■ CASE STUDIES

Oropharyngeal Kaposiform Hemangioendothelioma

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The kaposiform hemangioendothelioma is a very infrequent tumor proceeding from endothelial-derived spindle cells, more often found on the limbs, although peritoneal, retroperitoneal, and sacrum locations are also prevailing. Head and neck are exceptional locations. The kaposiform hemangioendothelioma is almost exclusively found in children and early adolescents, and it is highly associated with the Kasabach-Merritt syndrome and lymphangiomatosis. The main treatment is the tumor surgical removal, including wide margins, plus supporting therapy when Kasabach-Merritt syndrome is linked. We report an isolated oropharyngeal kaposiform hemangioendothelioma in a 19-year old male.

Key words: Kaposiform haemangioendothelioma. Kasabach-Merritt syndrome. Lymphangiomatosis. Capillary hemangioma.

Hemangioendotelioma kaposiforme de orofaringe
El hemangioendotelioma kaposiforme es un tumor raro que procede de células fusiformes de derivación endotelial cuya presentación más frecuente es en las extremidades; es frecuente también su localización peritoneal, retroperitoneal y en el sacro. La presentación en cabeza y cuello es excepcional. El tumor afecta casi exclusivamente a niños y adolescentes jóvenes y se asocia con frecuencia elevada al síndrome de Kasabach-Merritt y a linfangiomatosis. El tratamiento de elección consiste en la extirpación quirúrgica del tumor con amplios márgenes y el tratamiento de soporte en los pacientes con síndrome de Kasabach-Merritt. Presentamos un caso único de hemangioendotelioma kaposiforme localizado en la orofaringe en un varón de 19 años.

Palabras clave: Hemangioendotelioma kaposiforme. Síndrome de Kasabach-Merritt. Linfangiomatosis. Hemangioma capilar.

CASE STUDY

A 19-year-old male attended consultation for a chance finding of a right tonsil neoformation. On examination, a pseudo-polypoid tumouration was observed in the superior pole of the right tonsil bed; it had a smooth surface, was red-wine colour and of medium consistency, with a diameter of approximately 1 cm. The rest of the examination was normal.

Using local anaesthesia, the lesion was biopsied and resected, with pathology findings compatible with kaposiform hemangioendothelioma, as shown un Figures 1 and 2.

With this diagnosis and after reviewing the existing literature it was decided to perform a right tonsillectomy; the sample was tumour-free. The patient remains disease-free after 1 year of follow-up.

The authors declare they have no conflict of interest.

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DISCUSSION

Kaposiform hemangioendothelioma is a very infrequent tumour proceeding from endothelial-derived spindle cells and it has received various names in the literature: haemangioma with characteristics of a pseudo-Kaposi's sarcoma, infantile kaposiform hemangioendothelioma, Kaposi-like hemangioendothelioma, or simply hemangioendothelioma, although the term "kaposiform hemangioendothelioma," first adopted by Zukerberg et al[1] in 1993, has prevailed. The first case described dates from 1971[2] and few cases have been diagnosed since then; its presentation in the head and neck is exceptional.

Kaposiform haemangioendothelioma almost solely affects children and young adolescents, although cases have been diagnosed in adults.[3] It is very frequently associated with Kasabach-Merritt syndrome (thrombocytopenia and consumption coagulopathy)[4] and occasionally lymphangiomatosis.

The most frequent location is in superficial or deep tissue on the limbs,[5] and also in the peritoneal, retroperitoneal, and sacrum area. Retroperitoneal masses are usually extensive non-resectable lesions associated with Kasabach-

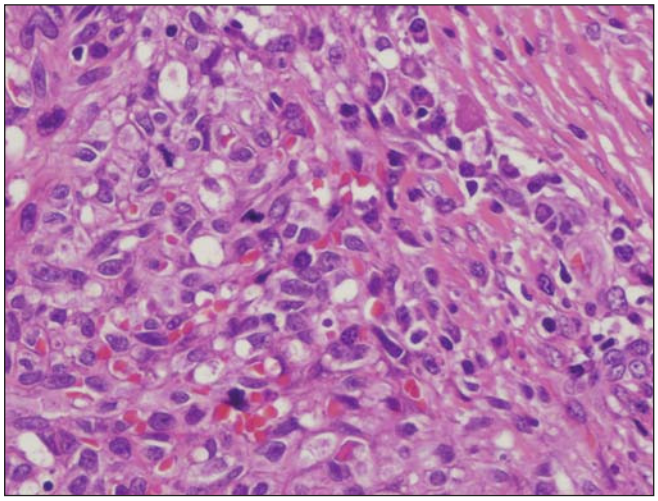

Figure 1. Detail of cellularity. Lengthened nuclei can be seen with discreet or moderate variations in size. There are 2 mitosis figures in the image. Clefts with red blood cells.

