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CASE REPORT

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Perivascular epithelioid cell tumor (PEComa) of the pterygopalatine fossa

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Abstract

Perivascular epithelioid cell tumors (PEComas) are a rare group of mesenchymal tumors associated with tuberous sclerosis. These tumors are typically treated with resection and rarely recur or exhibit malignant behavior. A 78-year-old woman presented with an incidentally discovered pterygopalatine fossa/retroantral mass. Excisional biopsy was performed and revealed pathology consistent with PEComa. Upon review of the literature, there have been 43 reported cases of PEComa of the head and neck. There is only one previously reported case of PEComa in the skull base, and none reported in the pterygopalatine fossa. Of note, the previously reported case of skull base PEComa involved an aggressive tumor with widespread metastasis. Here, we report the first case of a PEComa of the pterygopalatine fossa/retroantral region, which was treated conservatively. This rare pathology should be considered in the differential diagnosis for atypical skull base tumors.

KEYWORDS

head and neck, PEComa, pterygopalatine fossa, retroantral, skull base tumor

1 | INTRODUCTION

Perivascular epithelioid cell tumors (PEComas) are a family of rare mesenchymal neoplasms that show a perivascular tumor epithelioid cell (PEC) differentiation. This group of tumors includes angiomyolipoma, clear cell "sugar" tumor (CCST) of the lung and extrapulmonary sites, clear cell myomelanocytic tumor of the falciform ligament, and lymphangioleiomyoma. Perivascular epithelioid cell tumors have distinct histologic features such as epithelioid to spindle cell appearance with clear to granular eosinophilic cytoplasm and round to oval nucleus with inconspicuous nucleoli. Immunohistochemically, this family of tumors characteristically shows markers of PEC differentiation including myogenic and melanocytic markers. Perivascular epithelioid cell tumors have previously been associated with tuberous sclerosis,[1] although they have been reported to occur in the absence of this syndrome.[2] Perivascular epithelioid cell tumors are typically benign but are known to recur,[3] and malignant behavior has been reported from a number of primary sites.[4] Commonly reported sites of origin include abdominal and gynecologic soft tissue,[5] although cases in the head and neck region have been previously described.[6] Here, we describe the first reported case of PEComa of the pterygopalatine fossa.

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2 | CASE

A 78-year-old woman was transferred to our hospital for management of a left occipital intracerebral hemorrhage. A follow-up MRI incidentally revealed a T2 hypointense, T1 isointense left pterygopalatine fossa/retroantral region mass with postcontrast enhancement measuring 16 × 7 mm (Figures 1 and 2). The mass was noted to have enlarged since a CTA obtained 4 months prior, where the mass was noted in retrospect. The lesion was approached via an endoscopic, endonasal transmaxillary approach using navigation with excisional biopsy resection. During this procedure, the inferior turbinate was left intact, the middle turbinate was medialized for uncinectomy, the maxillary ostium was opened, ethmoid bulla was removed, posterior maxillary wall was resected, and internal maxillary artery

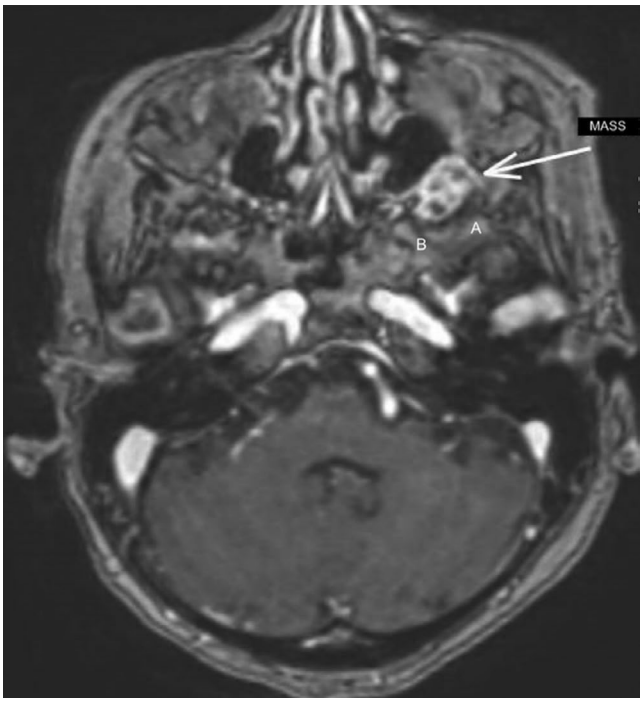

FIGURE 1 T1 postcontrast axial MRI. Arrow points to the mass lesion. A represents the lateral pterygoid muscle. B represents the pterygoid plate

