Composite Hemangioendothelioma: A Rare Presentation in the Pediatric Scalp and Review of Literature

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Abstract

Composite Hemangioendothelioma (CH) is rare vascular neoplasm's with a complex mixture of benign and malignant components. The clinical course in CH is variable; treatment is typically surgical, though optimal treatment is debatable. First described in 2000, less than 40 cases, nearly all adults, are described in the literature. We describe the presentation and management of CH in the scalp of an adolescent female. A review of the published pediatric cases of CH identified in the head and neck region is discussed. We conclude that while pediatric cutaneous neoplasms are infrequently malignant, a thoughtful approach to the surgical management is necessary.

Keywords: Composite hemangioendothelioma; Scalp mass; Pediatric malignancy

Introduction

Composite Hemangioendothelioma (CH) are rare vascular neoplasms consisting of a mixture of benign, low-grade malignant, and malignant vascular components [1,2]. CH neoplasms are categorized in between hemangiomas (completely benign tumors) and angiosarcomas (highly malignant neoplasm’s) [3]. Correct diagnosis is important to selecting suitable treatment as the clinical course varies considerably from prognostically less favorable vascular tumors. A scalp CH lesion affecting a pediatric patient has not been reported to date. We present a case of adolescent female who developed a CH at her Bone-Anchored Hearing Aid (BAHA) implant site. This case was deemed exempt by the Institutional Review Board (IRB) at Nemours Children’s Hospital.

Case Report

A 13-year-old African American female, with otherwise non-contributory medical and family history, was seen in our office with a right parietal scalp mass (Figure 1). She had long standing right sided conductive hearing loss, and in past years had undergone multiple otologic procedures including exploratory tympanostomies and BAHA abutment surgeries at outside institutions. Approximately two years prior to our visit, the BAHA attract system was implanted. This system had worked well until her family noticed a tender mass growing, believed to be a keloid, at the implant site preventing external device use. Examination revealed a multi-lobed nodular growth estimated 2.5 cm x 2 cm in the post auricular scalp that was tender to palpation but without fluctuance. A Computed Tomography (CT) scan of the temporal bone demonstrated asymmetric fullness of the parietal soft tissue in the region of interest (in addition to the middle ear anomalies necessitating the BAHA). Conservative management with oral antibiotics and steroids did not improve her symptoms, therefore surgical intervention was recommended. She underwent wide local excision of the lesion with removal of the BAHA magnet. At the time of surgery, an approximately 2.5 cm x 2 cm nodular, friable post auricular mass was noted. The lesion was at the inferior margin of a wider region of nodularity and scarring measuring 4 cm x 6 cm, and extended down to the level of periosteum. Closure thereafter was accomplished with a local advancement flap.

A diagnosis of composite Hemangioendothelioma was made with positive margins; this diagnosis was confirmed with a secondary institutional review. The specimen was composed of three components, morphologically consistent with venous malformation, Epitheloid Hemangioendothelioma, and retiform hemangioendothelioma (Figure 2). Although a large component of the tumor had the morphology of epitheloid hemangioma, the WWTR1-CAMT1
Discussion

First described by Nayler et al. [4] in 2000, this pathologic entity is composed of varying combinations of Hemangioendothelioma, principally epitheloid, retiform, and spindle cell subtypes. Composite hemangioendothelioma are rare, with less than 40 cases reported in the literature [2,5]. Classically described as an adult tumor, only a handful of pediatric cases have been reported [6]. The majority of reported tumors occur in the extremities; only 10 cases to date in the head and neck region, and none have been reported as a pediatric scalp lesion (Table 1A) [7,8]. There is a slight female predominance [2]. The exact pathologic etiology is not entirely clear. In some cases, there appears to be predilection for cutaneous sites with a history of some insult, having been reported to occur at prior trauma, surgery, and radiation sites (Table 1B) [3,5,9].

The differential diagnosis for pigmented scalp lesions includes congenital nevus, atypical or acquired nevi, Spitz nevus, halo and radiation sites (Table 1B) [3,5,9]. The clinical course for these vascular neoplasms is variable, and is most likely related to the presence of the epitheloid hemangioendothelioma component, as it is the most aggressive [9]. No cases of death attributed to composite hemangioendothelioma have been published. Recurrence, not uncommon, can occur in more than 50% of patients ranging from 18 months to 10 years post treatment. Regional and distant metastasis rates of 13% and 5% respectively have been reported. To date, there has been no reported history of metastasis in children [2]. Surgical excision with wide margins is the typical treatment, though optimal treatment is debatable. Adjuvant lymphadenectomy is appropriate if regional metastasis is present, but is not advocated empirically in the absence of disease. Adjuvant radiation or chemotherapy has been discussed for distant metastasis or disease recurrence [2].

