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Tufted hemangioma: clinical case and literature review

Viktoryia Kazlouskaya[1], Bohdan Lytvynenko[2], Elen Blochin[1]

1 Ackerman Academy of Dermatopathology, New York, NY, USA
2 "Euroderm" Clinic, Kyiv, Ukraine

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Corresponding author: Viktoryia Kazlouskaya, M.D., Ph.D., Ackerman Academy of Dermatopathology, 145 E 32 St, Fl 10, New York, NY 10016, USA. E-mail: viktoriakozlovskaya@yahoo.com

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].

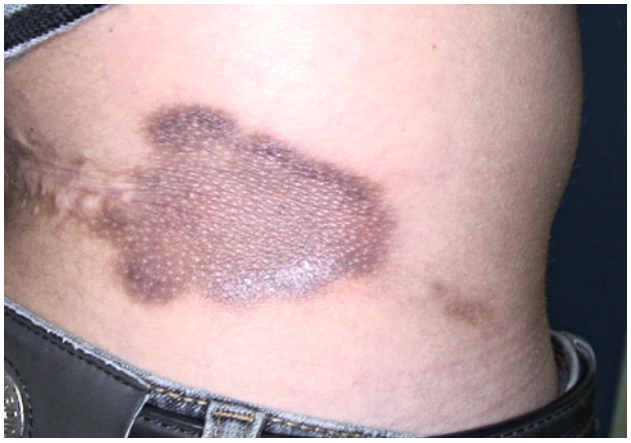

Figure 1. Large vascular lesion present since infancy (with the scar from the previous biopsy) and smaller lesions in the flank area. [Copyright: ©2014 Kazlouskaya et al.]

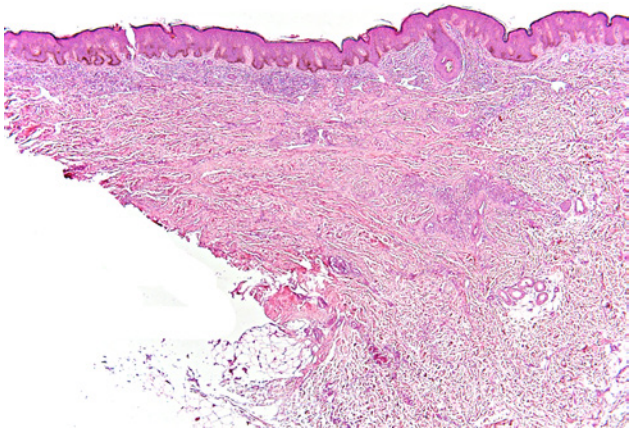

Figure 2. Vascular lesion with ill-defined vascular proliferations in a sclerotic stroma. Hematoxylin and eosin stain, x40. [Copyright: ©2014 Kazlouskaya et al.]

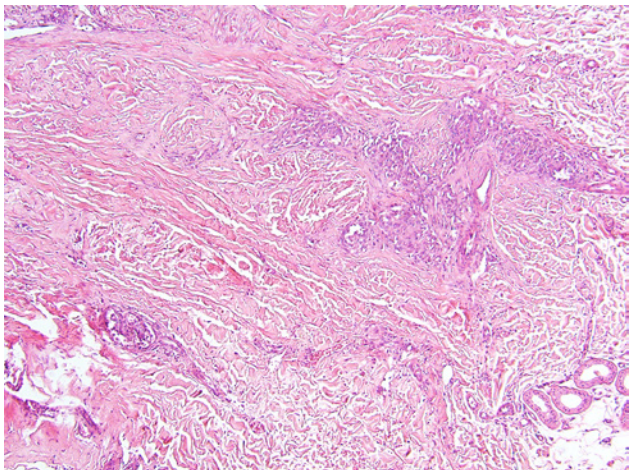

Figure 3. Ill-defined and round vascular proliferations in a sclerotic stroma. Hematoxylin and eosin stain, x100. [Copyright: ©2014 Kazlouskaya et al.]

