Atypical Meningothelial Meningioma Presenting as a Painful Palate Mass

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Abstract

Objectives: To describe a rare case of an extracranial meningioma presenting as a painful palate mass, with multiple cranial neuropathies and an unusual destructive pattern on imaging.

Case: We present a case of a 79 year old female who presented to our clinic with a painful palate mass, persistent otalgia and hearing loss. She additionally noted right facial hypoesthesia, ageusia, and dysphagia. CT and MRI were obtained showing a 4.6 cm hypervascular mass involving the right masticator and parapharyngeal space with erosion of the pterygoid plates and maxillary sinus. Transoral biopsy of the palate mass revealed atypical meningothelial meningioma (WHO Grade 2) with accelerated growth. The patient was treated via a multidisciplinary surgical approach with head and neck surgery, oral and maxillofacial surgery, and neurosurgery.

Conclusions: Extracranial meningioma presenting in the oral cavity is exceedingly rare, but should be considered when evaluating a soft tissue mass of the palate. In patients presenting with multiple symptoms and destructive tumors, surgical resection should be recommended.

Introduction

Meningiomas are tumors arising from the meningeal arachnoidal (meningothelial) cells. They are typically benign on histology, represent approximately 20-30% of intracranial tumors, affect women more commonly than men, and increase in incidence with age[1,2] Some risk factors have been correlated with meningioma occurrence (e.g. radiation exposure, obesity, prior head trauma, heritable traits such as neurofibromatosis type-2) and other factors have been hypothesized to be related (e.g. sex hormones).[1,3] Given their typically benign character, these tumors are often asymptomatic and go undiagnosed until incidentally found on imaging.

Up to 20% of primary intracranial meningiomas demonstrate secondary extracranial extension.[4-7] Only a small minority of primary meningiomas occur extracranially, with their recorded rates of primary occurrence ranging between 0.8-2%. Outside of direct extension, extracranial meningiomas have been theorized to occur from the arachnoid cells of cranial nerve sheaths, from extracranial embryonic arachnoid cells, and from metastases from intracranial meningiomas.[9] To our knowledge, the largest studies of extradural meningioma to date have involved analysis of 231 and 146 cases by Liu et al and Rushing et al, respectively.[8,10] These studies have demonstrated that the meningiomas most commonly present in the skull, scalp, nose, orbit, paranasal sinuses, middle ear, neck and skin.[8] Among these reviewed cases, only one involved the palate.

