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Embryonal mastoid rhabdomyosarcoma in a three years old child: A case report

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ABSTRACT

INTRODUCTION: Rhabdomyosarcoma is the most common malignant soft tissue tumor in the pediatric age, especially in the head and neck region; in the orbit and the nasopharynx. The middle ear is a very rare site for this neoplasm as it accounts for only 10 % of head neck rhabdomyosarcoma.
PRESENTATION OF CASE: We report here the case of a three years-old child who was admitted to the emergency room for a left parotid and retro-auricular swelling with grade V facial palsy. The patient experienced chronic otorrhea with left facial palsy for two months. Two weeks later, a left parotid swelling appeared and gradually increased in size with weight loss. Computed tomography showed a slightly dense tissue lesion in the left mastoid. The patient underwent a diagnostic mastoidectomy. Pathology and immunohistochemical study were compatible with an embryonic rhabdomyosarcoma.
CONCLUSION: Rhabdomyosarcoma should be considered as a differential diagnosis of any mastoiditis resistant to treatment in a young child. Because of its serious prognosis, rhabdomyosarcoma must be diagnosed at an early stage to increase chances of recovery.

1. Introduction

Rhabdomyosarcoma is the most common malignant soft tissue tumor in the pediatric population [1]. 35 % of all pediatric rhabdomyosarcomas are in the head and neck region [2,3]. The middle ear is a very rare site for this neoplasm as it accounts for only 10 % of head neck rhabdomyosarcomas [4].

The clinical presentation of middle ear rhabdomyosarcoma simulates suppurative chronic otitis media which might delay the diagnosis [5]. In an advanced stage, facial paralysis and signs of intracranial extension appear. This work is reported by following the surgical case report (SCARE) guideline [6].

2. Presentation of case

A three years-old child with no relevant personal nor family medical history was admitted to the emergency room for a left parotid and retro-auricular swelling with facial palsy. The patient experienced chronic otorrhea with left facial palsy for two months. Two weeks later, a left parotid swelling appeared and gradually increased in size with weight loss. Clinical examination showed a grade V left peripheral facial paralysis (House Brackmann score), a hard swelling extending from the mastoid region, the pinna to the parotid region and a bleeding whitish mass in the left ear canal (Fig. 1). Computed tomography (CT) showed a slightly dense tissue lesion in the left mastoid with marked contrast enhancement, measuring 58 × 45 × 37 mm. The tumor caused osteolysis of the mastoid bone, the walls of the middle ear cavity, the petrous apex and the carotid canal. It had an endocranial temporal development with extension to the left cerebellopontine angle (Fig. 2).

We informed the father's child about the surgical procedure, its potential complications and the expected outcomes which were accepted. The patient underwent a diagnostic mastoidectomy by a senior otologic surgeon. No post-operative complication was noted. Pathology and immunohistochemical study were compatible with an embryonal rhabdomyosarcoma (Figs. 3 and 4). The patient was referred to the pediatric oncology department where she started chemotherapy (ifosfamide 3 g/m2, vincristine 4.5 g/m2, dactinomycin 1.5 g/m2 and doxorubicin 30 g/m2). 1 month follow-up showed a decrease in otorrhea and disappearance of otalgia.

3. Discussion

Temporal bone neoplasms represents a challenge in pediatric population because of its rarity whose rhabdomyosarcoma is the most common [7].

Rhabdomyosarcomas include three histologic subtypes: embryonal, alveolar, and pleomorphic [8]. The embryonal type is most commonly found in the head and neck. Of all rhabdomyosarcomas in children, 30%–50% occur in the head and neck and the most frequent localizations are the nasopharynx and the orbit.

