

Embryonal rhabdomyosarcoma of the middle ear presenting with aural polyp and facial nerve palsy

Rafiqahmed Vasiwala, MS*, Ismail Burud, MS*, Siew Kheong Lum, FRCS Ed*, Rajveer Singh Saren, MS**

*IMU Clinical School, Seremban, Negeri Sembilan, Malaysia, **Hospital Tuanku Ja'afar, Seremban, Negeri Sembilan, Malaysia

SUMMARY

Rhabdomyosarcoma is a rare tumour in the middle ear and mastoid cavity in children and the diagnosis is difficult. Repeated histological examination may be essential to confirm the diagnosis. We report a 6 year old boy with a left aural polyp, otorrhoea and facial nerve palsy who was initially thought to have otitis media and mastoiditis. He had polypectomy and the tissue taken for histopathology suggested an inflammatory condition. Subsequently he had mastoidectomy. Tissue taken during mastoidectomy was however reported as rhabdomyosarcoma. The child developed a cerebral abscess and eventually succumbed. A literature review of the disease, radiological findings, immunohistochemical features and treatment options is described.

KEY WORDS:

Rhabdomyosarcoma, mastoiditis, aural polyp, facial palsy

INTRODUCTION

Rhabdomyosarcoma, a rare malignant neoplasm originating from voluntary muscles, is the most common soft tissue sarcoma in children. About 30% of this tumour occurs in the head and neck.^{1,2} However, this tumour is rarely reported in the middle ear and mastoid. Middle ear and mastoid rhabdomyosarcoma may present with variable symptoms and signs including aural polyp, otorrhoea and facial paralysis.³ we report below a case of embryonal rhabdomyosarcoma in the middle ear and mastoid in a 6 year old boy who presented with left aural polyp, otorrhoea and facial nerve palsy. This report aims to increase awareness amongst clinicians to consider rhabdomyosarcoma in the differential diagnosis of unresolved otitis media presenting with a polypoidal mass and facial palsy.

CASE REPORT

A 6-year-old male patient presented with a history of purulent left ear discharge and a polypoidal mass in the external auditory canal of 3 weeks duration (Fig 1). He was thought to have chronic otitis media with aural polyp and treated with Unasyn 300 mg/kg/day. In view of persistent symptoms, aural polypectomy was done and the tissue sent for histopathology. The next day the patient developed left facial palsy. Computer tomography (CT) of the head and mastoid area was suggestive of mastoiditis. A soft tissue mass was seen occupying the left mastoid cavity with erosion of the

tegmen tympani (Fig 2). Histopathology of the removed polyps was reported to be of inflammatory origin. We continued with the same antibiotics.

Two weeks later the left aural polypoidal mass recurred. The child was febrile, drowsy and had headache and vomiting. Examination revealed a polypoidal fleshy mass protruding from the left external auditory meatus and the mastoid area was oedematous. Left otitis media with mastoiditis and facial nerve palsy was suspected and mastoidectomy was performed. The polypoidal mass in the middle ear and mastoid cavity was scrapped and sent for histopathology. The tegmen tympani was noted to be eroded and the dura was exposed. The facial nerve was noted to be compressed by a fleshy granulation tissue mass, stage III disease. All the granulation tissue was removed and facial nerve decompression was done.

Unfortunately, the histopathology report revealed embryonal rhabdomyosarcoma of the middle ear and mastoid. This was confirmed by immunohistochemical stain. Multiagent chemotherapy was commenced. After one cycle of chemotherapy, the patient developed high-grade fever and signs of raised intracranial pressure. Repeat CT scan confirmed a left temporal lobe abscess. The patient's condition worsened and he succumbed in the hospital.

DISCUSSION

Rhabdomyosarcoma is a rare and aggressive malignant soft tissues tumour. In children less than 15 years old, the incidence of this tumour is estimated to be at 0.44 per 100,000 in Caucasians and 0.13 per 100,000 in Afro-descendants.⁴ This tumour has a bimodal age of presentation, with peaks at 2-5 years and in adolescence. More than 60% of the cases occur in children less than 10 years of age.⁴ Horn and Enterline classified the histological variants as embryonal, botryoid, and alveolar and pleomorphic types.⁴ Approximately 60% of all newly diagnosed rhabdomyosarcomas are of the embryonal type.

Rhabdomyosarcoma in the head and neck can occur in the orbit, pterygopalatine fossa, parapharyngeal space, nasopharynx but seldom in the middle ear and mastoid.³

Embryonal rhabdomyosarcoma should be considered in a child presenting with a polyp in the external auditory meatus with recent onset of facial nerve palsy. Advanced cases may

This article was accepted: 6 July 2015

Corresponding Author: Rafiqahmed Vasiwala, IMU Clinical School, Jalan Rasah, 70300 Seremban, Negeri Sembilan, Malaysia

Email: rafiq_ahmed@imu.edu.my



Fig. 1: External auditory canal mass.



Fig. 2: CT scan showing mass in mastoid and erosion of tegmen tympani.

present with aural discharge, facial weakness and swelling in the region of the mastoid area. Extensive destruction of the petrous bone may be revealed in the CT scan.² Immunohistochemical study of rhabdomyosarcoma is important as the alveolar variant has been reported to have a poorer prognosis and a greater frequency of disseminated metastasis. In spite of primary combined modality chemotherapy, radiation therapy and surgery the prognosis is poor. Younger patients, for unknown reasons, tend to have a more favourable prognosis.

CONCLUSION

Embryonal rhabdomyosarcoma of the middle ear and mastoid is a rare neoplasm. It should be considered in the differential diagnosis in a child with an aural polyp, otitis media and recent onset of facial palsy. Biopsy of the polyp in the external auditory meatus is recommended. Initial histopathology results may be inconclusive or misleading and repeat biopsy may be indicated. Early diagnosis and multimodal therapy is desirable. However, prognosis remains poor in most cases.

REFERENCES

1. Goto TK, Yoshiura K, Tanaka T, *et al.* A follow-up of rhabdomyosarcoma of the infratemporal fossa region in adults based on the magnetic resonance imaging findings: case reports. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1998; 86(5): 616-25.
2. Viswanatha B. Embryonal rhabdomyosarcoma of the temporal bone. *Ear Nose Throat J* 2007;86(4): 218, 220-2.
3. Durve DV, Kanegaonkar RG, Albert D, *et al.* Paediatric rhabdomyosarcoma of the ear and temporal bone. *Clin Otolaryngol Allied Sci* 2004; 29(1): 32-7.
4. Rodeberg DA, Paidas CN, Lobe TL, *et al.* Surgical Principles for Children/Adolescents with Newly Diagnosed Rhabdomyosarcoma: A Report from the Soft Tissue Sarcoma Committee of the Children's Oncology Group. *Sarcoma*. 2002; 6(4): 111-22.
5. Sbeity S, Abella A, Arcand P, *et al.* Temporal bone rhabdomyosarcoma in children. *Int J Pediatr Otorhinolaryngol*. 2007; 71(5): 807-14.