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Case Report on Rhabdomyosarcoma of Middle Ear in a 4 Year Old Child: A Rare Case

Solanki P*

Netaji Subhash Chandra Bose Medical College Jabalpur, India

***Corresponding author:** Preeti Solanki, Netaji Subhash Chandra Bose Medical College Jabalpur, State Cancer Institute Jabalpur, India, Tel: 9999427987; Email: drpsolanki@gmail.com

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Abstract

Introduction: Rhabdomyosarcoma accounts for up to 60% of childhood soft tissue sarcoma, occurring mostly in the head and neck region but rarely in the ear and temporal areas.

Case Report: A 4-year-old boy symptomaticaly treated at rural center for 3-4 months and then referred to our hospital due to a purulent discharge. Also seen mass in the right external auditory canal along with facial deviation to the left side. After histopathologic evaluation, it was diagnosed as embryonal rhabdomyosarcoma.

Conclusion: Rhabdomyosarcoma should be kept in mind as a differential diagnosis in all children presenting with chronic otitis media (CSOM). Biopsy should be considered who are not responding to medications and those worsening condition.

Keywords: Rhabdomyosarcoma; Chronic Otitis Media; Embryonal Rhabdomyosarcoma

Introduction

Rhabdomyosarcoma is a rare type of cancer that primarily affects children and adolescents [1]. This malignant tumor of mesenchymal origin arises from skeletal muscle lineage cells. RMS can occur in various parts of the body, including the head and neck, genitourinary tract, and extremities. However, when it develops in the middle ear of a child, things can get complicated quickly. The symptoms might go unnoticed for some time until they progress to severe levels, causing hearing loss or facial paralysis [2]. The most common sarcoma of childhood is rhabdomyosarcoma, this tumor is the third most common childhood solid tumor after neuroblastoma and nephroblastoma. Approximately 35% of all pediatric rhabdomyosarcomas occur in the head and neck. The most common site that involved by rhabdomyosarcoma is orbit (about one-third of cases). After that, in decreasing order, rhabdomyosarcoma affects oral cavity and pharynx (29%), the face and neck region (24%), involvement of the ear and temporal bone with rhabdomyosarcoma is uncommon [3]. Pathologic subtypes of rhabdomyosarcoma include: embryonal, botryoid, alveolar, pleomorphic, spindle cell, and anaplastic variants. The embryonal rhabdomyosarcoma includes about 60-70% of rhabdomyosarcoma cases .Most of the cases of rhabdomyosarcoma of middle ear present between 2-5 years of age.

Treatment plans for rhabdomyosarcoma of the middle ear typically include a combination of surgery, chemotherapy and radiation therapy [4]. In some cases, medical treatment may be tried first. However, if there is no clinical improvement, more aggressive treatment may be required. In this case, the patient underwent surgery followed by chemotherapy. The course of treatment and prognosis depend on the stage of the disease, the age and general health of the patient, and the response to treatment [1,5]. Close monitoring and follow-up are essential to ensure the best possible outcome for patients.

In this case report, we present the history of a 4-year-old boy who was diagnosed with rhabdomyosarcoma of the middle ear, an exceptionally rare disease [6]. A male patient presented with symptoms of purulent discharge and right ear and facial weakness. The diagnosis of rhabdomyosarcoma was not initially suspected, and the patient was initially diagnosed with chronic otitis media. Through this case report we want to increase awareness among healthcare professionals that rhabdomyosarcoma of middle ear can mimic CSOM.

Case Report

A 4-year-old boy presenting at rural center due to serosanguineous purulent discharge from his right ear and mild deviation of face towards left side. He was treated with antibiotics for 3-week period with attenuation in the amount of drainage. He received symptomatic treatment for 3-4 months there and then referred to our center. He came to our center with same complain. In physical examination, there was polypoidal growth in right external auditory canal and there was serous discharge from the ear canal and face deviated to left side (Figure 1). Biochemistry tests were normal. In computer tomography, soft tissue opacification of right middle ear, mastoid antrum, external auditory meatus with opacification of mastoid air cells, blunting scutum and dehiscence of tegmen tympani (Figure 2). Therefore, first diagnosis was neoplastic change and rhabdomyosarcoma was most probably because of patient age, metastatic neuroblastoma, lymphoma and leukemia was in differential diagnosis. Patient admitted and radical mastoidectomy was done [7]. During surgery external auditory canal and mastoid was full of polypoid granulomatous tissue. Samples were prepared and sent to pathology laboratory, pathologist reported malignant spindle cell rhabdomyosarcoma (Figure 3) and IHC was revealed. Immunohistochemistry showing spindle cell rhabdomyosarcoma. Then patient sent for multiagent chemotherapy and radiotherapy.

Follow-up and post-treatment monitoring are critical parts of the management of middle ear rhabdomyosarcoma in children. Patients treated for rhabdomyosarcoma of the middle ear require regular follow-up for signs of recurrence or complications [1]. Imaging studies such as CT scans and MRI may be required to assess response to treatment and detect any residual tumor [8]. In addition, audiometric testing and evaluation of facial nerve function may be warranted to monitor any long-term effects of treatment [1].

The long-term outcomes of treating middle ear rhabdomyosarcoma can have a major impact on a child's quality of life. Depending on the level of treatment required, children may develop hearing loss, facial nerve palsy, or other complications [9]. In addition, the psychological impact of cancer diagnosis and treatment can be substantial, and children may require on-going support to cope with the emotional and social impact of the disease [10]. Therefore, it is important to provide comprehensive care that meets the physical and emotional needs of children and their families.

