

Egyptian Dermatology Online Journal

Volume 1 Number 1

Tufted Angioma

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Egyptian Dermatology Online Journal 1 (1): 9, June 2005

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Summary

Tufted angioma is an uncommon benign vascular neoplasm, localized to the skin and subcutaneous tissues. Half of the cases present in the first year of life. We describe a case in a 34-year-old male who presented with different sized reddish nodules, occurring on multiple hyperpigmented erythematous macules situated on the proximal parts of upper limbs that first appeared three years earlier. Histopathology revealed circumscribed multiple foci of closely set vascular channels giving the cannon ball appearance and were of vascular origin .

Introduction

Tufted angioma is an uncommon benign vascular neoplasm, localized to the skin and subcutaneous tissues with no documented systemic or metastatic involvement. Tufted angioma were described in the literature under different names including Nakagawa's angioma, Nakagawa's angioblastoma, progressive capillary hemangioma, and acquired tufted angioma of the skin and subcutaneous tissue[1]. It is characterized by slow angiomatous proliferation with no racial predilection, and occurs equally in both sexes. The skin lesions occur without a history of preceding trauma, except for one case where tufted angioma has followed an arthropod bite[2]. The lesions are usually asymptomatic but painful episodes have been described [3].

Case Presentation

A 34-year-old male presented with multiple hyperpigmented erythematous macules on the proximal parts of the upper limbs, shoulders, and dorsal aspects of the forearms of 3-year duration. Nodules of different sizes appeared on top of some of the macules. The nodular lesions varied in color from light to dark red and were restricted to the macules, with no involvement of the normal skin.

The clinical differential diagnosis included Kaposi's sarcoma, pyogenic granuloma,

hemangioendothelioma, bacillary angiomatosis, and angiosarcoma.

Two biopsies were taken, one from the macular lesions, and the second from one of the nodules. The first biopsy (from the macules) showed basal cell hyperpigmentation, and dermal cellular infiltrate. The second biopsy (from one of the nodules) showed multiple, discrete, circumscribed dermal foci of closely set vascular channels, with the epidermis surrounding one of the foci in a claw-like manner. The tumor masses appeared as dilated capillary channels lined with spindle shaped infiltrating cells, with no atypia or mitotic figures, and there were RBCs inside the cavities with no extravasations. All tumor cells stained positively with CD34, while factor VIII was positive in some vessels.

CBC, prothrombin time, partial thromboplastin time, and coagulation factors, and bleeding time were normal.



Figure 1

Reddish nodules, occurring on multiple hyperpigmented erythematous macules.

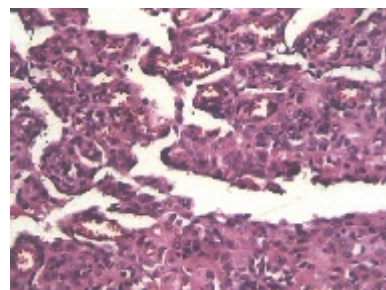


Figure 2

H&E section: tumor masses appeared as dilated capillary channels lined with spindle shaped infiltrating cells, with no atypia or mitotic figures, and there were RBCs inside the cavities with no extravasations..

Discussion

More than 50% of cases of tufted angioma occur within the first year of life, and of those, 15% are believed to have had a blemish in the area where tufted angioma appeared later, which suggests a congenital mode of presentation[4]. Most of cases (60-70%) of tufted angioma develop before the age of five. Fewer than 10% of cases with tufted angioma are older than 50 years. In individuals older than 60 years, the disease is very rare [5].

Tufted angioma presents clinically as ill-defined slowly progressive dull red, brownish or purple macules or plaques with mottled surfaces. It slowly enlarges over six months to ten years, after which no further growth occurs[6]. Papules and nodules may superimpose on the main lesions[1] as was observed in the present case.

The sites most commonly involved are the upper trunk, neck and shoulders, less

commonly the face, scalp, and proximal extremities. Rarely does it affect the feet and oral mucosa. Partial regression may occur, but complete disappearance is rare[7].

Hyperhidrosis may occur in the skin overlying the lesion, and when it occurs, it corresponds histologically with an area of abundant collagen[4]. Occasional association of the more serious platelet-trapping syndrome (Kasabach-Merritt syndrome) has been reported[8], so the presence of petechiae or ecchymotic patches should alert the physician to the development of those complications. Cases of tufted angioma have been reported on the skin of a liver allograft recipient, but regressed after modulation of immunosuppressive therapy[9].

Different modalities of treatment have been tried, including complete surgical excision, soft X-ray therapy, and electrocoagulation. Pulsed dye laser has also been tried without encouraging results[10]. Potent topical steroids have been used to reduce pain[2]. Interferon α_2 , and high doses of systemic steroids should be tried before proceeding to excision, particularly where lesions are excessive[11, 12].

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