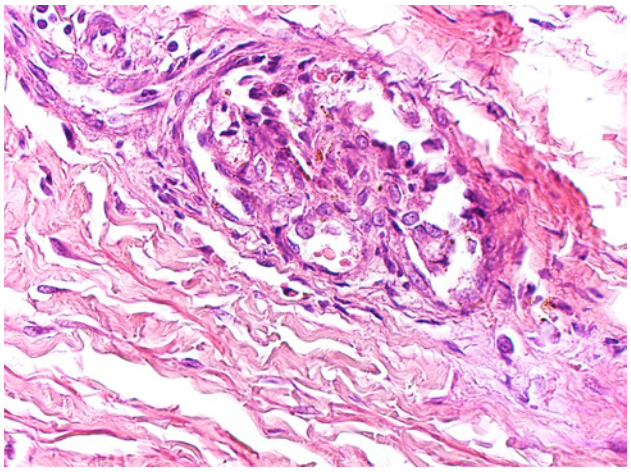

Figure 4. Rounded collections of epithelioid cells surrounded by slit-like vascular lumina. Small hyaline globules are noted. Hematoxylin and eosin stain, x600. [Copyright: ©2014 Kazlouskaya et al.]

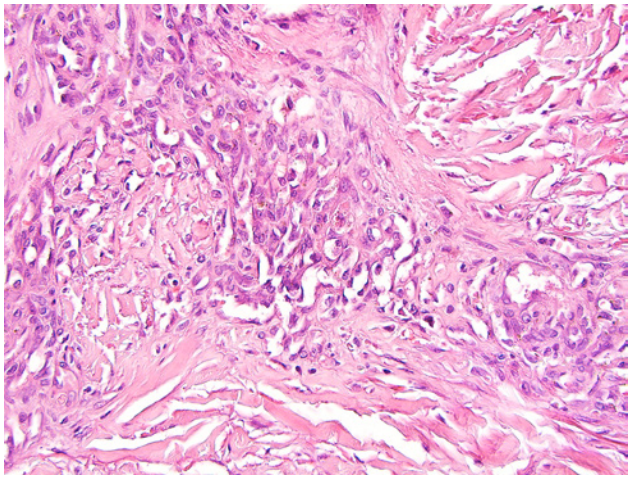

Figure 5. Collections of epithelioid cells with cracked spaces. Hematoxylin and eosin stain, x200. [Copyright: ©2014 Kazlouskaya et al.]

Tufted hemangiomas predominantly affect males and are located on the trunk, neck or extremities [1,2]. They clinically present as patches or plaques, although subcutaneous masses may be seen [3]. Hypertrichosis and hyperhidrosis are 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 distribution, encircled by empty cleft-like vessels and often surrounded by sclerotic dermis. Tufts are composed of epithelioid 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 resembles 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

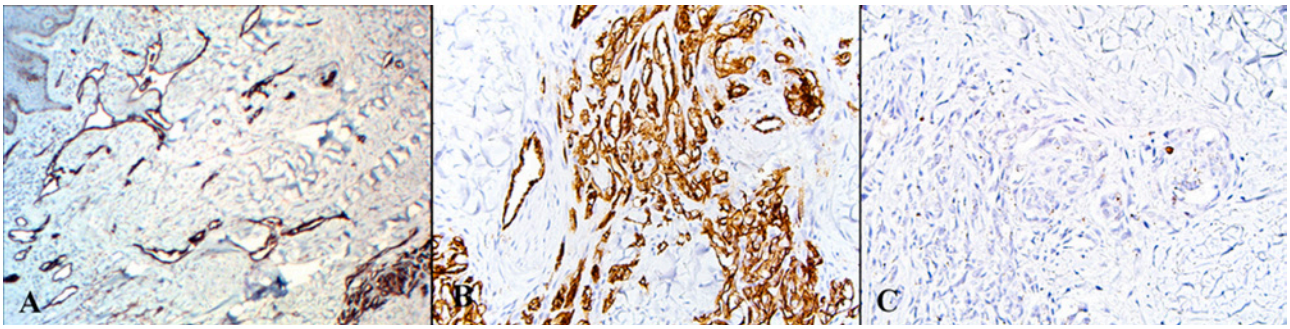

Figure 6. (A) D2-40 positivity in the small and large dilated vessels in the superficial dermis, x100. (B) Expression of CD31 in the small vessels, x200. (C) Sparse cells expressing Ki-67, x200. [Copyright: ©2014 Kazlouskaya et al.]

