CASE REPORT

Pediatric Rhabdomyosarcoma with Acute Otitis Media as Presenting Feature

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Abstract

Rhabdomyosarcoma is one of the most common neoplasms in pediatrics. However, despite being commonly found on the head and neck, its location in the temporal region is rare. We report a case of a 6-year-old male admitted to the emergency department with a history of recurring right otalgia, with progressive clinical worsening despite seriated antibiotics and corticotherapy. Image characterization with both computer tomography and magnetic resonance imaging were suggestive of a space occupying lesion, confirmed with an exploratory tympanomastoidectomy that revealed an obliterative polypoid lesion. The samples examination confirmed a rhabdomyosarcoma. The patient underwent combined chemotherapy and radiotherapy, initially with improvement, but clinical relapse followed by a rapid decline to death nine months after his first symptoms. This case highlights the importance of a strong clinical suspicion in children presenting with a persistence of common symptoms in pediatrics.

Keywords: Child; Ear Neoplasms; Otitis Media/diagnosis; Rhabdomyosarcoma/diagnosis; Rhabdomyosarcoma/diagnostic imaging; Rhabdomyosarcoma/drug therapy; Rhabdomyosarcoma/radiotherapy; Skull Neoplasms

Introduction

Rhabdomyosarcoma is the most common soft tissue sarcoma in childhood.[1]

It represents the third most common solid extracranial neoplasia in the pediatric age, after neuroblastoma and Wilms tumor, and has a bimodal distribution, between 2 and 6 years old and 10 and 18 years old.[1,2]

Approximately 50% of pediatric rhabdomyosarcomas occur in the cervical or head region. However, middle ear and mastoid involvement is rare, representing about 8% of the total.[3]

It typically occurs sporadically, with no known risk factors associated, but it may also be associated with familial syndromes such as Li Fraumeni and neurofibromatosis type 1.[2]

Rhabdomyosarcoma clinical presentation is variable depending on the anatomical region affected by the tumor. The involvement of the temporal region classically presents as severe otitis refractory to therapy. The most common symptoms and signs of presentation are otorrhea, cervical lymphadenopathies, hearing loss, ear polyps, or neurological changes, such as facial palsy.[4,5]

Rhabdomyosarcoma treatment has evolved considerably in recent decades. Cure rates have increased, largely due to the implementation of combination therapy. Combined chemotherapy and radiotherapy have had reports of over 70% cure rates in patients with localized disease.[2,6] However, the location in the middle ear is associated with highly aggressive behavior and a poor prognosis.[3,7]

In this case report, we present a rare presentation of a rhabdomyosarcoma, intending to create awareness regarding its diagnosis.

Case Report

A 6-year-old male, previously healthy, presented to the pediatric emergency department several times due to otological symptoms.

In the first episode, the patient was admitted with a three-day history of right otalgia, followed by homolateral facial edema and a deviation of the labial commissure to the left. He had no fever, respiratory symptoms, or headache. He also denied trauma.

Otoscopy revealed hyperemia and bulging of the right tympanic membrane, without other abnormalities. The

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diagnosis of acute otitis media complicated by peripheral facial nerve palsy was assumed. The patient was discharged with amoxicillin 90 mg/kg/day, prednisolone 2 mg/kg/day, and a follow-up appointment.

In the outpatient consultation seven days later, there was some clinical improvement but, due to the maintenance of inflammatory signs of the tympanic membrane, the antibiotherapy was changed to amoxicillin with clavulanic acid (90 mg/kg/day of amoxicillin) with the maintenance of the oral corticosteroids in a weaning scheme.

After a short period of improvement of around two weeks, the patient returned to the pediatric emergency department with a 12-hour history of right sided otalgia and pain on palpation to the pinna.

With no objective changes other than mild edema and erythema in the external auditory canal, the patient was discharged with the presumptive diagnosis of otitis externa and medicated with a topical ofloxacin.

Worsening of the symptoms led to a new visit to the pediatric emergency department six days later with recurrence of facial palsy. Otoscopy also showed bulging of the tympanic membrane in addition to signs of otitis externa. A new infection of the middle and outer ear was considered, and the patient was discharged with oral cefuroxime 40 mg/kg/day, a new course of corticosteroids, and an outpatient appointment scheduled for the following week.

In this consultation, the patient was found to be non-responsive to the prescribed antibiotics and the physical examination showed a mass in the posterior quadrants of the tympanic membrane. These findings raised the hypothesis of a cholesteatoma matrix. Considering the clinical evolution, further exams were suggested.

He underwent an analytical evaluation that revealed hemoglobin 14.3 g/dL, leukocytes 20,340 cells/µL, neutrophils 14,563 cells/µL, and reactive C protein < 0.03 mg/dL. A computed tomography (CT) of the ear was indicative of right external erosive otitis media with the total opacification of the ear and osteitis in several locations of the mastoid.

The patient was hospitalized and started on intravenous antibiotics with ceftriaxone 100 mg/kg/day and clindamycin 20 mg/kg/day as well as intravenous methylprednisolone 2 mg/kg/day and topical ciprofloxacin and dexamethasone.

