Case for diagnosis*

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CASE REPORT

14-year-old male patient had a 9-month history of tumoral lesion in the occipital region. The lesion showed progressive growth, bleeding upon trauma and was slightly painful. The patient denied comorbidities and previous lesions. On examination, we found a well-delimited, 3x2cm-sized, violet-colored erythematous tumor, with yellowish crusts on its surface, in the right occipital region (Figure 1).Complete excision of the tumor was performed.

The anatomicopathological examination showed an endothelial proliferation with multiple delicate papillary projections. It was composed of a single layer of endothelial cells without atypia that involved a collagenized axis containing occasional capillary vessels. There was presence of rare mitotic figures. The lesion was well circumscribed, without necrosis and pleomorphism (Figure 2). Immunohistochemistry revealed that: the endothelial cells were immunopositive for CD34; stromal and endothelial cells were immunopositive for vimentin; and Ki67 was positive in less than 3% of endothelial cells (Figure 3). There was no recurrence in the 8 month follow-up period.

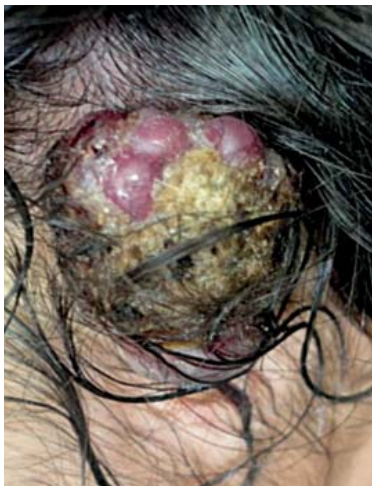

FIGURE 1: Well-delimited, violet-colored erythematous tumors with yellowish crusts on the surface

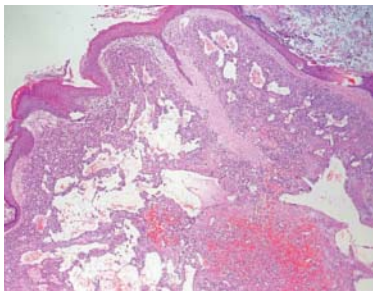

FIGURE 2: 40x HE. Vascular tumor with papillae, and area of thrombosis and hemorrhage.Courtesy of the Pathology Service of the Evangelical Hospital of Curitiba

DISCUSSION

The diagnosis of intravascular papillary endothelial hyperplasia (IPEH) was confirmed by clinical and histopathological findings. IPEH is a benign, vascular lesion caused by endothelial proliferation, and it corresponds to 2% of all vascular neoplasms of the skin.[1] It was first described by Masson, and the main significance of this intravascular endothelial hyperplasia is its clinical and histological resemblance to angiosarcoma.[2] Three forms of IPEH have been described.A primary form occurs in dilated vessels,without previous malformations.The secondary form of the disease occurs when there are preexisting lesions (hemangioma, pyogenic granuloma). A tertiary form may be found in hematomas.[3]

The disease is reactive to thrombotic or inflammatory stimuli in the vessel wall and may be associated with local trauma. One probable etiology is the production of fibroblast growth factor by endothelial cells in the tumor.[4] The incidence is higher in women and it is most common in the third and fourth decades of life.[2,4]

It consists of a violet-colored erythematous, firm, well-delimited and slow-growing nodule or mass. The most common sites are the fingers, head

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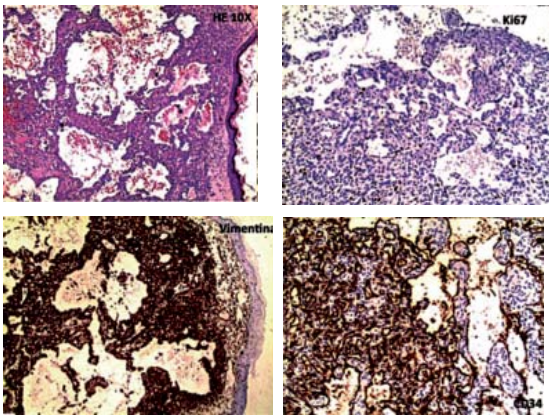

