A Toddler with Rhabdomyosarcoma Presenting as Acute Otitis Media with Mastoid Abscess

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Rhabdomyosarcoma (RMS) is a malignant tumor which involves the striated muscle, and it is most common in the pediatric age group. Usually, children with RMS present with persistent ear discharge, aural polyp and hearing loss which are similar to the symptoms seen in chronic otitis media (COM).[1,2] This similarity with COM often delays the diagnosis. The histological diagnosis of RMS is always a challenge because there are many other conditions which exhibit similar characteristic features such as an aural polyp.[3] We discussed an unusual case of RMS in a 15-month-old girl who presented with acute otitis media (AOM) and mastoid abscess. The biopsy of the aural polyp confirmed the diagnosis.

A 15-month-old girl presented with a history of right ear discharge for 1 month. Otoscopic examination was performed, and a polyp was observed. The tissue was sent for histopathological examination, and the report suggested pyogenic granuloma. Subsequently, she developed right facial asymmetry and mastoid swelling for 3 days. Clinically, the child was active, despite high-grade temperature at 38.7°C. There was a right mastoid swelling which was tender, inflamed, and fluctuant. Otoscopic examination revealed polypoidal tissue obscuring the view of the tympanic membrane. There was House-Brackmann Grade IV right facial nerve paresis. Diagnosis of right AOM with mastoid abscess was made. A high-resolution computed tomography of temporal showed extensive bony erosion of the right tegmen tympani and cortex of mastoid with soft tissue filling up the ear canal and middle ear [Figure 1]. A ring enhancing collection was observed in the soft tissue adjacent to the mastoid, and it was suggestive of an abscess formation. No hearing assessment was performed as the patient was admitted after office hours and audiology services were unavailable. She underwent examination under anesthesia and right cortical mastoidectomy with drainage of postauricular abscess which communicated with the external ear canal.

Intra-operatively, the mastoid cortex was noted to be breached with unhealthy tissue overlying it. There was extensive soft tissue filling up the entire mastoid cavity, antrum, middle ear, and external ear canal. The proximal part of the vertical portion of the facial nerve was exposed by the polypoidal tissue, and the nerve was found to be edematous. The polypoidal tissue was sent for histopathological examination.

She completed 2 weeks of intravenous Ceftriaxone and 1 week of intravenous Dexamethasone. The postauricular wound did not heal well and there was new granulation tissue observed at the inferior edge of the wound. The polypoidal tissue in the ear canal also persisted despite 2 weeks of IV antibiotic. The histopathological examination confirmed the diagnosis of embryonal RMS. Immunohistochemistry staining was positive for desmin and vimentin. This explained the poor response of the disease to antibiotics. She was referred to pediatric oncology and chemotherapy was

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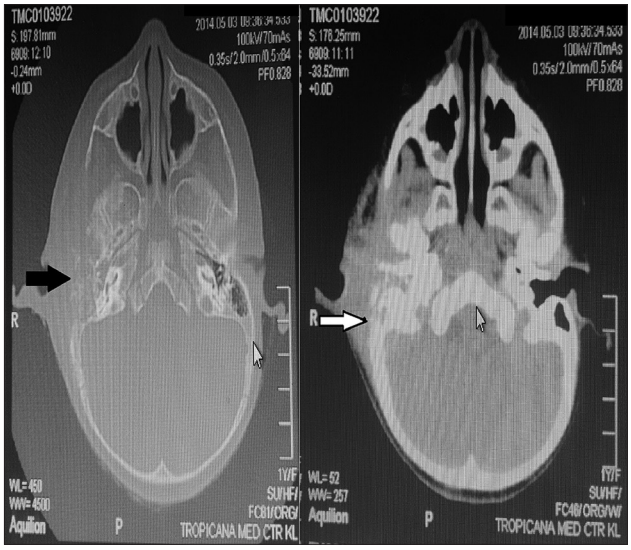

Figure 1: High-resolution computed tomography temporal of the patient showed extensive bony erosion of the right mastoid (black arrow) and abscess formation at the soft tissue adjacent to the mastoid (white arrow).

started. She recovered well and is currently in remission with normal hearing and normal facial nerve function.