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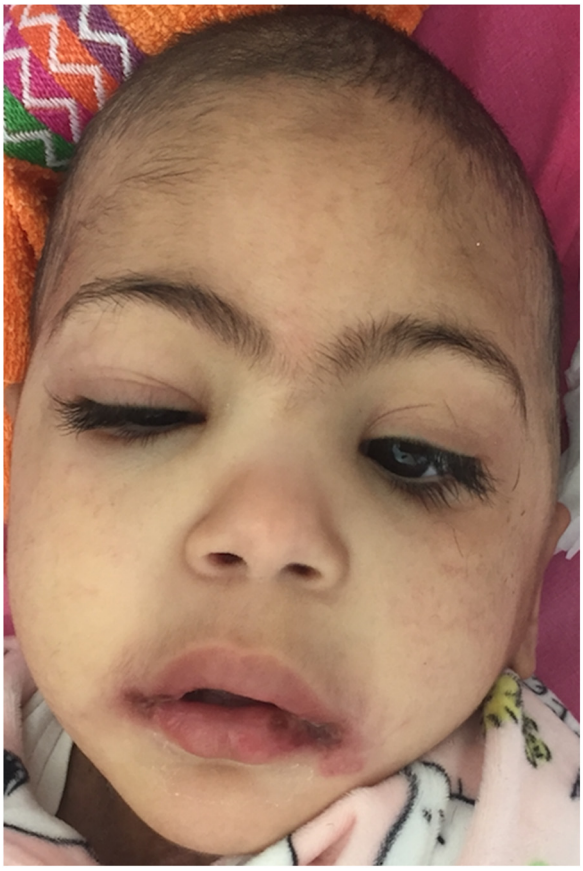

Fig. 1. Child admitted with left peripheral facial palsy grade V (House Brackmann score).

Sbeity and al, reported a serie of 6 children aged between 3 and 5 years old. Our child is 3 years old [5].

Chronic suppurative otitis media resistant to usual medical treatment can mimic temporal bone rhabdomyosarcoma [6].

Facial paralysis is often present during diagnosis due to the rapid course of the disease. Invasion of the petrous apex, the internal auditory canal and the skull base can lead to cranial nerve deficits [9].

Pratt and Gray reported the signs and symptoms of 50 patients with rhabdomyosarcoma of the middle ear. Mass is the ear region is the most commonly found (56 %) followed by polyp in the external auditory canal (54 %), aural discharge (40 %), bleeding (30 %), earache (22 %) and facial paresis (14 %) [10].

In this case, the patient presented with purulent otorrhea, facial paralysis and intracranial extension which is due to delayed consultation.

CT scan is more efficient in bone analysis. Temporal bone rhabdomyosarcoma usually shows aggressive bone destruction with obliteration of normal skull base landmarks. Magnetic Resonance Imaging (MRI) is preferred to CT scan to assess endo and exocranial involvement, as well as the relationship with the jugular vein and carotid arteries. However, rhabdomyosarcoma has a non-specific MRI signal characteristics. It is isointense to slightly hyperintense to adjacent muscle in T1 and hyperintense in T2 sequences [11].

Biopsy is highly recommended for children with otitis media resistant to medical treatment [5].

On histopathological examination rhabdomyosarcoma is a round cell tumor [12]. Immunohistochemistry rules out other small round cell tumors, such as lymphoma (CD20, CD3 positive), Ewings and primitive neuroectodermal tumors sarcomas (CD99 positive). Rhabdomyosarcoma is desmin positive and negative for the other markers [13].

The aim of the treatment is to achieve loco-regional control and to prevent distant metastases [13]. Some authors recommend primary surgical resection for localized tumors without intracranial extension or distant metastasis [14]. However, surgery is usually limited to biopsy for histopathological examination in other cases [5]. Chemotherapy with loco-regional radiotherapy are the main therapeutic mainstay [15].

Donaldson suggested a staging system which is a modified version of the Amercian Joint Committee for Cancer Staging and the International Union Against Cancer [16]. Patients with stage 1 and stage 2 have a far better outcome. However, intracranial invasion (stage 3) is a factor of poor prognosis. Sbeity and al reported two children with intracranial invasion who died after 1 and 6 months [5]. Our patient showed a good clinical evolution after 1 month follow up.

