Kaposiform Hemangioendothelioma Arising From the Maxillary Sinus: A Case Report

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Kaposiform hemangioendothelioma is a very infrequent, locally aggressive vascular neoplasm, characterized by fascicular spindle cell proliferation. It occurs almost exclusively in infants and adolescents, and is often associated with Kasabach-Merritt phenomenon. The tumor is predominantly located subcutaneously or in the deep soft tissue of the extremities and trunk, peritoneum, or retroperitoneum. However, this tumor can sometimes be located on the head and neck. We report a case of kaposiform hemangioendothelioma of the maxillary sinus in a 4-month-old female infant presenting with cheek swelling, thrombocytopenia, anemia, and disseminated intravascular coagulation. Sinus computed tomography presented an enhancing, bone-destructing tumor. Magnetic resonance images showed an uncommon appearance as mostly low signal intensity on T2-weighted images.

Key Words: head and neck, kaposiform hemangioendothelioma, Kasabach-Merritt phenomenon, maxillary sinus
(*Kaohsiung J Med Sci* 2010;26:154–7)

Kaposiform hemangioendothelioma (KHE) is a very infrequent tumor found predominantly in infants and adolescents with intermediate malignant and locally aggressive characteristics [1]. It is more often located in the limbs and is uncommon in the head and neck regions [2], as well as rarely involving the nose and paranasal sinuses [3]. Radiologically, hemangioendothelioma shows as hyperintense on T2-weighted images or inversion-recovery sequences and when well-enhanced, and it resembles a hemangioma. Hemangioendothelioma may contain flow voids of vascular structures or hemorrhagic foci [1]. We report a case of KHE of the maxillary sinus with unusual imaging manifestations.

Received: Jun 15, 2009 Accepted: Jul 6, 2009
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Case Presentation

A 4-month-old female infant had progressive swelling of the right cheek without associated local heat, tenderness, or redness for 2 weeks. In addition, swelling of the right soft palate and upper gingiva, nasal obstruction, sclera injection, and ecchymosis of the lower eyelids were also depicted on physical examination. She was not hospitalized for other medical or surgical disease and received vaccination as scheduled. Findings of laboratory studies included thrombocytopenia, consumption coagulopathy, and microangiopathic hemolysis.

Computed tomography and magnetic resonance imaging were arranged to determine the cause of swelling of the right cheek. Sinus computed tomography showed an isodense to hyperdense mass mainly in the right maxillary sinus with bone destruction, the right middle cranial fossa, and right orbital extension (Figure 1). Magnetic resonance imaging revealed that the mass was markedly hypointense on fat-suppressed T2-weighted sequences, and was slightly hypointense

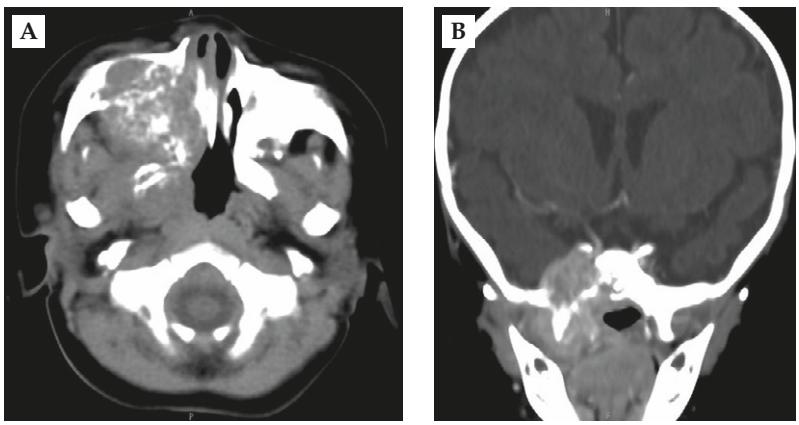

Figure 1. *(A) Axial non-enhanced computed tomographic image showing a soft tissue mass (arrowhead) in the right maxillary sinus with adjacent bony destruction. (B) Coronal contrast-enhanced computed tomography shows the enhanced soft tissue mass (arrowhead) involving the right middle cranial fossa.*

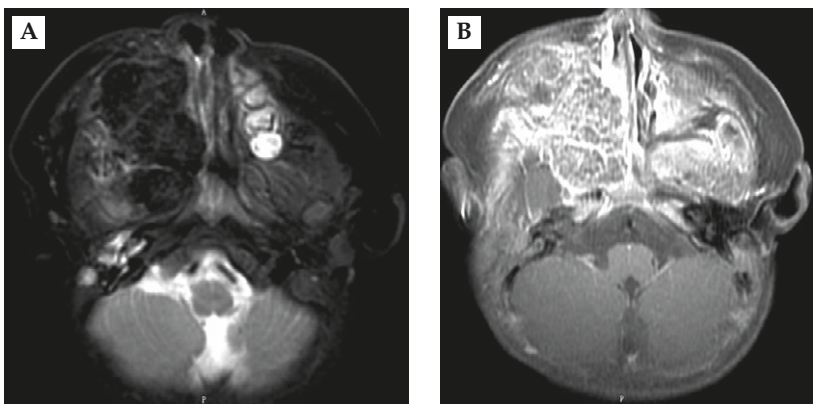

Figure 2. *(A) Axial T2-weighted image demonstrating an extremely hypointense expansile mass in the right maxillary sinus. (B) Gadolinium-enhanced T1-weighted axial image demonstrating a peculiar reticular enhancing tumor without definite vascular structure within the mass.*

on T1-weighted sequences with moderate peculiar reticular enhancement after intravenous administration of contrast material (Figure 2). There was no imaging evidence of prominent vascularity. A biopsy was performed and microscopic examination showed KHE with spindle cell proliferation with fascicular growth interspersed with slit-like vascular lumina containing numerous red blood cells. Nuclear atypia and mitotic figures were inconspicuous. Immunohistochemical study demonstrated CD34+ and CK– for the tumor, which confirmed the vascular nature (Figure 3).

