Tufted hemangioma: clinical case and literature review

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Keywords: tufted hemangioma, hemangioma, histopathology


Received: November 5, 2013; Accepted: January 11, 2014; Published: April 30, 2014

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Funding: None.

Competing interests: The authors have no conflicts of interest to disclose.

All authors have contributed significantly to this publication.

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ABSTRACT
Tufted hemangiomas are relatively rare benign vascular proliferations that are congenital or appear during the first years of life. Herein we present an additional case of tufted hemangioma that appeared one year after birth and discuss its histopathological criteria and differential diagnosis with malignant vascular proliferations including sarcoma Kaposi, angiosarcoma and kaposiform hemangioendothelioma.

Observation

A 26-year-male patient presented with a brownish plaque in the flank area. It appeared one year after birth and was partially excised shortly after. Histopathological examination was not performed at that time. Over the past several years the lesion has gradually enlarged with the appearance of new smaller lesions in the vicinity of the main lesion (Figure 1). The patient is otherwise healthy.

A biopsy revealed a vascular proliferation with round and poorly defined collections of epithelioid and spindle cells. Dilated slit-like vascular spaces, resembling those of lymphatic vessels, were seen in the sclerotic stroma (Figures 2-4).

Cracked irregular spaces were seen between the cells in the proliferation (Figure 5). Scarring from the previous excision was evident (Figure 2 and 3).

The vascular proliferation was positive for CD31, CD34, D2-40 and negative for HHV8. Ki-67 rate was low (Figure 6).

The diagnosis of tufted hemangioma was established on the basis of clinical presentation, history and histopathological presentation.

Discussion

Tufted hemangiomas are relatively rare benign vascular proliferations. Although initially described in the European and Japanese literature, the term “tufted hemangioma” was introduced in 1989 by Jones and Orkin [1] along with the description of the largest group of 20 patients. They usually appear during the first years of life and are congenital in up to 50% of cases [2].
sometimes present on the surface of plaques or surrounding
the lesions. In children the lesions may be painful.

Histopathologically the lesions are characterized by
collections of vessels in small tufts with a cannonball dis-
tribution, encircled by empty cleft-like vessels and often
surrounded by sclerotic dermis. Tufts are composed of epithe-
lioid and spindle cells with slit-like spaces resembling Kaposi
sarcoma. Hyaline globules may be seen within the tufts [2].
Widened vessels, resembling lymphatic ones, are often a
source of potential diagnostic mistakes if the biopsy is taken
at the periphery of the lesion [4]. Tufted hemangiomas express
endothelial and lymphatic vascular markers CD31, CD34,
VEGF-A and D2-40. Mitotic rate is usually low.

The differential diagnosis of tufted hemangioma is with a
variety of vascular tumors. Although Kaposi sarcoma resem-
bles tufted hemangioma histopathologically, it rarely affects
children. Clinical data (HIV status, immunodeficiency, and
African or Mediterranean origin) and positivity for HHV8
allow distinguish both entities with certainty. Presence of

Tufted hemangiomas predominantly affect males and are
located on the trunk, neck or extremities [1,2]. They clini-
cally present as patches or plaques, although subcutaneous
masses may be seen [3]. Hypertrichosis and hyperhidrosis are

Figure 1. Large vascular lesion present since infancy (with the scar
from the previous biopsy) and smaller lesions in the flank area.
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Figure 2. Vascular lesion with ill-defined vascular proliferations in
a sclerotic stroma. Hematoxylin and eosin stain, x40. [Copyright:
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Figure 3. Ill-defined and round vascular proliferations in a sclerotic
stroma. Hematoxylin and eosin stain, x100. [Copyright: ©2014 Ka-
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Figure 4. Rounded collections of epithelioid cells surrounded by slit-
like vascular lumina. Small hyaline globules are noted. Hematoxylin
and eosin stain, x600. [Copyright: ©2014 Kazlouskaya et al.]

Figure 5. Collections of epithelioid cells with cracked spaces. He-
matoxylin and eosin stain, x200. [Copyright: ©2014 Kazlouskaya
et al.]
siform hemangioendotheliomas (known as Kasabach-Merritt syndrome) may be a grave consequence of both conditions. The phenomenon is explained by entrapment and adhesion of the thrombocytes to the endothelial cells of the hemangioma. Subsequent activation of the thrombocytes leads to coagulopathy. Large hemangiomas may lead to congestive heart failure [7]. Osio et al. described a clinical variant of tufted hemangiomas with chronic coagulopathy, but without thrombocytopenia [2].

There are no standard guidelines for the treatment of tufted hemangiomas. If the lesion is not associated with Kasabach-Merritt syndrome no treatment is usually necessary. Kasabach-Merritt syndrome is aggressively treated with corticosteroids, vincristine or interferon-α7. Excision or laser modalities may be implemented for cosmetic reasons.

References