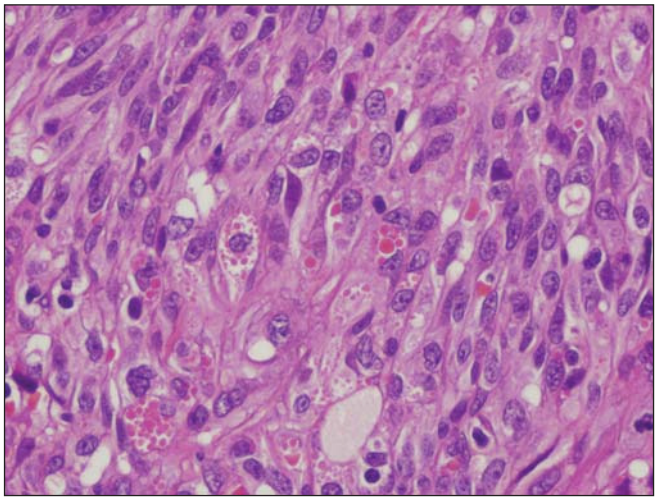

Figure 2. Oval and spindle-like cellularity with notable fasciculation that can be observed in the upper left corner. Presence of prominent hyaline cytoplasmatic globules.

Merritt syndrome, generally leading to the death of the patient.[1,2]

Kaposiform hemangioendothelioma may behave in a locally aggressive way and metastasize to the regional lymph-node glands, although there are no remote metastases. When bone structures are invaded, it is radiographically similar to aggressive fibromatosis and rhabdomyosarcoma.

Head and neck presentation is extremely infrequent: 14 cases have been published in the literature in English. Of these, only 1 was located in the oro-pharynx (soft palate).[6] The others were distributed on the skin of the face and neck or on the temporal bone. Most of these 14 cases were diagnosed before 2 years of age and the onset in all cases was as palpable wine-red or purple lesions in superficial, or deep locations.

Histologically, kaposiform hemangioendothelioma is characterized by combining characteristics typical of Kaposi's sarcoma, hemangioma, hemangioendothelioma, and hemangiopericytoma. Its defining histological characteristic is the lobular growth pattern with irregular nodes infiltrating the soft tissue around the lesion. These nodes are formed by fascicules of endothelial spindle cells which delimit small vascular buttonhole clefts (Figure 1). These clefts contain glomeruloid nests of endothelioid epithelioid cells with cytoplasmic vacuoles, deposits of haemosiderin and PAS diastase positive hyaline globules (Figure 2). Cellular atypia is minimal. Immunohistochemical findings show that the tumour represents a heterogeneous population of immature endothelial cells expressing CD31 and CD34. The nodes may be surrounded by cells with positive reactions for factor VIII.[7]

The treatment of choice consists in the surgical removal of the tumour with wide margins, whenever possible. In non-resectable tumours, vascular embolization was attempted. In these cases, support treatments were also important, especially in patients with Kasabach-Merritt syndrome. Although experience is limited, treatment with steroids, interferon, and various chemotherapy regimes have managed to reduce some tumours.[6,8] There is no consensus among the different authors with respect to the usefulness of radiation therapy.

The 2 cases described in the oro-pharynx (1 case located in the soft palate and the tonsil case described here) were well-localized lesions not associated with Kasabach-Merritt syndrome and their treatment consisted in local resection with margins. No other form of treatment was necessary in these cases. Nonetheless, in view of the scant cases reported, we cannot conclude that cases limited to the oro-pharynx imply a better prognosis.

REFERENCES

1. Zukerberg LR, Nickoloff BJ, Weiss SW. Kaposiform hemangioendothelioma of infancy and childhood. An aggressive neoplasm associated with Kasabach-Merritt syndrome and lymphangiomatosis. Am J Surg Pathol. 1993;17:321-8.
2. Al-Rashid RA. Cyclophosphamide and radiation therapy in the treatment of hemangioendothelioma with disseminated intravascular clotting. Cancer. 1971;27:364-8.
3. Mentzel T, Mazzoleni G, Dei Tos AP, Fletcher CD. Kaposiform hemangioendothelioma in adults: clinicopathologic and immunohistochemical analysis of three cases. Am J Clin Pathol. 1997;108:450-5.
4. Kasabach H, Merritt K. Capillary hemangioma with extensive purpura. Am J Dis Child. 1940;59:1063-70.
5. Lyons LL, North PE, Mac-Moune Lai F, et al. Kaposiform hemangioendothelioma: A study of 33 cases emphasizing its pathologic, immunophenotypic, and biologic uniqueness from juvenile hemangioma. Am J Surg Pathol. 2004;28:559-68.
6. DeFatta RJ, Verret DJ, Adelson RT, et al. Kaposiform hemangioendothelioma: case report and literature review. Laryngoscope. 2005;115:1789-92.
7. Fukunaga M, Ushigome S, Ishikawa E. Kaposiform hemangioendothelioma associates with Kasabach-Merritt syndrome. Histopathology. 1996;28:281-4.
8. Blei F, Karp N, Rofsky N, et al. Successful multinodal therapy for kaposiform hemangioendothelioma complicated by Kasabach-Merritt phenomenon: case report and review of the literature. Pediatr Hematol Oncol. 1998;15:295-305.