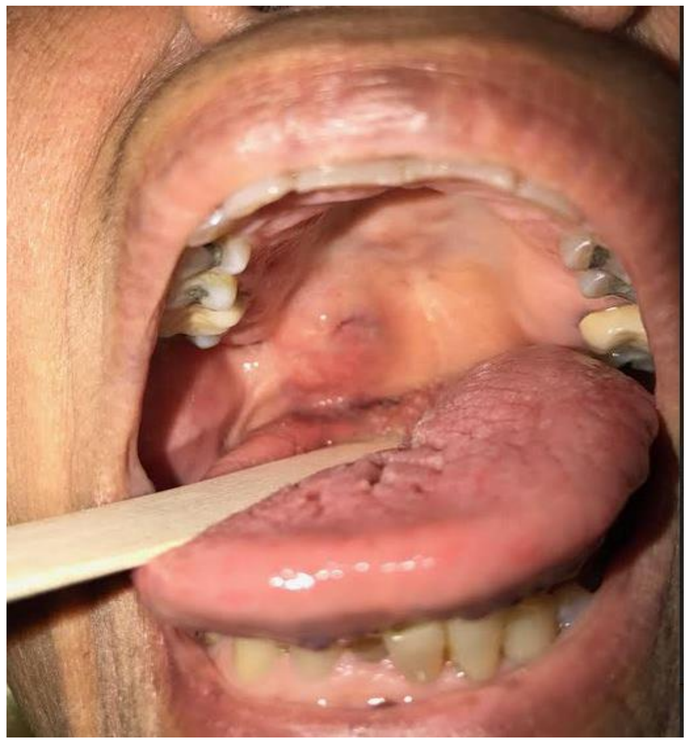

Figure 1. Palatal mass with healing biopsy site

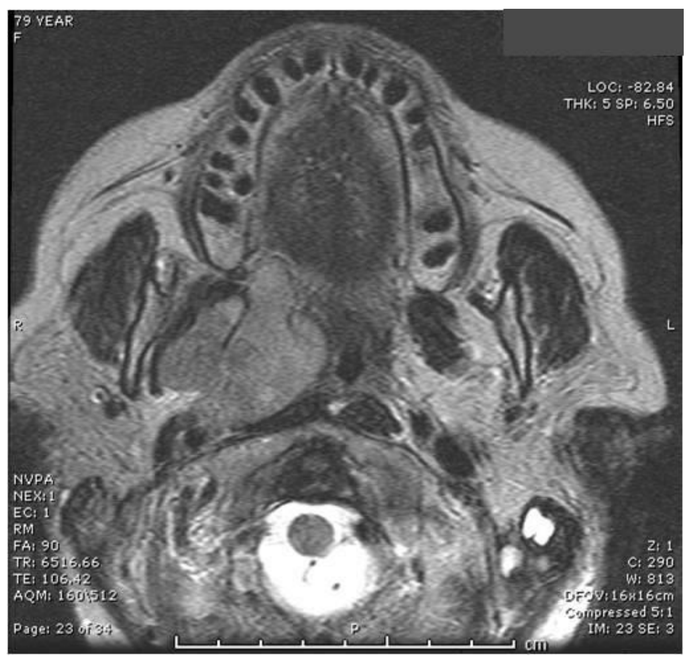

Figure 2. Axial MRI demonstrating soft tissue mass arising from the right pharyngeal mucosal space

Conclusions

Extracranial meningioma presenting in the oral cavity is exceedingly rare, but should be considered when evaluating a soft tissue mass of the palate. In patients presenting with multiple symptoms and destructive tumors, surgical resection should be recommended.

Case Report

A 79 year-old female presented to the emergency department with chief complaints of 3 years worsening hearing loss and ear pain on the right side. Past treatments included placement of a tympanostomy tube and polymyxin ear drops, without any improvement in her symptoms. At the time of presentation to Head and Neck Surgery clinic she noted a painful swelling of her palate, pain with mastication, mild dysphagia, dysgeusia and hemifacial paresthesia. Physical exam was significant for a tender mass of her right hard palate soft palate junction which extended to the nasopharynx. She additionally had decreased sensation in the right V3 distribution, right sided middle ear effusion, and decreased taste on the right.

Diagnostic studies included an audiometry, MRI and oral biopsy. Pure tone audiometry revealed mild to profound sensorineural hearing loss on the right and moderate to severe mixed hearing loss on the left. Word recognition scores were good (80-100%) bilaterally. MRI demonstrated a 4.6 cm hypervascular mass that appeared to be arising from the right pharyngeal mucosal space extending posterolaterally with involvement of the pterygoid muscles and apparent invasion of the skullbase on the right. The lesion was reported as perineural spread via the right V3 nerve. However, on histology the lesion was definitively diagnosed as atypical meningothelial meningioma, WHO Grade 2. Upon returning to clinic, surgical options were discussed and the patient elected to undergo resection. She is scheduled for and awaiting surgery.

Discussion

The World Health Organization classification distinguishes between three grades of meningioma based on histology: Typical or benign (Grade I); atypical with frequent mitosis (Grade II); and anaplastic with invasion (Grade III).[2] Rushing et. al demonstrated in a study of 146 primary extracranial meningiomas that the vast majority were WHO Grade I tumors (87.7%), followed by Grade II (9.6%) and Grade III (2.7%) tumors. Five year disease specific survival were shown to decrease from 92.4% (Grade I) to 88.9% (Grade II) to only 50% (Grade III).[10] Given that the strongest positive prognostic indicator in meningothelial meningioma is the completeness of surgical excision, surgical planning and technique are vastly important. The anatomical complexity of the head and neck and the critical importance of its structures present serious challenges with these invasive tumors. In order to achieve completed resection in this case, either a transpalatal approach or maxillary swing approach will be used.

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