RMS is a family of soft tissue tumors which is associated with skeletal muscle lineage, and it usually occurs in the pediatric population. It is the most common sarcoma in the head and neck region, accounting for 40% of all sarcomas in this region. Its site includes the orbit, oral cavity, nasopharynx, infratemporal fossa, rarely middle ear cavity, and mastoid.[4] The average age of presentation and diagnosis is at 4 years.[1,2] The age of patients ranged between 1 and 8.6 years in a cohort of 14 patients.[1] In the present case, the patient was merely 15 months old, i.e., a toddler.

The presenting complaints of RMS often mimic COM. The patients usually present with chronic aural discharge, hearing loss with aural polyp.[1,2] An early diagnosis is always a challenge because of the clinical features simulating COM. In this case, the patient presented with AOM complicated with facial nerve palsy and mastoid abscess. These were rare and unusual presentations. The clinical features seen in the present case was similar to a past research report which described symptoms such as bloody discharge, facial palsy, and lymphadenopathy.[1] Retroauricular mass, aural polyp, ipsilateral facial nerve palsy (tympanic facial nerve canal invasion), and cranial nerves V, VII, IX, XI, and XII palsies (intracerebral extension) are common features in extensive RMS.[2] In 2012, Vegari *et al*. reported a 3-year-old child with serosanguineous purulent ear discharge with aural mass for 3 weeks and biopsy revealed embryonal RMS. This showed that young children who presented with AOM and aural polyp must always be investigated carefully for malignancy, especially RMS.[5] A repeat biopsy might be needed if the initial histological result shows benign lesion as in this case.

There are many other causes of aural polyp including inflammatory polyps, cholesteatoma, chronic nonspecific inflammation, abscess, and squamous cell carcinoma, with malignancies involving two out of fifty cases.[3] Nevertheless, diagnosis of RMS relies on pathological evaluation. Therefore, repetitive biopsy is crucial to reach diagnosis, especially in children with recurrent aural polyp who do not respond to treatment. Four histological subtypes have been reported, most common being embryonal, which is also the subtype in this patient; followed by alveolar, others are pleomorphic and botryoid.[4]

RMS has a poor prognosis and in the earliest series reported in 1966, no case of survival was reported. The longest survival time was 22 months following presentation. Multimodal treatment includes multidrug chemotherapy with radiotherapy and/or surgery (international society of pediatric oncology protocols) and it leads to significant improvement regarding remission rates. Five-year survival rate was reported to vary between 41% and 81%.[2] Long-term follow-up is required to exclude recurrence and to recognize and treat all complications.

In conclusion, RMS is a rare tumor and it is associated with high mortality rate in the pediatric group, if diagnosed late. A high index of suspicion is needed to arrive at an early diagnosis. The present case illustrated a young patient with AOM, mastoid abscess, facial palsy, and aural polyp who required repetitive biopsies for the diagnosis of RMS. We opine that early diagnosis and multimodal treatment might be essential for a favorable outcome.

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Conflicts of interest

There are no conflicts of interest.

References

1. Durve DV, Kanegaonkar RG, Albert D, Levitt G. Paediatric rhabdomyosarcoma of the ear and temporal bone. Clin Otolaryngol 2004;29:32-7. doi: 10.1111/j.1365-2273.2004.00764.x.
2. Sbeity S, Abella A, Arcand P, Quintal MC, Saliba I. Temporal bone rhabdomyosarcoma in children. Int J Pediatr Otorhinolaryngol 2007;71:807-14. doi: 10.1016/j.ijporl.2007.02.002.
3. Agarwal NM, Popat VC, Traviad C, Srivastava A. Clinical and histopathological study of mass in ear: A study of fifty cases. Indian J Otolaryngol Head Neck Surg 2013;65 Suppl 3:520-5. doi: 10.1007/s12070-011-0434-0.
4. Parham DM. Pathologic classification of rhabdomyosarcomas and correlations with molecular studies. Mod Pathol 2001;14:506-14. doi: 10.1038/modpathol.3880339.
5. Vegari S, Hemati A, Baybordi H, Davarimajd L, Chatrbahr G. Embryonal rhabdomyosarcoma in mastoid and middle ear in a 3-year-old girl: A rare case report. Case Rep Otolaryngol 2012;2012:3. doi: 10.1155/2012/871235.