American Journal of Otolaryngology and Head and Neck Surgery

Case Report
Published: 19 Apr, 2018

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

Tyler A Janz[1], Aftab Patni[1], Craig Johnson[2], Lili Miles[3] and Cedric V Pritchett[4]*

[1]University of Central Florida, USA

[2]Department of Radiology, Nemours Children's Hospital, USA

[3]Department of Pathology, Nemours Children's Hospital, USA

[4]Department of Surgery, Nemours Children's Hospital, USA

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 × 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 × 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

OPEN ACCESS

Correspondence:

Tyler A Janz, College of Medicine, University of Central Florida, 6850 Lake Nona Blvd, Orlando, Florida, 32827, USA,

E-mail: tjanz@knights.ucf.edu

Received Date: *01 Mar 2018*

Accepted Date: *17 Apr 2018*

Published Date: *20 Apr 2018*

Citation:

Janz TA, Patni A, Johnson C, Miles L, Pritchett CV. Composite Hemangioendothelioma: A Rare Presentation in the Pediatric Scalp and Review of Literature. Am J Otolaryngol Head Neck Surg. 2018; 1(1): 1001.

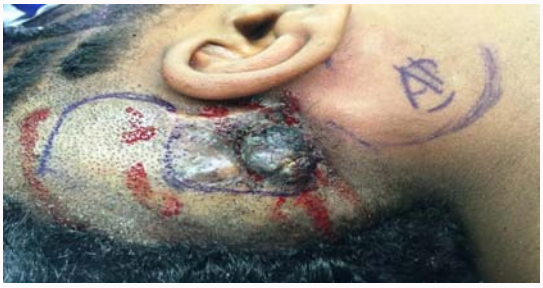

Figure 1: Right post auricular lesion.

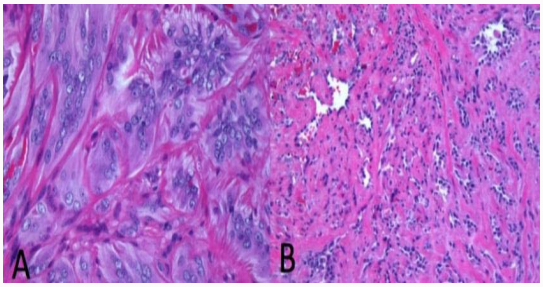

Figure 2: This composite hemangioendothelioma contains many components of the vascular tumors. Two key components shown here are epitheloid (A) and retiform (B) hemangioendothelioma like areas.

fusion gene test was negative, supporting the diagnosis of CH, especially with the presence of the aforementioned other two components. The patient was presented at both the Vascular Malformations and Multidisciplinary Tumor Board conferences. Given the unpredictable history of distant metastasis, she underwent CT imaging of the chest, abdomen, and pelvis. No evidence to suggest distant spread on the CT scan was noted. MR of the brain and neck was suggestive of residual local disease, but no regional metastasis. Re-excision was recommended, and subsequent resections were undertaken to achieve microscopically negative margins including at the level of the periosteum. Reconstruction was accomplished using a rotational scalp flap with split-thickness skin grafting. The patient healed well and continues to be followed with clinical surveillance. She has demonstrated no evidence of recurrence to date, at over a year from final resection.

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 pediatric scalp lesions includes congenital nevus, atypical or acquired nevi, Spitz nevus, halo nevus, blue nevus, melanoma, or congenital dermal melanocytosis [10]. In our patient, keloid topped our differential list. The most typical clinical appearance for CH is an ill-defined, often solitary cutaneous nodule, ranging from flesh colored to red or purplish black in color. CH lesions may have a hemorrhagic or scaly external surface, can range in size, and may appear fixed to underlying structures on palpation [2]. Histopathologically, the lesions are composed of different types of vascular lesions, most commonly, epitheloid and retiform hemangioendothelioma components, but are consistently negative for WWTR1-CAMT1 fusion gene [11].