Figure 3: Immunohistochemical study. Endothelial cells immunopositive for CD34; stromal and endothelial cells immunopositive for vimentin; and positive Ki67 in less than 3% of endothelial cells. Courtesy of the Pathology Service of the Evangelical Hospital of Curitiba

and neck. The main differential diagnoses are angiosarcoma, pyogenic granuloma, Kaposi's sarcoma, hemangioma, angioendothelioma, papular angioplasia, Kimura´s disease, intravascular atypical vascular proliferation and amelanotic melanoma.[4]

Due to the nonspecific clinical picture, histology is mandatory for diagnosis. Histopathologically, IPEH consists of fibrous connective tissue, lined by one or two layers of endothelial cells that form papillary structures. These papillary structures may fuse and form an anastomosing vascular network, as found in the lesion of the patient described here.[2] Endothelial cells are positive for CD31, CD34 and factor VIII-related antigen (as in angiosarcoma), and show positive reaction for vimentin in mesenchymal cells.[5,6] In order to differentiate it from angiosarcoma,distinguishing features of the benign disease should be considered, such as well-circumscribed lesion, papillary formation related to thrombotic material, one or two layers of endothelial cells, hyperchromatic endothelial cells, lack of cellular atypia, necrosis, tissue invasion and irregular capillary vessels, and rare mitotic activity.[3,7]

The treatment of choice is excision and the prognosis is good. Recurrence is rare and, when it occurs, is associated with secondary lesions and incomplete excision.[8] Since it is a rare, benign vascular neoplasm, it should be remembered in order to be differentiated from angiosarcoma, which has a bad prognosis and require invasive treatment. ❑

Abstract: Intravascular papillary endothelial hyperplasia is a benign vascular lesion caused by proliferation of endothelium. It is reactive to thrombotic or inflammatory stimuli in the vessel wall.We report the case of a 14-year-old male patient with a violet-colored erythematous tumoral lesion of progressive growth in the occipital region. The diagnosis of intravascular papillary endothelial hyperplasia (IPEH) was confirmed by clinical and histopathological findings. Total lesion exeresis was performed with no recurrence up to date. IPEH presents clinical importance due to its clinical and histological resemblance to angiosarcoma. In order to differentiate it from angiosarcoma, distinguishing features of the benign disease should be considered, such as lack of cellular atypia and rare mitotic activity.Prognosis is good.
Keywords: Scalp; Vascular neoplasms; Pathology

REFERENCES

1. Makosa C, Nikolaidoub AJ. Intravascular papillary endothelial hyperplasia (Masson´s tumor) of the oral mucosa.Presentation of two cases and review. Oral Oncology Extra. 2004;40:59-62.
2. Kim DS, Ryu DJ, Oh SH, Lee MG. Intravascular papillary endothelial hyperplasia simulating malignant melanoma. Acta DermVenereol. 2010;90:220-1.
3. Goulart JAVF, Pereira KMA, Galvão HC. Hiperplasia endotelial papilar intravascular oral: uma entidade rara. J Bras Patol Med Lab. 2006; 42: 31-5.
4. Tokyol C, Demir Y, Aktepe F. Cutaneous intravascular papillary endothelial hyperplasia of the hand: a case report and review of the literature. Turk J Pediatr. 2005;47:71-4.
5. Park KK, Won YS, Yang JY, Choi CS, Han KY. Intravascular papillary endothelial hyperplasia (Masson tumor) of the skull: case report and literature review. J Korean Neurosurg Soc. 2012;52:52-4.
6. Fleury LFF Jr, Sanches JA Jr. Primarycutaneous sarcomas. An Bras Dermatol. 2006; 81:207-21.
7. Inoue H, Miyazaki Y, Kikuchi K, Fujinami M, Yoshida N, Ide F, et al. Intravascular papillary endothelial hyperplasia of the oral cavity. J Oral Sci. 2011;53:475-80.
8. Sartore L, Voltan A, Tomat V, Bassetto F, Salmaso R.Masson´s disease in hand surgery: a clinicopathologic study of four cases. J Hand Surg Eur Vol. 2011;36:694-7.

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