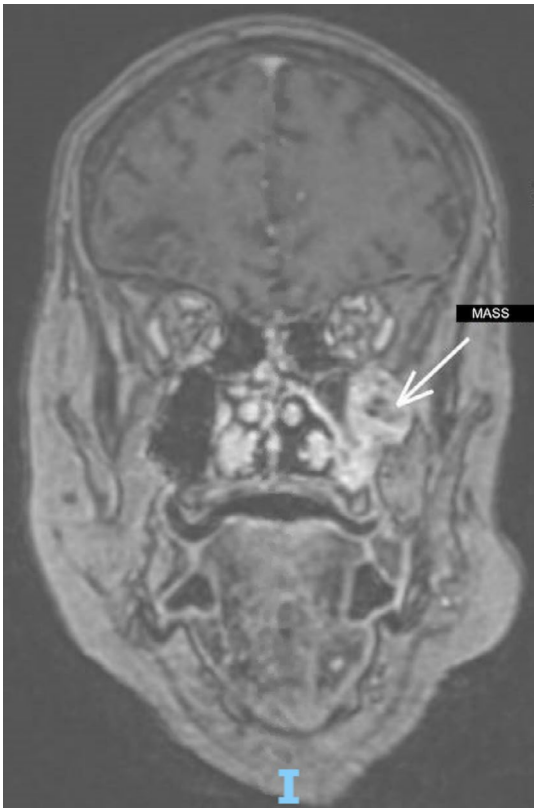

FIGURE 2 T1 postcontrast coronal MRI. Arrow points to the mass lesion

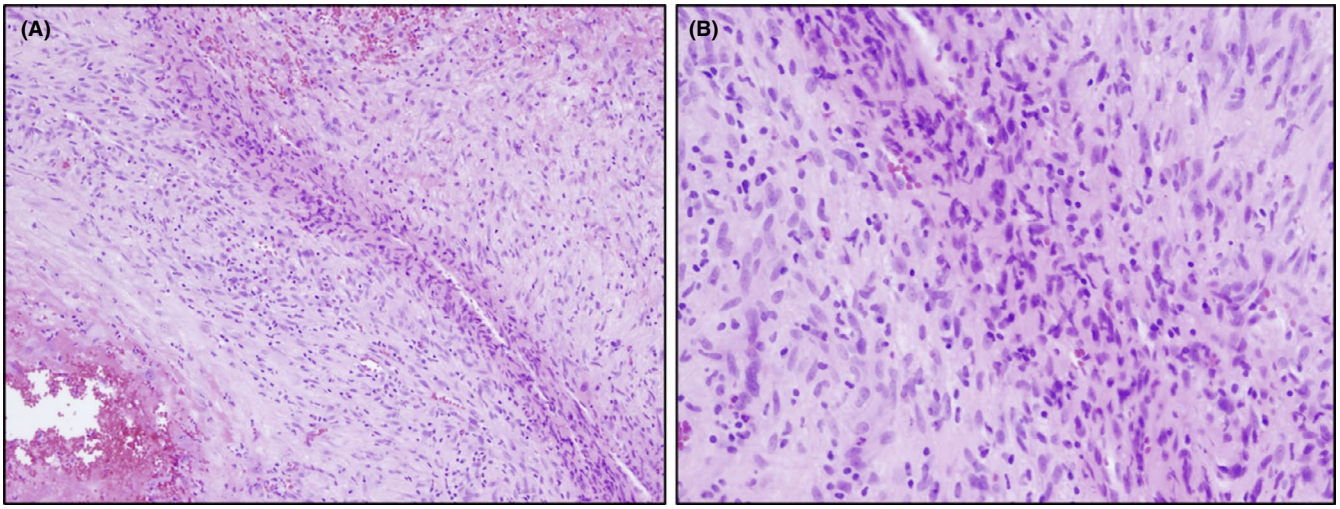

FIGURE 3 A, Organizing hematoma (arrow) surrounded by sheets of low-grade spindle cell proliferation with variable cellularity (200×, H&E). B, Bland spindle cells with eosinophilic cytoplasm and oval nuclei with small nucleoli in the absence of significant mitosis (400×, H&E)

TABLE 1 Literature review of PEComa of the Head and Neck. A review of the literature was performed with a Pubmed search of the terms "PEComa" and "head and neck."

