Pediatric Facial Kaposiform Hemangioendothelioma: A Case Report and Review of the Literature.

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Abstract

Objectives: To discuss facial kaposiform hemangioendothelioma (KH) and compare KH to tufted angioma (TA) by illustrating a rare pediatric case.

Study Design: Case report and literature review.

Methods: We report a unique case of a pediatric patient presenting with facial kaposiform hemangioendothelioma.

Results: An 11-month-old male with a history of spontaneously resolved nasolacrimal duct stenosis presented for evaluation of left facial swelling for 9 months. Six months prior to admission, a mass was identified on the left cheek which had progressively increased in size over the past 2 months. On exam, he was found to have a firm, mobile, non-tender mass just inferior to the left zygomatic arch. Ultrasound revealed a vascularized solid lesion measuring 12 mm x 15 mm x 16 mm limited to the superficial soft tissues. MRI showed a multi-lobular mass centered within the left masseter with increased T2 signal infiltrating into surrounding subcutaneous tissue. The patient subsequently underwent ultrasound-guided core biopsy. Initial histology was consistent with TA; however, upon further review, a diagnosis of KH was favored. The tumor was managed with low-dose aspirin and observation.

Conclusions: Cutaneous KH most commonly presents in the extremities and rarely in the cervicofacial region. This is one of the first reports of facial presentation reported in the pediatric population. KH and TA lie on the same spectrum, and differentiating between the two can be difficult as demonstrated in this case. Early and proper identification of this tumor is crucial given the concern for complication with Kasabach-Merritt phenomenon.

Introduction

- Tufted angioma (TA) and kaposiform hemangioendothelioma (KH) are vascular tumors with benign and locally aggressive growth potential, respectively.[1]
- These tumors are very rare with one study reporting a prevalence of approximately 0.91 cases per 100,000 children for KH.[2]
- Importantly, these tumors have the unique potential of developing into Kasabach-Merritt phenomenon (KMP), a life-threatening thrombocytopenia with mild to moderate coagulopathy due to intralesional platelet trapping.[3]
- KMP has been reported to occur in approximately 70% of KH and 10% of TA, and is more likely in larger, deeper lesions.[1-3]
- KH and TA most commonly present with cutaneous manifestations favoring the extremities.[2] TA has also been reported to present on the neck and upper trunk.[4,5]
- Facial presentation of KH is extremely rare, with only a few cases reported in the literature.

Methods and Materials

- We report a unique case of pediatric facial kaposiform hemangioendothelioma treated with low-dose aspirin and observation at our center.
- A comprehensive literature review was also performed using the MEDLINE database via PubMed.

Results

- An 11 month-old male presented with a 2-month history of enlarging left facial swelling and intermittent fevers.
- On exam, a firm, mobile, non-tender mass without overlying erythema or discoloration was identified just inferior to the left zygoma.
- The patient was otherwise healthy and baseline labs, including platelets, were within normal limits.
- Ultrasound showed a 16 mm solid vascular lesion of the left cheek. MRI with contrast revealed an enhancing soft tissue mass with indistinct margins centered within the left masseter muscle.

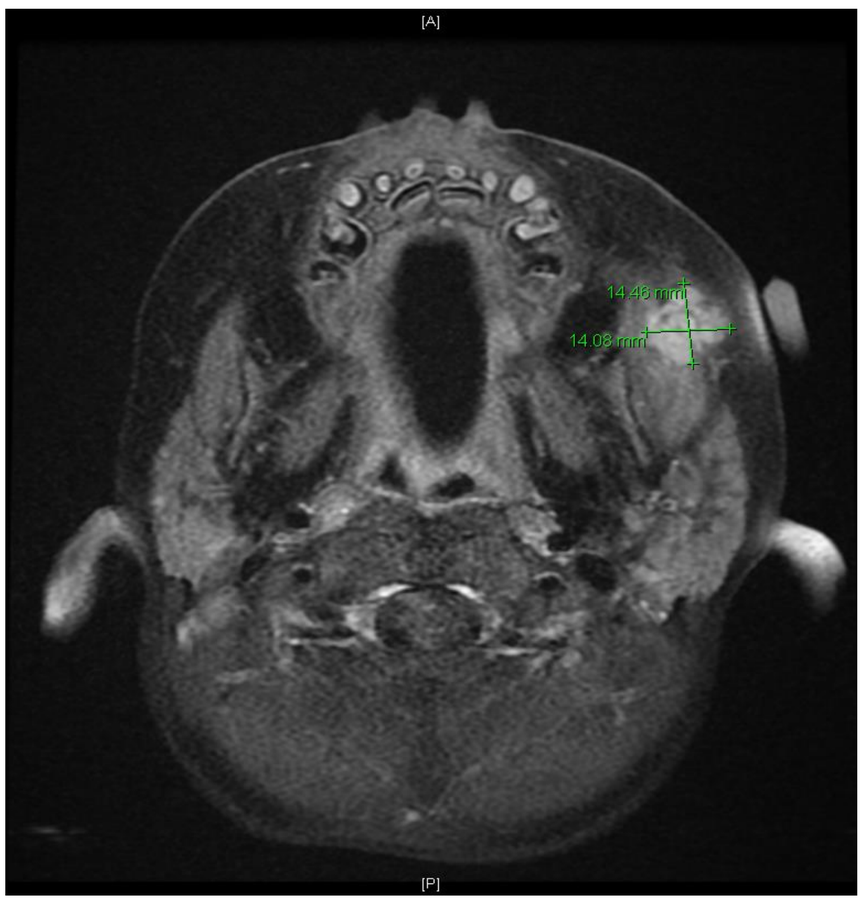

Figure 1. T2-weighted MRI revealed a homogenously enhancing 16 mm x 13 mm x 11 mm mass with indistinct margins centered within the left masseter muscle and protruding into the subcutaneous tissues laterally.

Results, continued

- Ultrasound-guided biopsy was performed and histological analysis revealed fibrovascular tissue containing compact nests of variably sized and ill-defined vascular channels.
- Immunohistochemistry staining showed the endothelial cells of the lesion were diffusely reactive for CD34 and CD31, partially reactive for D2-40, and negative for GLUT-1.
- Cytogenetic analysis was unable to be performed as there were no dividing cells present in the tissue sample.
- While a definitive diagnosis was unable to be achieved, KH was favored over TA due to location of the lesion within subcutaneous fat, solid and infiltrative components on MRI, and normal overlying skin on exam.
- The case was discussed at a vascular anomalies conference, and the decision was made to treat with low-dose aspirin and perform serial MRI imaging.

Discussion

- In the largest cohort study discovered in the literature, cervicofacial involvement was found to be the least common anatomic location for pediatric KH.[2]
- As seen in this patient, facial presentation of KH without classically associated dermatologic changes such as cutaneous discoloration is an extremely rare phenomenon[2] and presents a significant diagnostic challenge.
- TA and KH appear to lie on the same spectrum, but differentiating between the two may be of benefit as KH has been shown to be associated with a higher risk of KMP than TA.[1-3]
- One 2006 study by Shimizu et al. suggested that D2-40 immunohistochemistry staining patterns may be used to distinguish between KH and TA.[6]
- While numerous treatment options have been described including both medical and surgical management, evidence-based guidelines are limited.[1]

Conclusions

- This is one of the few cases of facial KH reported in the pediatric population.
- Although rare, KH should be considered in the differential for facial masses, including those without suggestive dermatologic changes.
- Identification of KH and TA can be difficult, especially with atypical presentations. However, the risk of progression to KMP warrants early diagnosis.

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