The Egyptian Journal of Otolaryngology

CASE REPORT Open Access

Temporal bone rhabdomyosarcoma mimicking otitis media complicated by facial nerve palsy

Ahmed Galal[1], Omneya Ahmed[1*], Azza M. Rizk[2], Hanan Yehia Tayel[2] and Rania G. Aly[2]

Abstract

Background: Rhabdomyosarcoma is common in childhood, especially, the head and neck region, yet involvement of the temporal bone is rare.

Case presentation: We reported a case of an embryonal rhabdomyosarcoma in a 4.5-year-old boy presenting with external auditory canal polyp and purulent otorrhea that later developed grade 6 facial palsy. Imaging showed soft tissue mass involving the middle ear, mastoid cavity, parotid gland, and parapharyngeal space. Subtotal petrosectomy with blind closure of the external auditory canal was performed with facial nerve decompression and debulking biopsy followed by combined chemoradiation.

Conclusion: Middle ear rhabdomyosarcoma is a rare pathology, usually present in childhood by symptoms similar to suppurative otitis media not responding to medical treatment leading to delayed diagnosis and development of complications.

Keywords: Middle ear rhabdomyosarcoma, Temporal bone rhabdomyosarcoma, Complicated otitis media, Facial palsy, Case report

Background

Rhabdomyosarcoma (RMS) is the third most common neoplasm in childhood after neuroblastoma and nephroblastoma. It is derived from striated muscles arising in any part of the body [1]. RMS is a malignant tumor, that is highly aggressive, locally invasive, and pertaining poor prognosis [2]. It accounts for about 60% of soft tissue tumors in children most commonly between 2 and 5 years [3, 4]. About 30–50% of cases present in the head and neck area [5, 6], with only 3% presenting within the middle ear (ME) or the temporal bone (TB) which is occasionally associated with cranial nerves affection [4, 6].

Pathologic variants of rhabdomyosarcoma include embryonal, alveolar, pleomorphic, and spindle cell/sclerosing. The most common is embryonal rhabdomyosarcoma accounting for about 60–70% of the cases [7, 8]. Rhabdomyosarcomas spreads locally and metastasizes to distant sites via hematological means, and the lungs and bone are the most common sites for secondary deposits. Clinical presentation depends on the site of the tumor. Symptoms in the TB and ME are similar to the clinical picture of chronic suppurative otitis media and its complications, e.g., polypoid masses in the external auditory canal, otorrhea, ear pain with or without facial nerve palsy. This fact frequently causes a delay in diagnosis [9].

Case presentation

This was a 4.5-year-old boy who started complaining since October 2019 of left-sided purulent, profuse offensive otorrhea for 2 weeks. The patient sought medical help outside our center and received treatment in the form of systemic and aural antibiotics for 2 weeks. However, there was no improvement. Then, the patient

* Correspondence: doctoromneya@gmail.com
[1]Department of Otorhinolaryngology, Faculty of Medicine, Alexandria University, Alexandria, Egypt
Full list of author information is available at the end of the article

started to develop a postauricular swelling and torticollis. He additionally, developed left-sided acute facial palsy (HB grade 6) with irrelevant medical, family, and social history.

Consequently, he was referred to our center. His otoscopy revealed a soft tissue mass extruding through the left external auditory canal (EAC) associated with offensive, purulent discharge. The postauricular swelling was red, hot, and tender, however, was firm and not cystic (Fig. 1). No neck lymph nodes were detected. His pure tone audiometry (PTA) revealed right mild conductive hearing loss (CHL) and left moderate CHL of 45 dB ABG with bilateral type B tympanometry. The patient was admitted and received IV antibiotics. Meanwhile, due to suspicion of neoplasm based on firm swelling and the facial palsy development, imaging was ordered. CT and MRI with contrast revealed a left well-defined complex multiloculated lesion filling the ME, mastoid cavity, eustachian tube (ET), and EAC. It reached the left parotid gland with a size of 2.8×2.5×3 cm, and a left parapharyngeal extension of 3.5×2.7cm was noted (Fig. 2). Left multiple reactionary-looking enlarged upper jugular chain LN was also observed.

Informed and written consent for surgery and for case reporting was obtained from the patient's guardian (the father). Surgery was scheduled urgently. It was performed threefold. First to provide a generous biopsy of the mass, second to stop the discharge, and third to decompress the FN. Therefore, the decision was to perform a subtotal petrosectomy via postauricular incision with blind end closure of the EAC and plugging of the ET. The tumor was found intra-operatively to be filling the mastoid cavity, retro-facial air cell tract. This area was the most suspected place of facial nerve invasion causing facial palsy since the deep surface of the mastoid segment of the FN was uncovered and partially invaded by the tumor. The tumor also filled ME, ET, and EAC. The postauricular swelling was found to be formed of tumor tissue and was excised. Maximum debulking was performed. Nonetheless, complete tumor resection was neither achieved nor planned. The FN was decompressed from geniculate ganglion to stylomastoid foramen (Fig. 3).