4. Conclusion

Rhabdomyosarcoma should be considered as a differential diagnosis of any mastoiditis resistant to treatment in a young child especially with facial palsy. Because of its serious prognosis, rhabdomyosarcoma must be diagnosed at an early stage to increase chances of recovery.

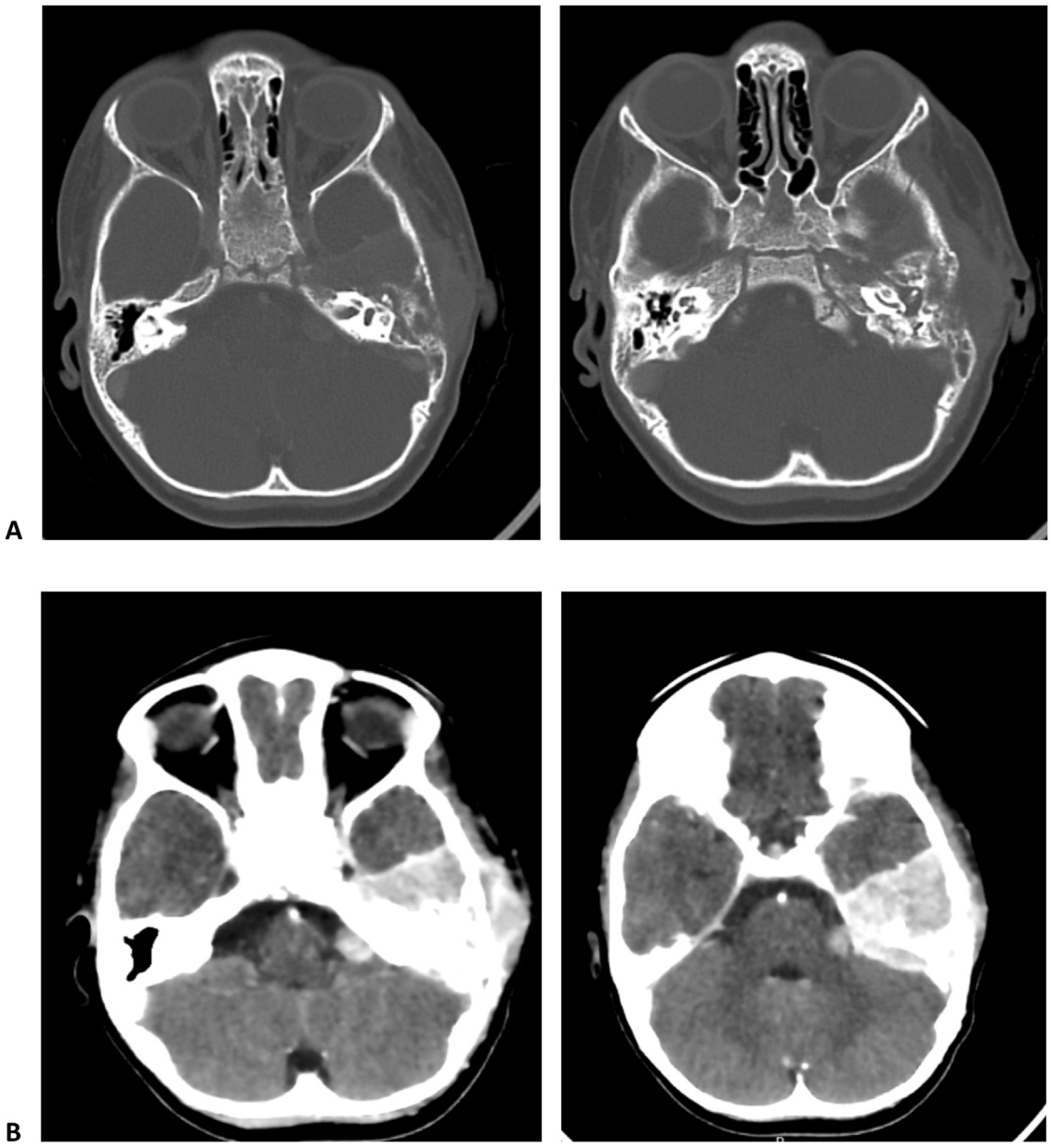

Fig. 2. A: Axial CT scan showing bone lysis by a lesional process responsible for mastoid ostelysis with lysis of the petrous apex.
B: Axial CT showing temporal endocranial development with extension to the cerebellopontine angle.

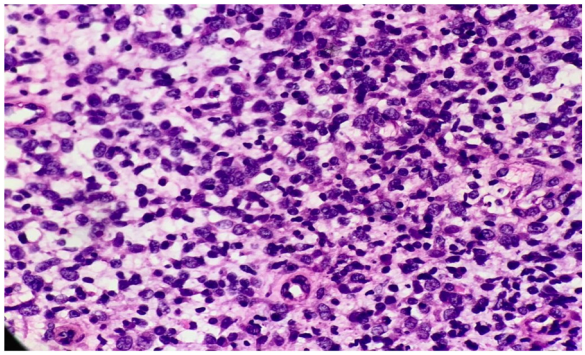

Fig. 3. Hematein and eosin stain of the tumor with a high-power view (x40) showing dense cellularity of a monomorphic, ovoid or slightly angular nuclei diffuse tumor proliferation with finely granular chromatin. The cytoplasm is scanty, ill defined. The stroma is thin including slightly tickened wall vessels.