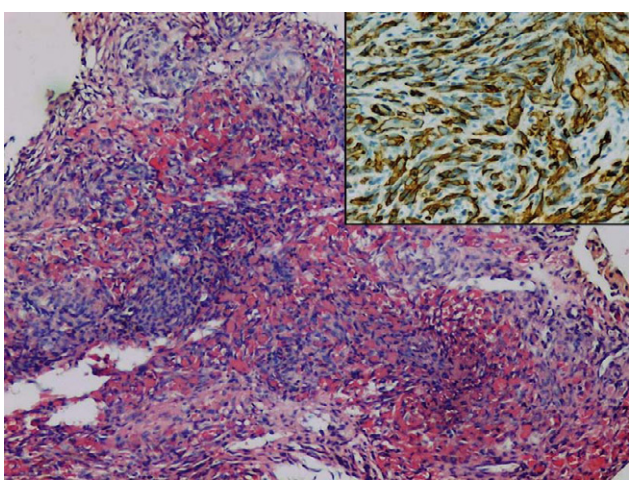

Figure 3. *Microscopy shows spindle cell proliferation with fascicular growth interspersed with slit-like vascular lumina containing numerous red blood cells (original magnification, 100×). The spindle cells are labeled with the endothelial cell marker CD34 (insert; original magnification, 400×).*

Discussion

KHE is a rare vascular tumor that occurs almost exclusively in children. Only a few cases have been reported in the literature since the first described case

in 1971 [2]. Nearly 75% of cases present as cutaneous lesions of trunk and limbs and the second most common location (18%) is in the retroperitoneum [4]. Cases in the head and neck regions are uncommon; we found only one report of a KHE in the ethmoid sinus of an 8-year-old girl [5]. We present here KHE of the maxillary sinus in an infant.

KHE is classified as an intermediate malignant vascular neoplasm [1] due to locally aggressive behavior but without distant metastasis, and can present as multifocal lesions [6]. In contrast to infantile hemangiomas, there is no tendency of spontaneous regression for KHE.

Radiologically, KHE can present as an expansile osteolytic lesion with irregular margins and extension into the adjacent soft tissue. Hemorrhagic foci and flow voids of vascular structures may be seen [1]. KHE typically has a well-enhancing soft tissue component with magnetic resonance hyperintensity on T2-weighted images and hypointensity on T1-weighted images compared with muscle [7]. Unlike these characteristic imaging appearances, our case showed mostly extremely low signal intensity on fat-suppressed T2-weighted images and irregular enhancing margins. Histologically, KHE consists of infiltrative multilobular spindle cell proliferation with slit-like vascular spaces resembling capillary hemangioma and Kaposi's sarcoma [6]. Differentiation from the latter two is made by the presence of well-formed spindle cell fascicles and the absence of periodic-acid-Schiff-positive globules [1]. Lesions demonstrate low or no mitotic appearance and minimal cellular atypia in the endothelial cells. Immunohistochemically, there is a heterogeneous population of immature endothelial cells expressing CD31 and CD34. Positive reactions for factor VIII or alpha smooth muscle actin also can be detected [5].

KHE is frequently associated with Kasabach–Merritt phenomenon (KMP) including thrombocytopenia, consumption coagulopathy, and microangiopathic hemolysis, especially in younger patients and patients with larger vascular tumors [8]. This association is known as a poor prognostic predictor. Another feature of KHE is the involvement of adjacent bony structures [8]. In our case, there was destruction of the maxillary bone, as well as elevation of D-dimers, and low platelet and red blood cell counts. More than 90% of cases with KMP are secondary to KHE [7].

It is important to consider the diagnosis of KHE when KMP is present, even in patients with an uncommon location and atypical imaging findings.

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上頜竇卡波西樣血管內皮細胞瘤：病例報告

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卡波西樣血管內皮細胞瘤是一種非常罕見的，具局部侵犯性的血管腫瘤，其特徵為束狀梭形細胞增生。它幾乎全部發生在嬰幼兒和青少年，經常合併有卡 - 梅綜合徵。腫瘤主要位於皮下或深部軟組織的四肢和軀幹，腹腔或後腹膜腔。然而，頭部和頸部是例外。我們報告一例上頜竇的卡波西樣血管內皮細胞瘤在 4 個月大的女嬰呈現臉頰腫脹，血小板減少，貧血，瀰漫性血管內凝血。鼻竇電腦斷層呈現一個有顯影的骨破壞腫瘤。而磁共振成像顯示少見地在 T2 加權像大部分以低信號強度作為表現。

關鍵詞：頭部和頸部，卡波西樣血管內皮細胞瘤，卡 - 梅綜合徵，上頜竇

（高雄醫誌 2010;26:154–7）

收文日期：98 年 6 月 15 日

接受刊載：98 年 7 月 6 日

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