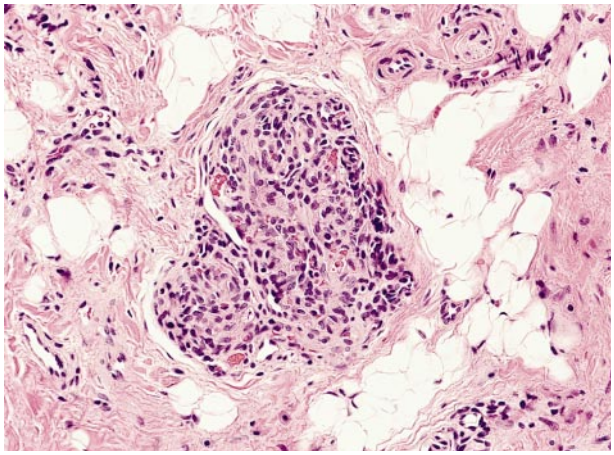

Figure 3.

Diagnosis and Discussion

Tufted Angioma

HISTOPATHOLOGIC FINDINGS

Histopathologic examination of the biopsy specimen showed numerous lobules ("tufts") of plump, cytologically bland spindle cells located at various levels in the dermis. The tufts contained sparse cleftlike vascular lumina. Some tufts protruded into thin-walled, semilunar vessels that partially surrounded the tufts.

DISCUSSION

In 1989, Wilson Jones and Orkin[1] described 20 patients with benign, progressive vascular lesions that had a distinctive histopathologic appearance. They termed these lesions *tufted angioma.* Previously, similar lesions had been described as "angioblastoma" in the Japanese literature[2,3]; these are now considered by many to be identical to acquired tufted angioma.[1,4-6]

A review of 37 cases in the English-language literature reveals that tufted angioma has a number of characteristic clinical features. Onset of lesions most commonly occurs in early childhood (14% at birth, 49% at <1 year of age, and 77% at <10 years of age). Both sexes are affected equally. Typically, lesions occur on the neck, trunk, and back. Tufted angioma usually consists of dull erythematous or brown macules, papules, and/or nodules that slowly enlarge and then become stable. They are frequently tender. At presentation, most of the lesions have been clinically diagnosed as vascular malformations, although in several cases, granulomatous disease or connective tissue abnormalities were initially suspected. Patients often report that the lesions are tender. Unlike congenital capillary hemangioma, tufted angioma does not ordinarily undergo spontaneous involution, although this has been noted in three cases.[5,7,8] Despite progressive enlargement, tufted angioma behaves in a benign manner.

It is by histopathologic examination that the definitive diagnosis is made. Low-power microscopy shows round to ovoid cellular tufts of capillaries, most prominent in the middle to lower dermis. Wilson Jones and Orkin[1] described this as a "cannonball" distribution. At higher magnification, the capillaries have narrow, cleftlike lumina that often lack erythrocytes. A distinctive feature of some tufts is the presence of an ectatic vessel into which the lobular tuft protrudes, imparting a glomeruloid appearance. Cellular atypia is not present.

The histologic differential diagnosis includes juvenile capillary (strawberry) angioma, Kaposi's sarcoma, angiosarcoma, and lobular capillary hemangioma. The first three lack the lobular architecture of tufted angioma; also, angiosarcoma and Kaposi's sarcoma show nuclear atypia. Lobular capillary hemangioma differs in that it shows good circumscription, larger numbers of open capillaries, its own stroma, and thick-walled feeding vessels outside the main capillary aggregates.[1] Padilla et al[4] have suggested that while tufted angioma is a distinct clinicopathologic entity, it may be closely related to lobular capillary hemangioma, as supported by immunocytochemical and electron microscopic observations.

Treatment of this condition to date has not been satisfactory. Simple excision, excision and grafting, and cryosurgery have been successful in a few cases. However, most lesions are too large for these treatment modalities, and there are reports of recurrence up to several years after removal. There has been one report of a lesion treated with pulsed dye laser, with a satisfactory result.[9] However, because the lesions are cellular and often are poorly canalized and involve deep dermis, pulsed dye laser therapy is not likely to be successful. Although these lesions can progress, it is important to reassure the patient that tufted angioma is a benign condition.

Selected from Arch Dermatol. *1996;132:703. Off-Center Fold.*

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