

Pseudomyogenic hemangiioendothelioma of the head and neck: Case report and review of literature

Nicole Wershoven, MD¹, Susan Potterveld, MD² Brain Cervenka, MD¹

¹ Department of Otolaryngology, Head and Neck Surgery, University of Colorado, Anschutz Medical Campus

² Department of Pathology, Head and Neck Surgery, University of Colorado, Anschutz Medical Campus

ABSTRACT

Pseudomyogenic hemangiioendothelioma is a newly recognized rare vascular tumor that is predominantly found in the lower limbs of young males. Recently, pseudomyogenic hemangiioendothelioma (PMHE) has been reported in several other areas of the body, including the head and neck. This entity is challenging to diagnose given the similarity to other tumors, inconspicuous morphologic characteristics, and lack of immunohistochemical features. Here we describe the first known case of pseudomyogenic hemangiioendothelioma with multiple fascial plane involvement of the head and neck in addition to a review of literature of the other known cases of PMHE in the head and neck. We recommend to the Otolaryngology community that pseudomyogenic hemangiioendothelioma be on the differential diagnosis for an indistinct tumor of the head and neck.



Figure 1. Clinical photos of PHE involving left temple/malar region as well as left upper eyelid

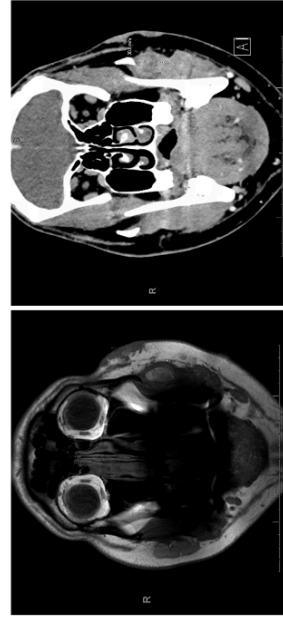


Figure 2. including CT neck/MRI face which demonstrated a heterogeneously enhancing nodule with the left masseter muscle measuring 2.3 x 1.7 x 3.1 cm (AP X TR X CC) and multiple subcutaneous nodules in the areas of overlying skin change which included nodules along the left cheek, temporalis fossa, and periorbita (Figure 2).

CASE REPORT

A 23-year-old male presented to clinic presents for surgical evaluation of PMHE that first was noted over a year prior as a small pimple on the left cheek and diagnosed via biopsy in a dermatology clinic.

- He subsequently received six cycles of Gem/Tax neoadjuvant therapy.
- On physical exam, skin lesions left temple/malar region as well as left upper eyelid (Figure 1). There was a palpable mass in left masseter region that was tender to palpation (Figure 1).
- Underwent radical excision of left facial skin and soft tissue, left parotidectomy, excision deep soft tissue masseter muscle mass, partial removal left zygomatic arch, left neck dissection, levels II and upper III, excision left upper eyelid with free flap reconstruction left face with left ALT musculocutaneous free flap, and left upper eyelid reconstruction with full thickness skin grafts.
- Final pathology revealed multiple foci of pseudomyogenic hemangiioendothelioma (PMHE) present in the dermis, subcutaneous adipose, and skeletal muscle (Figure 3).
- Post-operatively, the patient received no additional adjuvant treatment and is 14 months out without evidence of recurrence.

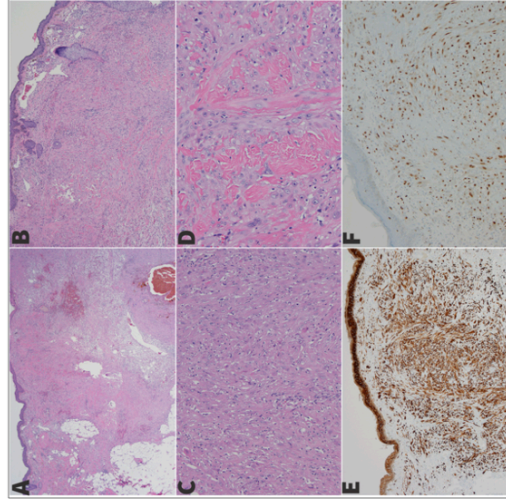


Figure 3. Pseudomyogenic Hemangiioendothelioma of the Head and Neck
 A. Proliferation of tumor cells within the dermis haphazardly infiltrating the underlying superficial subcutaneous adipose tissue (H&E, 2x).
 B. Tumor cells dissect between background collagen fibers and residual adnexal structures (H&E, 4x).
 C. Sheets of plump tumor cells admixed with a sparse inflammatory infiltrate composed of primarily neutrophils (H&E, 10x).
 D. Individual tumor cells with abundant brightly eosinophilic cytoplasm, large round to spindle nuclei with vesicular chromatin and prominent nucleoli. Occasional mitotic figures are present (H&E, 20x).
 E. An immunohistochemical study for pancytokeratin (AE1/AE3) shows strong diffuse cytoplasmic positivity in the tumor cells (4x).
 F. An immunohistochemical study for ERG shows strong diffuse nuclear positivity in the tumor cells (10x).

DISCUSSION

Demographic	Location	Treatment	Recurrence	Metastasis
33, F	Oral cavity	WLE	No	No
21, M	Nose/scalp	WLE	No	No
20, F	esophagus	Surgical excision with lymph node dissection	Yes	Yes- regional lymph nodes
60, F	Oral cavity	En bloc incision	No	No
35, M	Maxilla	Subtotal resection (List), en bloc resection (2nd) + chemotherapy	Yes	No
17, M	Diffuse head and neck ST	Chemotherapy (docetaxel and telatamib)	No	No
21, F	Oral cavity	Local excision	No	No
47, F	Scalp	WLE (2)	Yes	No
47, M	Nose	Local excision	No	No
34, M	Forehead	Local excision	No	No
25, M	Multifocal face	E bloc resection with ALT free flap	No	No

Table 2. Published cases of PMHE of the head and neck.

- There have been about 100 cases of Pseudomyogenic Hemangiioendothelioma reported in the literature, from 1993 to 2019
- PMHE of the head and neck is one of the rarest locations, 11 reported cases of PMHE of the head and neck
- The mean age was 32 and there was a 5:6 male to female ratio
- Most of these tumors were surgically removed either by piecemeal excision or en bloc resection
- 3/11 cases were also treated with chemotherapy
- Neoadjuvant chemotherapy was recommended in this case due to significant potential for local recurrence
- This therapy likely altered the extent of resection, as there was appreciable decrease in the size externally after seven cycles
- Of the 11 cases published, none of the patients received radiation therapy.
- There were three cases of local recurrence, however this could be limited due to the time of follow up for these patients
- We recommend to the Otolaryngology community that PMHE be recognized as part of the differential diagnosis for an indistinct tumor of the head and neck and that reporting these cases is imperative for improving the understanding and treatment of this tumor.

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