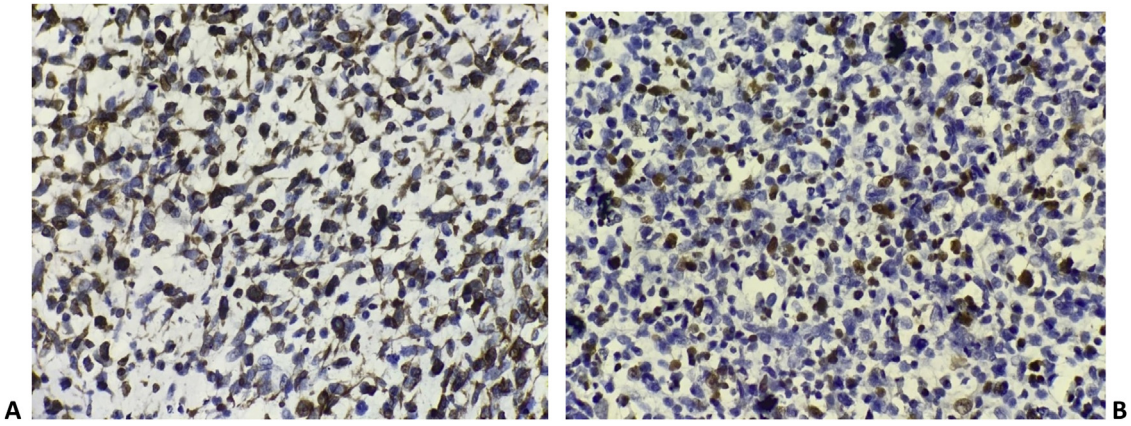

Fig. 4. Immunohistochemical staining (x40) shows cytoplasmic positivity for desmin (A) and nucleic positivity for myogenin (B).

Declaration of Competing Interest

The authors declare that they have no competing interests.

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Ethical approval

I certify that this kind of manuscript does not require ethical approval by the Ethical Committee of our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chied of this journal on request.

Author contribution

M. Beghdad: conception and design of the study
A. Mkhatri: acquisition of data
O. Berrada: drafting the article
R. Abada: revising the article
M. Mahtar: final approval of the version to be submitted

Registration of research studies

This is a case report that does not require a research registry.

Guarantor

M. Beghdad.

Provenance and peer review

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