A case of head and neck kaposiform hemangioendothelioma simulating a malignancy on imaging

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Introduction
Kaposiform hemangioendothelioma (KH) is a locally aggressive endothelial-derived spindle cell neoplasm that occurs almost exclusively in infants and adolescents [1]. We present the case of KH in a 1-year-old girl that involved the right temporal and occipital bones, the temporomandibular joint, the mandibular condyle, and atlas. The imaging findings and nature of this mass suggested malignancy in what proved to be a benign lesion.

Case report
A 1-year-old girl presented with a mass behind the right ear and over the mastoid bone. The patient had been born with a macular discoloration of the right temple and had a history of ear infections earlier in her life. At the time of admission, the patient presented with an erythematous and tender lesion with associated fever. The mass measured 2 cm × 2 cm, and felt cystic in nature. Antibiotic treatment was given without success. The patient was subsequently referred for a surgical excision of the mass with pre- and post-antibiotic therapy. Her complete blood cell count showed a decreased platelet count and the coagulation study was normal including fibrinogen, but the fibrin-split products were very elevated [findings consistent with Kasabach-Merritt phenomenon (KM)].

Under general anesthesia, an incision was made and a hemangioma-like lesion with extensive vascularity was noted in the muscular compartment of the muscles of the scalp, with the mass eroding the adjacent bone. A portion of the mass was removed and sent to pathology for tissue diagnosis. The pathology review demonstrated infiltrating or interconnecting sheets and nodules of slender endothelial cells lining slit-like vessels. The final diagnosis confirmed an incompletely resected KH.

A postbiopsy computerized tomograph of the temporal bones was then obtained and demonstrated abnormal enhancing soft tissue within the tissue superficial to the right temporal bone, posterior to the auricle (Fig. 1a,b). The mass extended into and partially destroyed the temporal bone at the level of the external auditory canal and tympanic cavity with multiple areas of erosion, including both sides of the temporomandibular joint. There was evidence of cortical destruction involving the mandibular condyle and deep on the medial surface of the petrous portion of the temporal bone just below the internal auditory canal. Erosion was also seen superficially within the temporal bone in the mastoid region (Fig. 1c). Cortical destruction was seen within the squamosal portion of the temporal bone and occipital bones in the posterior fossa. Lytic foci were noted in the right lateral mass of C1 vertebra. Multiple lymph nodes were noted in the deep and posterior cervical region bilaterally, the submandibular region, and the submental region which were most likely reactive in nature (Fig. 1a). Based on the imaging findings, the diagnosis entertained included rhabdomyosarcoma and aggressive fibromatosis. The final pathologic diagnosis was KH. The patient was discharged on oral prednisone. Simultaneous treatment with vincristine was also considered for
Fig. 1  a Axial contrast-enhanced CT image demonstrates a right soft-tissue mass with subcutaneous infiltrating changes. Bilateral adenopathy is visualized at levels Ib and II. b Axial contrast-enhanced CT image demonstrates the superficial component of the soft-tissue mass extending along the subcutaneous tissue adjacent to the temporal bone, posterior to the auricle. c Axial nonenhanced CT with bone window through the right temporal bone illustrates the soft-tissue mass eroding into the temporal bone and involving the vestibule, facial nerve canal, and outer cortex of the mastoid. The mandibular condyle is also eroded by the mass.

treatment. The patient was discharged in good health and is being followed up at regular intervals.

Discussion

KH is a rare, locally aggressive, endothelial-derived spindle cell neoplasm that occurs exclusively in the pediatric age group. It grows rapidly with focal extension into the adjacent skin, soft tissue, and bone [1]. Previous reports emphasize the predominant location in the peritoneum and retroperitoneum [2]. Cases of KH have been reported occurring within the lower and upper extremities. There has been documentation of a KH enlarging in an upper extremity in an 18-month-old child that was diagnosed at 6 years of age [3]. The cervicofacial region, trunk and shoulder, and even the scalp have been involved in previous cases. Often this vascular tumor is associated with KM syndrome [2]. The KM “syndrome” consists of thrombocytopenia, microangiopathic hemolytic anemia, and localized consumption coagulopathy. Previously, this phenomenon was most likely referred to as “platelet trapping hemangioma” KM syndrome. Characteristic features of this entity paralleled those of KM and consisted of an increased capillary bed with thrombocytopenia, often severe. These patients were often dependent on platelet transfusions. Patients with tumors on the trunk or proximal extremities have a higher incidence of lymphangiomatosis and KM, whereas those tumors limited to distal extremities are benign and more easily cured with surgical excision [1]. The KH lesion is known to be nonmetastasizing [1]. Prognosis depends on the extent and location of the disease. Tumors localized to the skin are benign whereas those that invade visceras have been fatal. Treatment is usually wide local excision and supportive therapy for associated symptoms.

Differentiation from Kaposi’s sarcoma is made by absence of periodic-acid-Schiff (PAS)-positive globules whereas differentiation from capillary hemangioma is made on the basis of well-formed spindle cell fascicles of KH [4]. Histologically, the tumors are characterized by infiltrating, interconnecting sheets of irregular nodules of slender endothelial cells lining crescentic or slit-like vessels and, less commonly, rounded capillary-type vessels [2].

The current case was confusing since it presented in the first decade of life with a locally aggressive soft-tissue mass in the head and neck. Bone destruction was evident on imaging studies, a rare occurrence with this tumor. The patient also had Kasabach-Merritt phenomenon. The discoloration of the mass first suggested a clinical diagnosis of a hemangioma or hemangiomatosis, whereas the radiological picture suggested a differential diagnosis of rhabdomyosarcoma or aggressive fibromatosis.