

Acquired tufted angioma: Case report

Edinsel tufted angioma: Olgu sunumu

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ABSTRACT

Tufted angioma is a rare benign vascular neoplasm, localized to the skin and subcutaneous tissues. Half of the cases present in the first year of life. In the histopathological examination, multiple ovoid and round lobules of endothelial cells and capillaries arranged in “cannon ball” appearance in the dermis were seen. Here we report a case of the tufted angioma on the back of an adult woman patient and cutaneous lesion has been present for 3 years. The awareness of occurrence of tufted angioma in adult is important for it is differential diagnosis of skin lesions, which includes complete surgical removal, to prevent recurrence.

Key words: Hemangioma, adult, skin

INTRODUCTION

Acquired vascular neoplasms, labelled as a “tufted angioma”, are rare angiomatous lesions first described in 1976 by Wilson-Jones [1]. Subsequently, he and Orkin [2] described 20 cases, while other authors described individuals. Tufted angioma was described in the literature under different names including Nakagawa’s angioma, Nakagawa’s angio-blastoma, progressive capillary hemangioma, and acquired tufted angioma on the skin and subcutaneous tissue [3,4]. These tumors occur primarily on the trunk and extremities of children and have a benign course of slow angiomatous proliferation, without any evidence of aggressive behavior or metastases. The lesions are usually asymptomatic but, rarely paroxysmal painful episodes may also be associated with it. It occurs equally in both sexes without any predilection [5,6].

ÖZET

Tufted angiom deri ve deri altında lokalize nadir görülen benign vasküler bir neoplazmdır. Olguların yarısı yaşamın ilk yılında görülür. Histopatolojik incelemesinde, dermiste kapiller ve endotelial hücrelerin oluşturduğu ovoid ve yuvarlak lobüler yumaklar görülür. Biz burada 3 yıldır sırtında deri lezyonu mevcut olan ve tufted angioma tanısı alan erişkin kadın hastayı sunuyoruz. Erişkinlerde görülen tufted anjiomanın cilt lezyonlarından ayırımında akılda tutulması ve nüksün önlenmesi için total olarak çıkarılması önemlidir.

Anahtar kelimeler: Hemanjiom, erişkin, deri

CASE REPORT

A 49-year-old woman presented with solitary a reddish-brown elevated papule about 2 mm, on the upper back. The lesion has been present for at least 3 years. There were painful episodes, pruritus and tenderness but no history of trauma, bleeding from the lesion or from other parts of the body, or increased sweating or discharge from the lesion. Other parts of the skin, hair, nails, and mucosa were normal. Systemic examination and complete biochemical blood tests were normal. The clinical differential diagnosis included Kaposi’s sarcoma, pyogenic granuloma, hemangioma and bacillary angiomatosis. The lesion was excised completely.

In the histopathological examination, the lesion was composed of multifocal, tightly packed glomerular lobules and tufts of spindle or polygonal cells associated with endothelial cells. The epidermis and

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the subcutis were not involved. The glomerular lobules consisted of multiple capillaries with bloodless lumina, surrounded by dilated crescent-shaped vascular channels. The proliferated tumor cells were principally uniform with poorly outlined, eosinophilic cytoplasm and partly fusiform, partly circular nuclei (Figure 1). There was no cellular atypia or mitotic figures. Immunohistochemical studies showed strong positive staining of endothelial cells with CD34 (Figure 2), while factor VIII and D2-40 was negative. Apart from this, pericytes which were positive for smooth muscle actin were identified. Up to 10% of the tumor cells were positive for Ki-67 (MIB-1).

