

## Case Report

### Kaposiform Hemangioendothelioma of the Skull in a 3 Year Old Boy

Seyed Hamid Madani<sup>1</sup>, Etrat Javadi Rad<sup>2</sup>, Ahmad Faramarzi<sup>3</sup>

1. Dept. of Pathology. Kermanshah University of Medical sciences, Kermanshah, Iran

#### ABSTRACT

Kaposiform hemangioendothelioma (KHE) appears as a single lesion at birth or early infancy in an equal sex ratio. A rare aggressive vascular proliferation has been recognized as a separate entity from other childhood vascular neoplasm. A 3-year-old Iranian boy with a rapidly enlarging mass in his (posterior aspect of skull at the midline) skull was present here. Physical examination revealed a dark-red, firm mass measuring 5 × 4.5 cm in the posterior aspect of skull. No association with Kasabach- Merritt syndrome (KMS) was observed despite its size. Histologically, KHE was composed of infiltrating nodules with slitlike or crescentic vessels that are poorly canalized and lined by spindle endothelium cells. Immunohistochemically, both spindle and epithelioid cells were immunoreactive for CD34 and CD31, while negative for EMA, cytokeratin or S100 protein.  $\alpha$ -SMA were detected in pericytes surrounding spindle cells. Recurrence occurred 2 month after first operation. Wide resection was performed at second operation and the patient was still alive during the 1-year follow- up period.

**Key words:** Hemangioendothelioma, Vascular neoplasms, Immunohistochemistry, Skull

#### Introduction

In the last decade, pathologists began to describe a distinctive vascular tumor, called Kaposiform hemangioendothelioma (KHE) that often was associated with thrombocytopenia. (1-3). The name was coined for its unique morphology, characterized by a Kaposi sarcoma- like spindle growth pattern (4, 5). This rare locally aggressive vascular neoplasm has a predilection for the trunk, extremities, retroperitoneum, mediastinum and deeper soft tissue. Lesions involving the skull were rarely described (1,4,

6). Herein, we report extremely unusual case of KHE occurring in the posterior aspect of the skull, with special reference to immunohistochemical study.

#### Case Report

A 3- year- old male child presented with a rapidly enlarging mass in his posterior aspect of skull at the midline, which was first noticed 2 month previously. Physical examination revealed ill-defined, reddish, firm mass measuring 5×4.5cm. There were no

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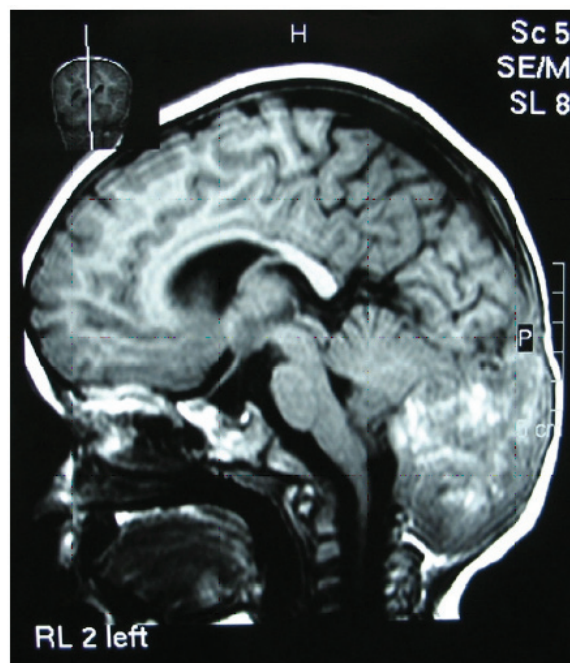
Received: 4 September 2008

Accepted: 20 January 2009

Address communications to: Dr Seyed Hamid Madani, Department of pathology. Kermanshah University of Medical sciences, Kermanshah, Iran

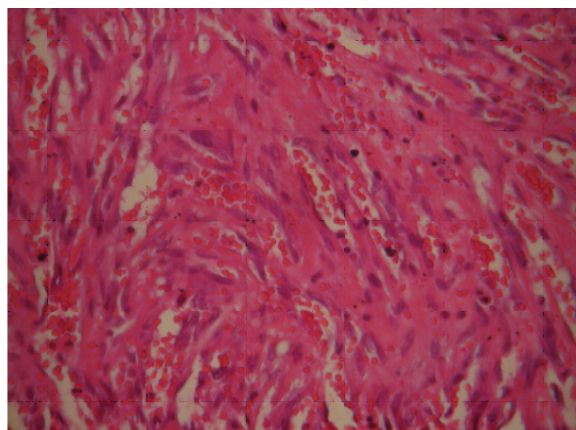
Email: shmmadani@yahoo.com

neurologic signs and no lymphadenopathy in the head and neck region. Laboratory findings, including complete blood count, blood biochemistry, fibrinogen and fibrin-split products and urine analysis were all within normal limits. Imaging demonstrated a diffuse, enhancing, T2 hyperintense lesion with ill-defined margins and bone destruction (Fig. 1).



**Fig.1:** Diffuse,enhancing lesion with ill-defined margins

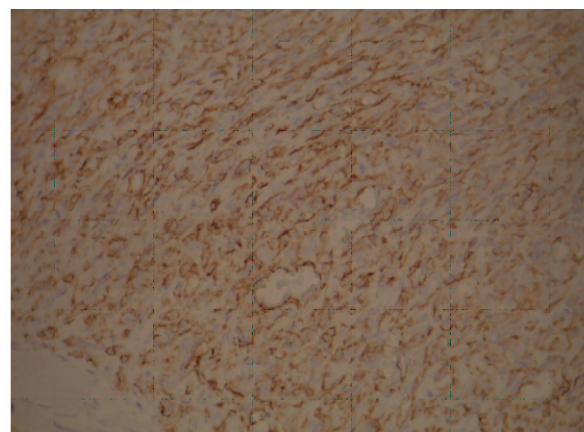
Morphologically, the tumor consists of dense spindle cells with a nodular growth pattern as small round or slitlike vascular channels (Fig. 2). The spindle tumor cells may appear plump with vesicular nuclei or small with dark nuclei but without cytologic atypia. Pericytic cells had rather similar cytologic features.