Author (Year)	Location	Age/Gender	Treatment	Outcome
Lehman (2004)[11]	Posterior skull base	49-y-o female	Partial resection	Metastasis to lung and spine, death
Weindling et al (2015)[13]	Maxillary sinus	78-y-o male	NR	NR
Leavers et al (2012)[14]	Maxillary nasal process	74-y-o female	Resection	No recurrent disease
Banerjee et al (2001)[15]	Nasal cavity	34-y-o female	Resection	No recurrent disease
Kuroda et al (2009)[16]	Nasal cavity	79-y-o male	NR	NR
Afrogheh et al (2013)[17]	Nasal cavity	19-y-o female	Resection	No recurrent disease
Bocciolini et al (2013)[18]	Nasal cavity	40-y-o female	Resection	No recurrent disease
Panelos et al (2009)[19]	Nasal septum	50-y-o female	Resection	No recurrent disease
Erkilic et al (2005)[20]	Nasal cavity	52-y-o male	Resection	NR
Iwata et al (2013)[21]	Nasal cavity	60-y-o male	Resection	NR
Moreira et al (2011)[22]	Nasal cavity	54-y-o male	Resection	No recurrent disease
Tardio et al (2002)[23]	Nasal cavity	45-y-o male	Resection	No recurrent disease
Gatalica et al (1994)[24]	Nasal cavity	64-y-o male	NR	NR
Watanabe et al (1999)[25]	Nasal cavity	66-y-o male	Resection	No recurrent disease
Watanabe et al (1999)[25]	Nasal cavity	88-y-o female	Resection	No recurrent disease
Dawlatly et al (1988)[26]	Nasal cavity	52-y-o male	NR	No recurrent disease
Gana et al (2012)[2]	Nasal cavity	22-y-o female	Resection (positive margins), re-resection	No recurrent disease
McGregor et al (2017)[27]	Nasal cavity	54-y-o female	Resection	No recurrent disease
Bandhlish et al (2011)[6]	Nasal cavity	18-y-o female	Embolization, resection (positive margins), re-resection	No recurrent disease
Bandhlish et al (2011)[6]	Nasal cavity	71-y-o female	NR	NR
Bandhlish et al (2011)[6]	Glottis	26-y-o female	Resection	No recurrent disease
Huai-yin et al (2009)[28]	Glottis	38-y-o female	Resection	Re-resection for recurrence at 15 mo, no evidence of disease
Huai-yin et al (2009)[28]	Laryngeal vestibule	42-y-o male	Resection	No recurrent disease
Huai-yin et al (2009)[28]	Hypopharynx	47-y-o male	Resection	No recurrent disease
Koutlas et al (2005)[29]	Hard palate	46-y-o female	Resection	No recurrent disease
Saluja et al (2018)[12]	Oropharynx	28-y-o female	Resection, everolimus, radiation	No recurrent disease
Foschini et al (1999)[30]	Parotid	68-y-o female	Resection	No recurrent disease
Ghazali et al (2010)[31]	Cheek	32-y-o female	Resection	No recurrent disease
Greveling et al (2013)[32]	Cheek	44-y-o male	Resection (positive margins), re-resection, radiation	No recurrent disease
Girardi et al (2018)[33]	Cheek	69-y-o male	Resection, radiation	No recurrent disease
Calder et al (2008)[34]	Scalp	76-y-o male	Resection	No recurrent disease
Argani et al (2010)[35]	Scalp	80-y-o male	NR	NR
Folpe et al (2005)[5]	Scalp	80-y-o male	Resection	Lost to follow-up
Folpe et al (2005)[5]	Neck	77-y-o female	Resection, re-resection, radiation	No recurrent disease