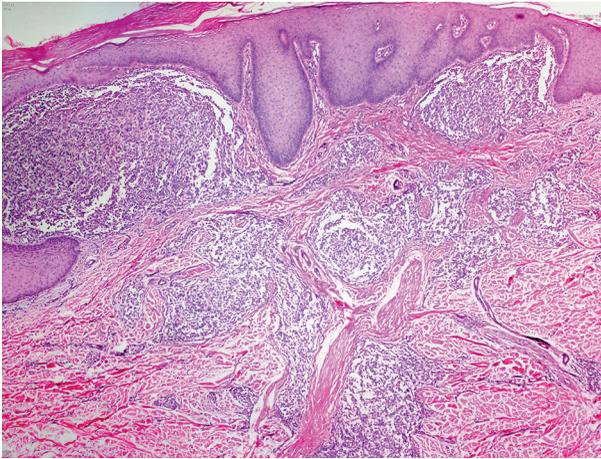


Figure 1. Capillary clusters in papillary dermis and reticular dermis (H&E x100)

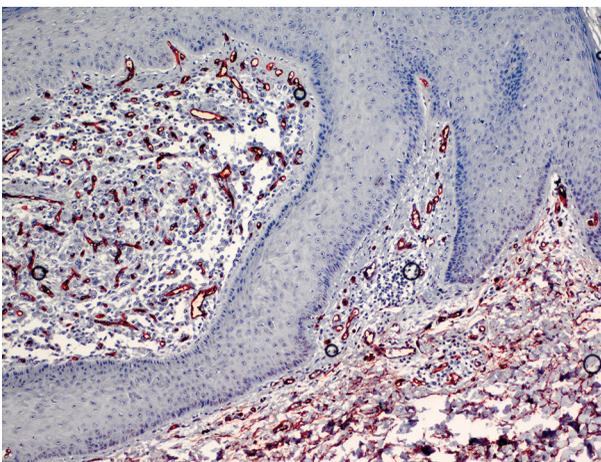


Figure 2. Immunostaining for CD34 was positive in the tumor (x200)

DISCUSSION

Tufted angioma is a benign and distinctive angiomatous condition. It usually occurs between the ages of one and five, involving both sexes equally. Occasionally, the lesion can be present at birth, and a delayed onset on fifth and sixth decades have also been reported [2,7]. The lesion of tufted angioma enlarges slowly over 5 months to 10 years, after which no further growth occurs. Although multiple lesions may occur, tufted angioma usually presents as a solitary nodule or papule. Clinically, tufted angioma appears as a deep red or purple patch or plaque with superimposed angiomatous papules that predominantly appear on the upper trunk, neck, and shoulders. Face, scalp and proximal extremities are rarely involved [5].

Tufted angioma can have variable presentations of mimicking pyogenic granuloma, hemangioma, vascular malformation or Kaposiform hemangioendothelioma (KHE) [8,9]. Our case was on the upper back in the presentation and it was a very well-defined papular lesion without any signs of vascular pathogenesis like a port wine stain, telangiectasia or any deeper tissue swelling. Most cases of reported childhood tufted angiomas couldn't distinguish from KHE. Both occur principally in children, are characterized by infiltrating nodules of tumor with focal glomeruloid structures, display a lymphatic component, and have a similar immunophenotype. Several reports even comment on the fact that some lesions exhibit features of both tumors or show transformation between the two. KHE and tufted angioma sometimes show similar histopathological features. KHE, sheets of spindle cells show discrete aggregates of epithelioid endothelial cells and hyaline globules, Well-formed vascular channels showing intraluminal thrombi are often seen at the periphery of the tumor nodules. Differential diagnosis for immunohistochemistry markers may be helpful [7].

Various treatment options are available for the tufted angioma. The recurrence rate is very high after surgical excision. Few cases have shown good response to a high dose of systemic steroids. Potent topical steroids have been used to reduce pain. Cases of tufted angioma have been reported on the skin of a liver allograft recipient, but regressed after modulation of immunosuppressive therapy. Subcutaneously administered interferon alpha has shown

a significant response in a child with tufted angioma involving the jaw and neck. Other treatment options are radiotherapy, cryotherapy and pulsed dye laser [6,7,9].

We excised the complete lesion surgically and no recurrence occurred after 2-year of follow-up. This case is presented for it is acquired onset, which is rare.

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