For a better characterization of the CT scan findings, a magnetic resonance imaging (MRI) was performed, which revealed a bulky tumefactive lesion involving the right temporal bone (Fig. 1).

During his hospital stay, an exploratory tympanomastoidectomy was also performed, revealing a large polypoid lesion, with mastoid, pericortical, and cortical invasion.

The anatomopathological examination of the lesion biopsies and surrounding bone region confirmed the diagnosis of rhabdomyosarcoma of the right ear, and was later confirmed to be of the embryonal subtype.

Combined chemotherapy and radiation on the tumor site were started, with improvement, and three months after the initial treatment, a new mastoid biopsy was performed, with no neoplastic cells being found.

Despite the initial positive response, clinical relapse occurred one week after the end of treatment, with headaches and seizures as presenting symptoms.

Imaging exams and a lumbar puncture were completed. Cytochemical examination of the cerebrospinal fluid revealed cells with a suspected sarcomatous nature. The CT and MRI showed extensive leptomeningeal dissemination in the cranial cavity and spinal canal. Fig. 2 shows a schematic timeline of the clinical case.

Unfortunately, the patient had a rapid decline to death, approximately nine months after the first symptoms.

Discussion

The diagnosis of ear and temporal region neoplasms represent a clinical challenge, particularly due to its rarity and differential diagnosis with other more common and benign pathologies. Therefore, a high index of suspicion is required in cases that do not follow the expected course. In this case report, the initial presentation was characterized by otalgia, with otoscopic findings suggestive of acute otitis media. Upon examination, facial palsy was found, which was initially attributed to the inflammatory involvement of the facial nerve, a well described complication.[8]

Further reevaluations showed little to no clinical improvement.

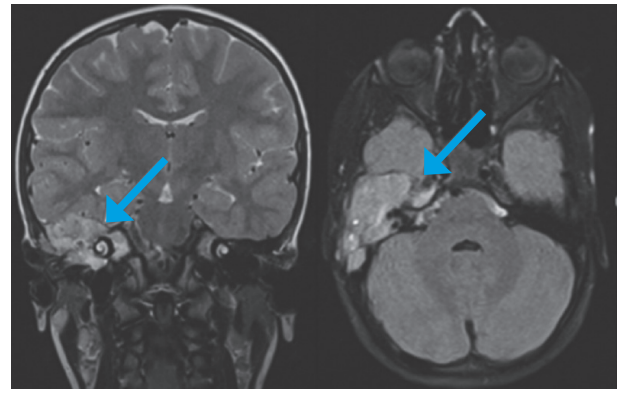
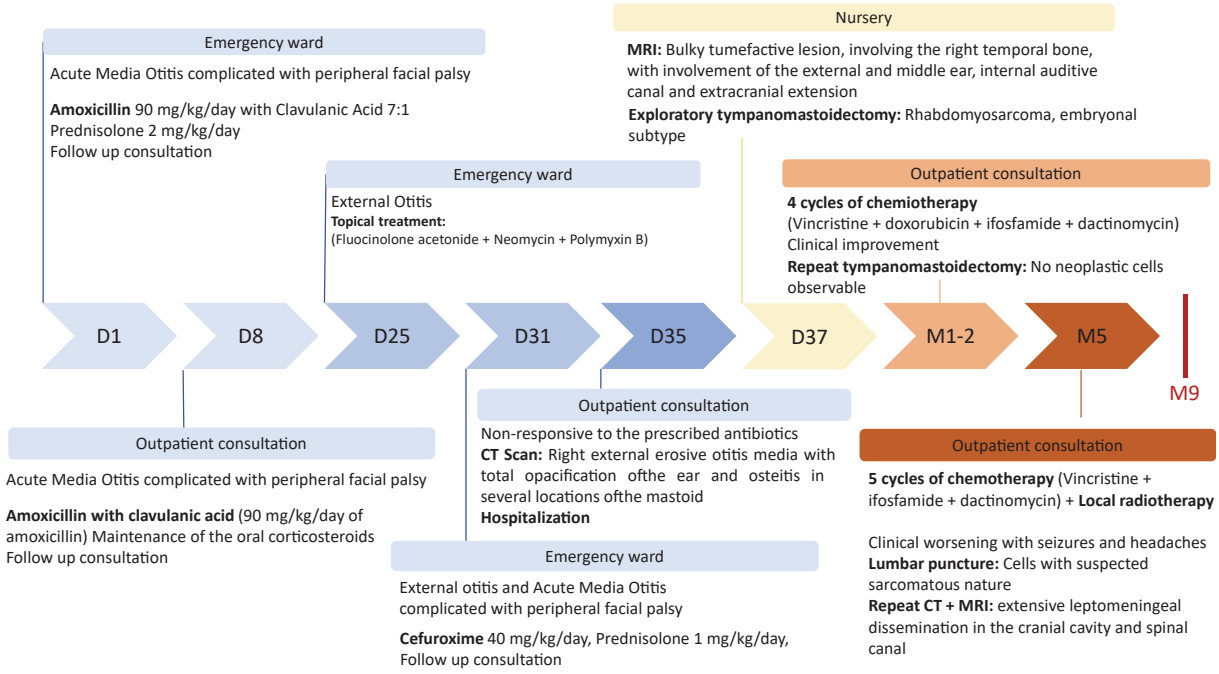


Figure 1. Coronal and transverse plane on magnetic resonance image revealing a bulky tumefactive lesion, involving the right temporal bone.