(Continues)

TABLE 1 (Continued)

Author (Year)	Location	Age/Gender	Treatment	Outcome
Iyengar et al (2004)[36]	Orbit	9-y-o female	NR	NR
Paliogiannis et al (2016)[37]	Orbit	46-y-o male	Resection	No recurrent disease
Nair et al (2018)[38]	Orbit	9-y-o female	Resection	No recurrent disease
Lubo et al (2016)[39]	Orbit	47-y-o male	Resection	No recurrent disease
Guthoff et al (2008)[40]	Orbit	54-y-o male	Resection	No recurrent disease
Alam et al (2017)[41]	Orbit	5-y-o male	Resection, Chemotherapy	No recurrent disease
Furusato et al (2010)[42]	Eyelid	26-y-o female	Excisional biopsy, re-excision	No recurrent disease
Furusato et al (2010)[42]	Ciliary body	7-y-o male	Resection	No recurrent disease
Goto et al (2015)[43]	Ciliary body	13-y-o female	Resection	No recurrent disease

was clipped to provide adequate hemostasis. Tumor specimen was sent for frozen section, which returned as chronically inflamed polypoid tissue. Due to the apparent benign nature of the mass, the tumor was simply debulked, forgoing further dissection superiorly to the inferior orbital fissure. Microscopic examination revealed an organizing hematoma surrounded by sheets of spindle-shaped cells with eosinophilic cytoplasm and oval nuclei with small nucleoli and low mitotic count (Figure 3A,B). A panel of immunohistochemical stains was performed that revealed positivity only for HMB45 and smooth muscle actin. This distinct immunohistochemical profile is suggestive of PEC differentiation. In addition, negative in situ hybridization for EWS and TFE3 was supportive of the diagnosis of conventional PEComa. She did not have a personal or family history of tuberous sclerosis. Head and neck tumor board recommended serial imaging given her medical comorbidities, with possible radiation therapy if significant progression was noted. She died 3 months later from complications of her intracerebral hemorrhage.

3 | DISCUSSION

Pterygopalatine fossa masses are rare and have a broad differential diagnosis including epidermoid cyst, meningocele, mucosal carcinoma, schwannoma, chordoma, teratoma, and neurofibroma.[7] The majority of these masses are benign in nature, although they can be locally invasive. The workup of pterygopalatine fossa lesions generally consists of nasal endoscopy, CT scan, and contrast-enhanced MRI. Biopsy is indicated to rule out malignancy except in the case of suspected juvenile nasopharyngeal angiofibroma, and the decision to radically resect can be made based on tumor histology.[8] PEComas typically exhibit benign behavior although they have been reported to metastasize, most commonly to cutaneous sites,[4,9] although Tynski et al reported a case of PEComa metastasis to the orbit.[10] Our literature review found 43 reported cases of PEComa of the head and neck; however, most were cutaneous in the nasal cavity or in the orbit (Table 1). Only one case of skull base PEComa has been reported, and there are no previously reported cases of this pathology in the pterygopalatine fossa. The case of posterior skull base PEComa reported by Lehman involved a tumor with histologic evidence of malignancy that had extensive local invasion of the petrous/occipital bones, clivus, and foramen magnum as well as eventual metastasis to the lung and spine resulting in the patient's demise.[11] Our case of PEComa of the pterygopalatine fossa did not display signs of malignancy histologically or clinically, although long-term surveillance was not possible due to the patient's unrelated death shortly after diagnosis. Management of these tumors generally consists of resection alone due to their benign behavior although observation is a reasonable option depending upon the location of the tumor and resulting signs/symptoms, evidence of malignant behavior, and patient comorbidities. Radiation therapy has also been used with some success,[12] but there is no established indication for adjuvant radiation.[6]

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

MD: collected data, assembled figures, wrote key portions of the manuscript, and prepared manuscript revisions. SP: participated in the care of the patient and edited/revised manuscript at all stages. AG: participated in the care of the patient, collected histopathology images, and wrote portions of the manuscript including pathologic descriptions. JM: participated in the care of the patient, wrote key portions of the manuscript, and edited/revised manuscript at all stages.

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