Tufted Angioma and Juvenile Xanthogranuloma

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This tender lesion on the right cheek of a 4-year-old white girl had appeared shortly after her birth. It had subsequently enlarged for about 3 years before stabilizing. Physical examination revealed an erythematous arcuate plaque with a slightly thickened border that extended from the right oral commissure onto the right cheek. There was no family history of similar lesions, and the child was otherwise healthy.

Figure
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Case 1:

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What is this lesion? What disorders would you include in the differential?

Case 2:

A 10-month-old child presented with this gradually enlarging lesion on his upper back that had first appeared when he was 2 months old. On physical examination, there was a 2.5-cm yellow plaque with surrounding similar-appearing satellite papules. Histological examination of a biopsy specimen showed a histiocytic infiltrate and foamy histiocytes. The patient was asymptomatic.

Do you recognize this lesion?

**Dermclinic-Answers**

**Case 1: Tufted Angioma**

The differential diagnosis included hemangioma of infancy, angiosarcoma, Kaposi sarcoma, and eccrine angiomatous hamartoma, in addition to other vascular malformations and tumors. A biopsy was obtained and histopathological examination revealed a "cannon ball" pattern of highly cellular capillaries without cellular atypia. These findings confirmed a diagnosis of tufted angioma.

Treatment with a 585-nm pulsed dye laser did not diminish the lesion. The patient was followed in the clinic for 3 years, during which time the angioma remained stable and the tenderness decreased. Tufted angiomas—also known as angioblastoma of Nakagawa—are rare benign vascular tumors predominantly seen in patients younger than 5 years. However, the tumors can occur at any time throughout life. Both sexes are affected equally.

The name is derived from the presence of clusters of capillary tufts that are surrounded by lymphatic vessels in the lower dermis. Tufted angiomas are most often located on the trunk, neck, and shoulders and can range from several millimeters to several centimeters in size. They generally grow slowly over several months to years. Unlike hemangiomas of infancy, tufted angiomas rarely regress. A few cases of congenital tufted angiomas have been reported, and it has been suggested that lesions that appear at or shortly after birth may have a greater rate of regression than those that appear later in life.

Clinically, the tumors generally have a rubbery consistency and are often tender. They may also be covered by lanugo hair. Proliferation of eccrine glands can lead to hyperhidrosis. There are rare reports of tufted angioma as a cause of Kasabach-Merrit syndrome.

Histological examination of a biopsy specimen is required to confirm the diagnosis. Laboratory study results are generally normal. MRI may be indicated if muscle or fascia involvement is suspected. Observation is the recommended therapy for isolated tufted angiomas. NSAIDs can provide relief during painful episodes, and some studies have suggested that topical, intralesional or systemic corticosteroids, and interferon alfa may be beneficial. However, data are limited because of the rarity of the condition. Surgical excision may be indicated for cosmetic reasons, if functionality is compromised, or if the lesion is painful. However, surgery is not always practical for large lesions. Other treatments, including cryosurgery, radiation, electrocoagulation, pulsed dye laser and intense pulsed light, have shown limited success in this setting.

**Case 2: Giant Juvenile Xanthogranuloma**

Juvenile xanthogranuloma (JXG) most often manifests at birth, infancy, or childhood. It is a benign, self-limited, red to yellow papule or nodule that may be solitary or multiple. It usually regresses spontaneously with little change to the affected skin. It is most often found on the skin of the head, neck, trunk, and upper extremities. However, manifestations can occur in the eye or visceral organs.

The cause of JXG is unknown. Biopsy findings reveal a time-dependent progression. Early specimens reveal a dense histiocytic infiltrate in the dermis. Older lesions contain giant cells and foamy histiocytes. JXG may represent a granulomatous reaction to an unidentified stimulus. JXG itself is benign, but it can be associated with a variety of other multisystemic diseases, including type 1 neurofibromatosis (NF1). Retrospective analysis shows that JXG develops in as many as 1 in 5 children who have NF1 before age 3 years. The appearance of multiple JXGs has also been
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associated with juvenile myelomonocytic leukemia, especially in patients with coexisting NF1. Therefore, patients who have both NF1 and JXG need to be monitored for the development of leukemia.\(^4\) Ocular lesions of JXG can lead to glaucoma and blindness; affected patients should be referred to an ophthalmologist.\(^5\)

JXG is said to be giant when lesions are greater than 2 cm in diameter. The clinical diagnosis of such lesions can be difficult, and biopsy is often necessary to rule out more worrisome tumors. The natural history of giant JXG is uncertain. However, it appears to favor the benign, self-limited nature of the smaller lesions.\(^6\) Nonoperative management with observation is advisable, with surgical intervention available as necessary.

References:

**Case 1**


**Case 2**


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