Case Report

Intracranial Rhabdomyosarcoma of the Cerebellopontine Angle in a 6-year-old Child: A Case Report

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Rhabdomyosarcoma (RMS) is the most common soft-tissue malignancy in children under 15 years of age. Approximately, 35% of RMS cases originate from the head and neck region. Among various sites in the head and neck region, intracranial extension is more likely to occur with parameningeal tumors, which is also considered an unfavorable prognostic factor in children with RMS. About 20% of RMS occurs in a parameningeal site. Intracranial RMS are rare tumors that usually arise from parameningeal sites or metastasis from an extracranial site. Primary intracranial types are even rarer, with only 50 reported cases of primary intracranial RMS in the literature.

Here, we report the case of a 6-year-old boy who presented with clinical and radiologic features of a cerebellopontine angle lesion, which turned out to be a RMS.

Keywords: Cerebellopontine angle, children, intracranial, parameningeal, rhabdomyosarcoma

Introduction

Rhabdomyosarcoma (RMS) is the most common soft-tissue malignancy in children under 15 years of age.[1] Approximately, 35% of RMS cases present in the head and neck region.[2] The orbit is the most common site within the head and neck, and accounting for 25%–35% of head and neck RMS, followed by parameningeal sites (including nasal and paranasal sinuses, middle ear and mastoid sinus, parapharyngeal space, and pterygopalatine and infratemporal fossae). Of all the sites, intracranial extension is more likely to occur with parameningeal tumors and is generally considered an unfavorable prognostic factor in children with RMS. About 20% of RMS occurs in a parameningeal site.[3] Intracranial RMS are rare tumors that usually arise from parameningeal sites or metastasis from an extracranial site.[4] Primary intracranial types are even more rare, with only 50 reported cases of primary intracranial RMS in the literature.[5]

Here, we report the case of a 6-year-old boy who presented with clinical and radiologic features of a cerebellopontine angle (CPA) lesion mimicking a vestibular schwannoma or osteosarcoma, but was histologically revealed to be a RMS.

Case Report/Case Presentation

Case. A 6-year old boy who presented with progressive gait disturbance, right facial weakness, and oropharyngeal dysphagia with difficulty swallowing over a 2-month time frame. He was oriented to time, person, and place. Neurological examination revealed moderate right hemiparesis, ataxia, left sixth CN paresis, weak left infra-nuclear facial paresis, impaired left gag reflex, and left-sided cerebellar signs.

Imaging. Brain magnetic resonance imaging (MRI) with and without intravenous contrast was performed, which showed an extraaxial 4.6 cm × 4.4 cm T2W hyperintense and T1W hypointense lesion [Figure 1].
The lesion’s epicenter was the right petrous temporal bone with posterior extension to right CPA and mass effect and medial displacement on the right cerebral and middle cerebellar peduncles and mild posterior displacement of the right cerebellum. A small amount of vasogenic edema was noted in the medial right middle cerebellar peduncle. The lesion also extended anteriorly to involve the middle cranial fossa.

Figure 1: Brain MRI with and without intravenous contrast showing an extraaxial (4.6 cm × 4.4 cm) T2W hyperintense (first row) and T1W hypointense (second row) lesion with heterogeneous enhancement after contrast administration (third to fifth rows). The lesion’s epicenter was the right petrous temporal bone with posterior extension to right cerebellopontine angle and mass effect and medial displacement on the right cerebral and middle cerebellar peduncles and mild posterior displacement of the right cerebellum with a small amount of vasogenic edema in the medial right middle cerebellar peduncle. The lesion also extended anteriorly to involve the middle cranial fossa.
showed heterogeneous enhancement after contrast administration, making vestibular schwannoma and osteosarcoma the most likely diagnoses. There was no hydrocephalus despite partial effacement of the fourth ventricle.

Operation. A right retrosigmoid craniectomy was performed. Because of extensive tumor adherence to major vascular and brainstem structures, a near total resection was performed. On gross examination, the tumor had a firm consistency and was moderately vascular.

Histology. Hematoxylin and eosin stained material showed hypo- and hypercellular areas with myxoid stroma containing round and spindled cells with hyperchromatic nuclei and scanty cytoplasm. The neoplastic cells were strongly positive for desmin and MyoD1 [Figure 2].

Post-op and discharge. The patient could not be extubated because of weak gag reflex. Therefore, a tracheostomy tube was placed for him. He was discharged on the 14th postoperative day and referred to a pediatric oncologist for chemotherapy.

Follow-up. At 3-month follow-up, his hemiparesis and cerebellar signs had improved and he could walk with assistance. His gag reflex was acceptable and the tracheostomy tube was discontinued. At 6-month follow-up his general condition was well and MRI of the brain showed no recurrence of the tumor.

DISCUSSION

CPA lesions are uncommon in pediatric population with an incidence rate of less than 10%. Unlike adult population where the vast majority of CPA lesions are vestibular schwannomas and meningiomas, pediatric patients experience more various tumor types comprising 35%–45% of CPA lesions (e.g., arachnoid cyst, lipoma, and cavernous hemangioma).[6] Malignant sarcomas constitute less than 1% of pediatric CPA tumors.[5]

RMS is the most common soft-tissue malignancy in children, accounting for 3%–4% of all pediatric malignancies.[1]

RMS of the middle ear occurs almost exclusively in children. At the time of diagnosis, the tumor has often invaded the external canal, mastoid, and meninges.[7] Microscopically, they are most often of embryonal type, including the botryoid variety.[8]

Intracranial RMS is an aggressive, rapidly growing, and vascular tumor that carries a very poor prognosis with only nine reported case survived beyond 2 years.[3] Unfortunately, RMS cannot be distinguished from other primary or metastatic tumors of the brain with imaging alone.[4]

Multimodality approach including total surgical resection with subsequent chemotherapy and radiotherapy is regarded as the best course of treatment.[9,10]

CONCLUSION

CPA lesions are generally uncommon in pediatric population. Various tumor types other than schwannomas and meningiomas are seen in CPA in pediatric population. Malignant sarcomas constitute less than 1% of these pediatric CPA tumors. Intracranial RMS is an aggressive, rapidly growing tumor with a very poor prognosis. Multimodality therapy is regarded as best treatment.
Ethical policy and institutional review board statement
This study was approved by the Ethics Committee on Human Research of the Shiraz University of Medical Sciences.

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Conflicts of interest
There are no conflicts of interest.

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