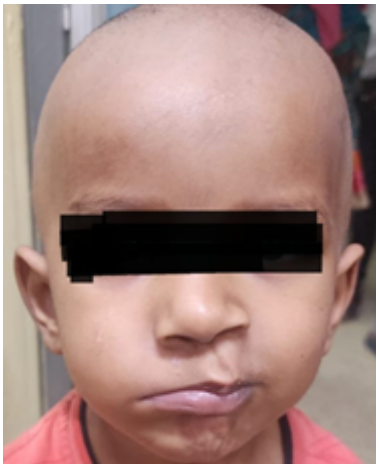

Figure 1: Polypoidal growth in right external auditory canal and there was serous discharge from the ear canal and face deviated to left side.

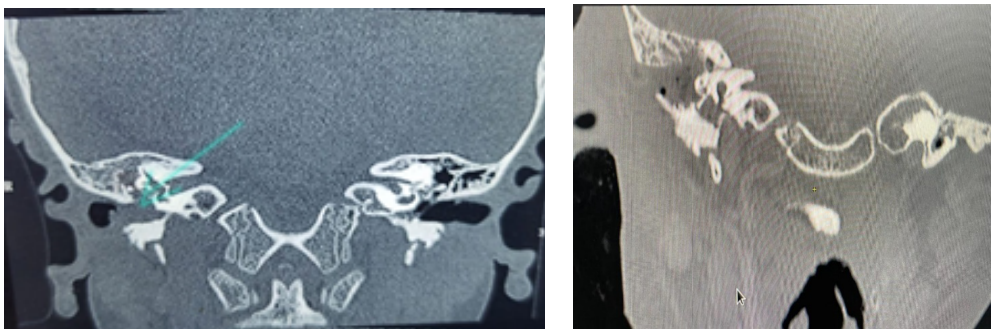

Figure 2: External auditory meatus with opacification of mastoid air cells, blunting scutum and dehiscence of tegmen tympani.

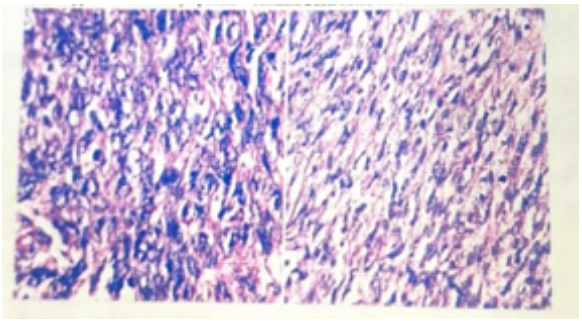

Figure 3: Malignant spindle cell rhabdomyosarcoma.

Discussion

Rhabdomyosarcoma is a rare type of soft tissue malignancy that arises from embryonic mesenchymal cells, which have the potential to differentiate into skeletal muscle cells [1]. Rhabdomyosarcoma commonly occurs in children, and it is the most common soft tissue sarcoma in this population [4]. However, rhabdomyosarcoma of the middle ear is an exceptionally rare occurrence in children, accounting for only 10% of head and neck rhabdomyosarcomas [11]. This type of cancer is often misdiagnosed as an aural polyp, leading to delayed diagnosis and treatment [1]. Therefore, it is essential to report rare cases of rhabdomyosarcoma of the middle ear in children to increase awareness and facilitate early diagnosis and treatment [12].

Rhabdomyosarcoma of the middle ear is a rare and challenging diagnosis, particularly in children. A case report by Galal A, et al. [5] described a 4.5-year-old boy with an embryonal rhabdomyosarcoma of the middle ear who presented with an external auditory canal polyp and purulent discharge [5]. Similarly, Menzies-Wilson et al. reported a case of a 4-year-old boy with a botryoid embryonal rhabdomyosarcoma of the right middle ear. These cases highlight the importance of considering rhabdomyosarcoma as a possible diagnosis in children with middle ear masses, even in the absence of typical symptoms.

Early diagnosis and appropriate treatment are critical in the management of rhabdomyosarcoma of the middle ear in children. A case report by Bhargava S, et al. [13] described an unusual case of rhabdomyosarcoma of the middle ear in an adult, highlighting the poor prognosis associated with this condition [13]. Similarly, Ragab AA, et al. [14] reported a case of a 4-year-old boy with rhabdomyosarcoma of the middle ear who was initially treated as a case of chronic ear infection [14]. Therefore, awareness and suspicion of this rare condition are essential, and prompt referral to a specialist is necessary for optimal management and improved outcomes.

In recent years, the prognosis and survival of children with rhabdomyosarcoma of the middle ear have improved due to advances in treatment. In localized disease, the 5-year overall survival rate has been improved to over 80% by the combined use of surgery, radiotherapy, and chemotherapy [15]. However, the prognosis of children with advanced or metastatic disease remains poor [16]. Therefore, early detection and timely treatment are crucial to improve the prognosis of middle ear.

Conclusion

The case report of a 4-year-old child with rhabdomyosarcoma of the middle ear is significant because it illustrates the rarity and difficulty of diagnosing this cancer. Because rhabdomyosarcoma is a malignant soft tissue disease that primarily affects children, it is important to increase awareness among healthcare professionals that it can occur in the middle ear [1]. This case report highlights the importance of early detection and diagnosis to improve the prognosis of patients with rhabdomyosarcoma of the middle ear [17]. It is important that healthcare providers remain vigilant in identifying potential symptoms and performing appropriate diagnostic testing to ensure a timely and accurate diagnosis.

Patient Consent

Consent has been taken by the parents of child for presenting this case report.

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