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 hypo pharynx 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 1A: Characteristics of scalp composite hemangioendothelioma.

Case	Reference	Age/ Sex	Site	Lesion size	Metastasis	Histology including EHE	Management/ Follow up
1	Liau et al. [7]	24 y/F	Scalp (left temporoparietal)	3 cm patch with 1.5 cm nodule	No	Yes	Surgical excision. Skin graft closure. NR 1 year.
2	Mahmoudizad et al. [8]	68 y/M	Scalp (vertex)	6.3 cm multilobulated nodule with adjacent papules	Yes; right posterior neck	Yes	Poor surgical candidate; radiation therapy with tumor shrinkage. Loss to follow up.
3	Current case	13y/F	Scalp (right parietal)	4 × 6 cm; prominent 2.5 cm nodule	No	Yes	Surgical excision. Skin graft closure. NR to date.

Table 1B: Characteristics of pediatric composite hemangioendothelioma in head and neck region.

Case	Reference	Age/ Sex	Site	Lesion size	Histology	Management/ Follow up
1	Tsai et al [9]	15y/F	Hypopharynx (left pyriform sinus)	5.5 × 4.2 × 3.2 cm bulging ulcerative mass	Spindle cell hemangioma and angiosarcoma-like	Primary resection. NR followed to 18 months.
2	Current case	13y/F	Scalp (right parietal)	4 × 6 cm; prominent 2.5 cm nodule	Venous malformation, EHE, and retiform hemangioendothelioma.	Surgical excision. Skin graft closure. NR to date.

EHE: Epitheloid Hemangioendothelioma; NR: No Recurrence

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

1. Requena L, Kutzner H. Hemangioendothelioma. Semin Diagn Pathol. 2013;30(1):29-44.
2. Shang Leen SL, Fisher C, Thway K. Composite hemangioendothelioma: clinical and histologic features of an enigmatic entity. Adv Anat Pathol. 2015;22(4):254-9.
3. McNab PM, Quigley BC, Glass LF, Jukic DM. Composite hemangioendothelioma and its classification as a low-grade malignancy. Am J Dermatopathol. 2013;35(4):517-22.
4. Nayler SJ, Rubin BP, Calonje E, Chan JK, Fletcher CD. Composite hemangioendothelioma: a complex, low-grade vascular lesion mimicking angiosarcoma. Am J Surg Pathol. 2000;24(3):352-61.
5. Leen SLS, Clarke PM, Chapman J, Fisher C, Thway K. Composite Hemangioendothelioma of the Submandibular Region. Head Neck Pathol. 2015;9(4):519-24.
6. Tsai JW, Huang HY, Lee JC, Yen YS, Tung CL, Huang CC, et al. Composite haemangioendothelioma: report of four cases with emphasis on atypical clinical presentation. Pathology. 2011;43(2):176-80.
7. Mahmoudizad R, Samrao A, Bentow JJ, Peng SK, Bhatia N. Composite hemangioendothelioma: An unusual presentation of a rare vascular tumor. Am J Clin Pathol. 2014;141(5):732-6.
8. Liau JY, Lee FY, Chiu CS, Chen JS, Hsiao TL. Composite hemangioendothelioma presenting as a scalp nodule with alopecia. J Am Acad Dermatol. 2013;69(2):e98-9.
9. Requena L, Luis Diaz J, Manzarbeitia F, Carrillo R, Fernandez-Herrera J, Kutzner H. Cutaneous composite hemangioendothelioma with satellitosis and lymph node metastases. J Cutan Pathol. 2008;35(2):225-30.
10. Jain S. Pediatric Dermatology. Dermatology: Illustrated Study Guide and Comprehensive Board Review. New York: Springer; 2012. p. 27-76.
11. Perry KD, Al-Lbraheemi A, Rubin BP, Jen J, Ren H, Jang JS, et al. Composite hemangioendothelioma with neuroendocrine marker expression: an aggressive variant. Mod Pathol. 2017;30(11):1589-602.