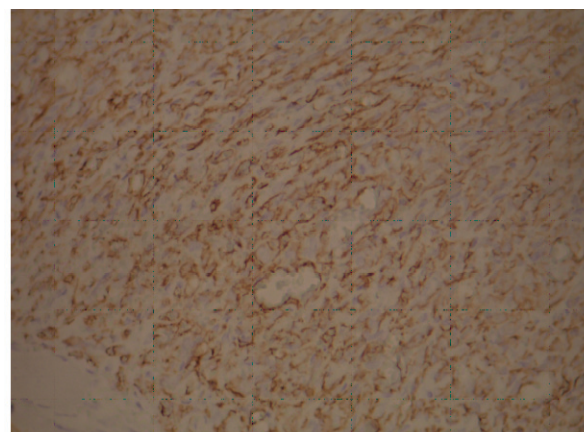
**Fig. 2:** Spindle cell proliferation with slitlike vascular channels(H&E staining x400)

Mitosis was not seen or rare. Hemosiderin was present in endothelial and pericytic cells. Also, there was no encapsulation. The solid spindle cells areas associated with slitlike lumen containing red blood cells were reminiscent of kaposi's sarcoma. But the gaped lumen and nodular growth pattern, permitted differentiation with Kaposi's sarcoma.

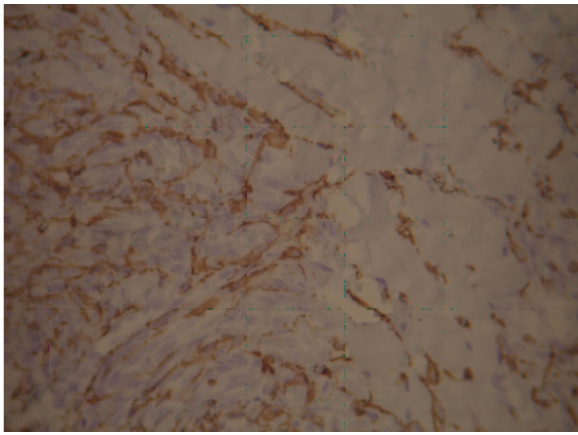
Immunohistochemical studies showed that the tumor cells, whether epithelioid or spindled strongly expressed CD<sub>31</sub>(1:200 ,DAKO) and CD<sub>34</sub> (1:500 ,DAKO) but not factor VIII related antigen (1:200 ,DAKO), epithelial membrane antigen (EMA,1:200 ,DAKO), cytokeratin (AE<sub>1</sub>/AE<sub>3</sub>,1:50 ,DAKO) or S100 protein (1:100, DAKO). Spindle cells surrounding endothelial lining and epithelioid cells were immunoreactive to alpha-smooth muscle actin ( $\alpha$ -SMA, 1:400, DAKO) (Fig. 3, 4, 5). Two months post- operatively, however, a 7 cm recurrent lesion appeared at the surgical site, with similar clinical appearance as the primary tumor. This time, the recurrent tumor widely was excised, and confirmed on pathology. One year after the initial examination, the boy was alive and well.



**Fig. 3 :** CD 31 expression of the tumor cells



**Fig.4:** CD 34 expression of the tumor cells



**Fig. 5:** Spindle cells surrounding endothelial lining and epithelioid cells were immunoreactive to alpha-smooth muscle actin

### Discussion

Kaposiform hemangioendothelioma is a rare locally aggressive vascular tumor of the skin, deep soft tissue, and bone in children, of intermediate malignant potential in the latest WHO classification of soft tissue tumor (3-5, 7), characterized by infiltrating nodules and sheets of spindle cells, and unmistakable resemblance to Kaposi's sarcoma (7-9).

It is also known as a subgroup of "malignant endovascular papillary angioendothelioma or Dabska tumor (10).

KHE involving head and neck is very rare, often presenting as a single mass, which may infiltrates peripheral tissue (4, 11-13). While the possibility of KHE, may be considered on clinical and radiological grounds, tissue diagnosis is required, but biopsy is now uncommonly performed in cases associated with KMS because of the potential for hemorrhage (2, 6, 14).

Despite its unusual site, the tumor in our patient showed rather typical morphology of KHE, with a deeply infiltrative nodular growth, spindle cells with slitlike vascular lumen (14, 15).

In our patient, both epithelioid and spindle tumor cells expressed endothelial markers (CD31 and CD<sub>34</sub>) but not factor VIII-related antigen.  $\alpha$ -SMA was expressed by pericytes that outlines tumor spindle cells, but not by these spindle cells, results consistent with the reported observations (4,15).

The precise biologic potential and pathogenesis of KHE remains uncertain. It was classified as borderline malignant because of its locally aggressive behavior, causing significant morbidity and mortality as a result of the compression and invasion of surrounding

structures.

Prognosis in this tumor was mainly related to the size, anatomic site, and extent of the neoplasm (4). In the present case, limited excision was not effective, resulted in recurrence, which required wide excision.

KHE is extremely rare, and the skull is an unusual location (4, 6). Our report demonstrates that KHE should be considered in the differential diagnosis of a rapidly growing vascular lesion.

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