At the time of this review, only 11 cases in head and neck region (including our case) have been reported in the English language [5]. Our case marks only the third case identifying CH in the scalp, and the first in the scalp a pediatric patient (Table 1A). To date, only one other case of head and neck CH in a child has been reported; this neoplasm was located in the hypopharynx of a 15-year-old [6]. She presented with three months of hemoptysis and globus sensation, and was noted to have a bulging ulcerative mass on laryngoscopic exam.

<table>
<thead>
<tr>
<th>Case</th>
<th>Reference</th>
<th>Age/ Sex</th>
<th>Site</th>
<th>Lesion size</th>
<th>Metastasis</th>
<th>Histology including EHE</th>
<th>Management/ Follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Liu et al. [7]</td>
<td>24 y/F</td>
<td>Scalp (left temporoparietal)</td>
<td>3 cm patch with 1.5 cm nodule</td>
<td>No</td>
<td>Yes</td>
<td>Surgical excision. Skin graft closure. NR 1 year</td>
</tr>
<tr>
<td>2</td>
<td>Mahmoudizad et al. [8]</td>
<td>68 y/M</td>
<td>Scalp (vertex)</td>
<td>6.3 cm multilobulated nodule with adjacent papules</td>
<td>Yes; right posterior neck</td>
<td>Yes</td>
<td>Poor surgical candidate; radiation therapy with tumor shrinkage. Loss to follow up</td>
</tr>
<tr>
<td>3</td>
<td>Current case</td>
<td>13y/F</td>
<td>Scalp (right parietal)</td>
<td>4 × 6 cm; prominent 2.5 cm nodule</td>
<td>No</td>
<td>Yes</td>
<td>Surgical excision. Skin graft closure. NR to date</td>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>Tsai et al [9]</td>
<td>15y/F</td>
<td>Hypopharynx (left pyriform sinus)</td>
<td>5.5 × 4.2 × 3.2 cm bulging ulcerative mass</td>
<td>Spindle cell hemangioma and angiosarcoma-like</td>
<td>Primary resection. NR followed to 18 months</td>
</tr>
<tr>
<td>2</td>
<td>Current case</td>
<td>13y/F</td>
<td>Scalp (right parietal)</td>
<td>4 × 6 cm; prominent 2.5 cm nodule</td>
<td>Venous malformation, EHE, and retiform hemangioendothelioma.</td>
<td>Surgical excision. Skin graft closure. NR to date</td>
</tr>
</tbody>
</table>

EHE: Epitheloid Hemangioendothelioma; NR: No Recurrence

Figure 1: Right post auricular lesion.

Figure 2: This composite hemangioendothelioma contains many components of the vascular tumors. Two key components shown here are epitheloid (A) and retiform (B) hemangioendothelioma like areas.
Imaging studies (CT and MRI) revealed a 5 cm mass involving left pyriform sinus and glottis without evidence of regional metastasis. She was treated with primary surgical resection, and remained free of recurrence through at least 18 months of follow up.

Similar to our case, in the previously reported scalp cases, the lesions were sizable, at least 3 cm in diameter, and contained a nodular component. Interestingly, their management was different. Regional metastasis was present in one of the two cases, a 68-year-old male with 10 months of pain and pruritis (Table 1A) [7]. He underwent radiation therapy treatment, being deemed a poor surgical candidate secondary to co morbidities, but was unfortunately lost to follow up. The other case, a 24-year-old female with several months of alopecia, was without metastasis and was treated with surgical resection. She had no evidence of recurrence at one year of follow up (Table 1A) [8]. In light of these experiences, similar to other parts of the body, primary surgical resection is the preferred modality of choice for CH management in the head and neck region.

**Conclusion**

Our case extends the known presentation for composite hemangioendothelioma and illustrates the variable nature of exhibition. Clinicians should be aware of the diagnosis. While pediatric cutaneous neoplasms are infrequently malignant, a thoughtful approach to the surgical management is necessary. In the case of a rare tumor, multidisciplinary discussion and close clinical follow up are advisable. In the case of composite hemangioendothelioma, early surgical intervention is recommended for optimal outcomes.

**References**