CT - computed tomography; D - day; m - month; MRI - magnetic resonance imaging.

Figure 2. Schematic timeline of the case report.

This is in accordance with retrospective studies that describe acute otitis media as not responsive to treatment as the most frequent finding prior to the diagnosis of middle ear and mastoid rhabdomyosarcoma. The most frequent signs are in decreasing order, polypoid masses (54%), otorrhea (40%), otorrhagia (39%), otalgia (22%), and facial nerve palsy (14%). The average time from first symptoms to diagnosis varies in different series, 7-21 weeks, which is also supported by our case report.[7,9]

In the presence of clinical suspicion, a diagnostic imaging test of the affected area should be performed. The most specific exam for the diagnosis of ear and mastoid rhabdomyosarcoma is contrast-enhanced MRI.[7]

A biopsy and anatomopathological evaluation of the lesion further confirms the diagnosis. Pathologic subtypes include embryonal, representing most cases of head and neck rhabdomyosarcoma, including our case report, botryoid, spindle cell, alveolar, and pleomorphic.[10]

Metastasis can occur by hematogenous, lymphatic, or contiguous destruction of the surrounding tissue. A common route is the infiltration of the facial nerve and surrounding structures. The presence of metastases should be promptly investigated after diagnosis with the lung being the most frequent site. However, there may also be hepatic, cerebral, or bone involvement.[9] Thus, the study should proceed with chest radiography, bone scintigraphy, and lumbar puncture if intracerebral involvement is suspected.[7]

The recommended treatment is the combination of chemotherapy and radiotherapy.[11]

In our case report, the patient was treated with surgical resection followed by combined chemotherapy and local radiotherapy. Initially, he had a good response with negative biopsies for tumor cells right after the first treatment cycle. However, clinical relapse occurred, with extensive leptomeningeal dissemination, which was soon followed by death.

Despite the advances achieved, the prognosis of this pathology remains reserved. Survival rates vary in different studies from 81% at five years to 41% at four years.[9]

Therefore, it is imperative to suspect the diagnosis of a middle ear tumor when facing a patient with acute middle otitis, severe enough to develop facial nerve palsy, but with no other respiratory symptoms or fever, and especially in the absence of evident clinical improvement despite the prescription of the correct therapy.

The described case report illustrates not only the diagnostic challenge and importance of high clinical suspicion, but also the aggressive nature of this neoplasm.

WHAT THIS CASE REPORT ADDS

- Despite the rhabdomyosarcoma frequency, its location on the temporal region is a rare finding, making it a diagnostic challenge.
- The involvement of the temporal region classically presents as severe otitis refractory to therapy.
- The most specific exam for the diagnosis of ear and mastoid rhabdomyosarcoma is contrast-enhanced magnetic resonance imaging.
- The recommended treatment for ear and mastoid rhabdomyosarcoma is the combination of chemotherapy and radiotherapy.
- The location in the middle ear is associated with highly aggressive behavior and a poor prognosis.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Consent for publication

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Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Otite Média Aguda como Característica de Apresentação de Rabdomiossarcoma Pediátrico

O rabdomiossarcoma é uma das neoplasias mais comuns na idade pediátrica. Contudo, apesar de ser um achado frequente na região da cabeça e pescoço, a sua localização na região temporal é rara. Descreve-se o caso clínico de uma criança de 6 anos de idade, do sexo masculino, admitida no serviço de urgência pediátrica por queixas recorrentes de otalgia direita, com agravamento progressivo, refratária à antibioterapia e corticoterapia. A caracterização imagiológica através de tomografia computorizada e ressonância magnética foi sugestiva de lesão ocupante de espaço, confirmada por timpanomastoidectomia exploratória, revelando uma lesão obliterativa polipoide. O exame histopatológico confirmou tratar-se de um rabdomiossarcoma. Foi iniciada quimioterapia e radioterapia combinada, com boa resposta inicial, mas seguida por recidiva, com declínio rápido para a morte, nove meses após a apresentação inicial. Este caso enfatiza a importância de uma forte suspeita clínica aquando a persistência de sintomas comuns na idade pediátrica.

Palavras-Chave: Criança; Neoplasias Cranianas; Neoplasias do Ouvido; Otite Média/diagnóstico Rabdomiossarcoma/diagnóstico; Rabdomiossarcoma/diagnóstico por imagem; Rabdomiossarcoma/radioterapia; Rabdomiossarcoma/tratamento farmacológico