INTRODUCTION

Rhabdomyosarcoma (RMS) is a rare diagnosis in adult patients but is one of the most common head and neck sarcomas in the pediatric population.1-3 The differential of pterygopalatine fossa masses is broad and must include RMS.4 In adults, RMS is generally associated with unfavorable characteristics: parameningeal location, tumor size, and aggressive histology.3 Survival and local control are greatly reduced relative to the pediatric population.1,3 Pediatric populations predominantly display alveolar and embryonal histology, while pleomorphic histology is displayed more often in adults. The spindle cell variant of RMS is rare in both adult and pediatric populations and carries the least favorable prognosis.

METHODS

The case of an adult patient with a right pterygopalatine fossa mass is reviewed. Clinical history, imaging and histology are examined to allow for comparison and discussion.

CASE PRESENTATION

The patient is a 24-year-old male with a 6-week history of right-sided otalgia, ipsilateral chin and cheek hypesthesia, and migraine-type headaches.

RESULTS

The CT and MRI of neck with IV contrast (Fig. 1) demonstrated a 2.8 x 3.2 cm right pterygopalatine fossa mass extending to the skull base. Endoscopic examination and transantral biopsy were performed (Fig. 2). Histopathology demonstrated characteristic spindle-shaped rhabdomyoblasts with atypical, hyperchromatic nuclei and positivity for desmin and myogenin (Fig. 3). Due to the extension of the mass to the skull base with involvement of the foramen ovale and foramen rotundum with intracranial extension along V3 seen in MRI of neck with contrast (Fig. 1 a), the tumor was determined to be unresectable. He underwent two cycles of VADc (vincristine, actinomycin, and cyclophosphamide) followed by 50.4Gy radiotherapy with concomitant ifosfamide/etoposide. Due to continued mass growth throughout the above therapies (Fig. 1 b-f), and persistent unresectable disease, the patient is scheduled to enroll in ARST0921 – A randomized phase II trial of bevacicuzumab (IND# 7921, Avastin) and temsirolimus (IND# 61010, Torisel) in combination with vinorelbine and cyclophosphamide for recurrent/refractory RMS.

DISCUSSION

The incidence of RMS in adult populations is considerably lower than the pediatric counterpart.1,3 While rare, the rapid diagnosis of RMS must be considered in light of the many important factors that have been shown to influence outcomes with the neoplasm; namely, anatomical site, tumor size, patient age, and histological subtype. These characteristics help determine the most appropriate treatment and counsel.

Due to its internal location, pathology within the pterygopalatine fossa often goes undetected until imaging studies are obtained. It is critical that clinicians recognize symptomatology that may suggest a contained mass and indicate the need for imaging of the face/neck (CT/ MR): unilateral facial pain or swelling, hearing loss, otalgia, facial hypesthesia or paresthesia, or nasal obstruction.4

CONCLUSIONS

Rhabdomyosarcoma remains a rare malady, particularly in the adult population, but the awareness and appreciation of this malignant disease process is important for providing early diagnosis and appropriate clinical care and counseling. There remains a very strong reliance on histopathological as well as radiological diagnostic techniques, and therapy continues to be centered on conventional chemotherapy (VADc), radiation therapy, and surgical excision.1,2

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REFERENCES


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