spindle cells, formation of new vessels around the preexisting cutaneous structures ("promontory sign"), variable cellular polymorphism may help in the diagnosis of Kaposi sarcoma. Angiosarcomas usually affect elderly patients and have a predilection for the scalp. Multiple mitotic figures and marked atypia are the hallmarks of angiosarcomas. The differential diagnosis of kaposiform hemangioendothelioma and tufted hemangioma is challenging. Some authors consider both to be within the spectrum of the same condition. Tufted hemangiomas tend to be located more superficially compared to superficial kaposiform hemangoenothelioma which may affect subcutis and retroperitoneum, invade the internal organs and is characterized by aggressive growth. Podoplanin (D2-40) was proposed as a useful marker for the differential diagnosis of tufted hemangioma and kaposiform hemangioendothelioma by Arai et al [5]. In their study D2-40 was positive in small capillaries in the Kaposi sarcoma-like part of the proliferation and was negative in the widened superficial vessels in kaposiform hemangioendothelioma. In contrast, in tufted hemangiomas, D2-40 labeling was seen in the widened superficial vessels and was negative or patchy in the capillaries of the cannonball vessels [5]. Other studies, as well as our case report, showed inconsistency of these findings and demonstrated that D2-40 may also be positive in the small capillaries in tufted hemangiomas [4]. Infantile hemangiomas are seen only in newborns and usually involute. They appear mostly commonly on the face and neck. Prox1 has been recently described as a potential useful marker for the differential diagnosis of tufted hemangiomas/kaposiform hemangioepitheliomas versus infantile hemangiomas and pyogenic granuloma [6]. It is usually positive in tufted hemangiomas/kaposiform hemangioendotheliomas, but is not expressed in infantile hemangiomas and pyogenic granulomas.

The clinical course of tufted hemangiomas is variable. More often the lesions gradually progress. Cases of spontaneous involution have been reported [2]. About 30% of patients may develop thrombocytopenia. Thrombocytopenia and coagulopathy in patients with tufted hemangiomas and kaposiform 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

1. Jones EW, Orkin M. Tufted angioma (angioblastoma). A benign progressive angioma, not to be confused with Kaposi's sarcoma or low-grade angiosarcoma. J Am Acad Dermatol. 1989;20:214-25.
2. Osio A, Fraitag S, Hadj-Rabia S, et al. Clinical spectrum of tufted angiomas in childhood: a report of 13 cases and a review of the literature. Arch Dermatol. 2010;146:758-63.
3. Herron MD, Coffin CM, Vanderhooft SL. Tufted angiomas: variability of the clinical morphology. Pediatr Dermatol. 2002;19:394-401.
4. Sadeghpour M, Antaya RJ, Lazova R , Ko CJ. Dilated lymphatic vessels in tufted angioma: a potential source of diagnostic confusion. Am J Dermatopathol. 2012;34:400-3.
5. Arai E, Kuramochi A, Tsuchida T, et al. Usefulness of D2-40 immunohistochemistry for differentiation between kaposiform hemangioendothelioma and tufted angioma. J Cutan Pathol. 2006;33:492-7.
6. Le Huu AR, Jokinen CH, Rubin BP, et al. Expression of prox1, lymphatic endothelial nuclear transcription factor, in Kaposiform hemangioendothelioma and tufted angioma. Am J Surg Pathol. 2010;34:1563-73.
7. Rodriguez V, Lee A, Witman PM, Anderson PA. Kasabach-Merritt phenomenon: case series and retrospective review of the mayo clinic experience. J Pediatr Hematol Oncol. 2009;31:522-6.