Biopsy was sent to our pathology department and revealed an embryonal rhabdomyosarcoma, botryoid variant which was confirmed by positive nuclear staining reaction for myogenin (Fig. 4). The patients' wound healing was uneventful, and the discharge stopped. Four months later, FN improved to HB II. After surgery, the patient was sent for consultation at our Medical Oncology Department. A scan for metastasis revealed none (CT chest, ultrasound abdomen, and bone scan). The patient started within a month his therapy. He started with 3 chemotherapy cycles in the form of IVA (Ifosfamide, Vincristine, Actinomycin). This led to regression that was incomplete. Therefore, a concomitant radiochemotherapy was decided. This was in the form of additional 6 chemotherapy sessions and 30 radiotherapy sessions. These chemotherapy sessions were administered without actinomycin due to synergistic toxicity with radiation. The patient is currently alive and well, and imaging showed a regressive course. At the time of writing this work 16 months after presentation, the patient has no complaints and follow-up MRI showed remission and a stationary course.

Discussion

Rhabdomyosarcoma is a common soft tissue tumor in the pediatric age group. It possesses a bimodal pattern of age distribution, a peak between 2 and 5 years and another peak in late adolescence [10]. About 63% of cases are under 10 years [11].

Rhabdomyosarcoma in the head and neck affects the orbit, pterygopalatine fossa, parapharyngeal space, and

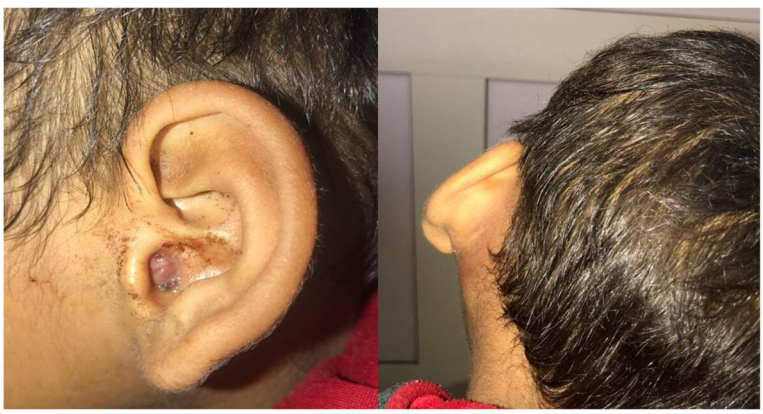

Fig. 1 On the left showing the left middle ear polyp with surrounding active discharge, the right figure shows left post-auricular and parotid swelling elevating the auricle

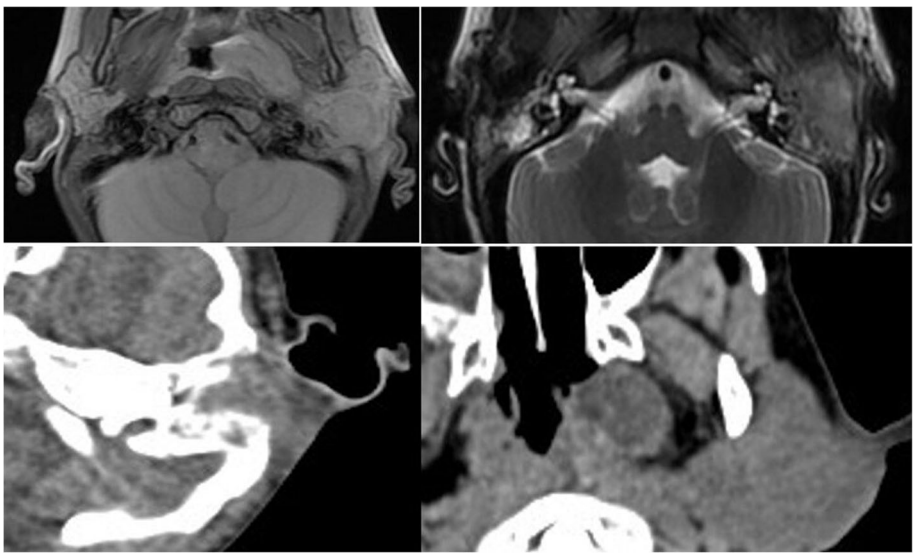

Fig. 2 Pre-operative imaging of the patient. Upper left: Axial MRI T1 with no contrast showing the mass of the left side occupying both the parotid region and the parapharyngeal space. Upper right: Axial MRI T2 at the level of internal auditory canal showing the mass in the left mastoid and middle ear region. Lower left: axial CT bone window of the left petrous bone showing total opacification by the mass of the external and middle ears and mastoid cavity and its erosion. Lower right: Axial CT bone window at the level of the nasopharynx showing left parapharyngeal and parotid region affection by the mass

nasopharynx and rarely affects the middle ear and mastoid. Signs and symptoms of this area are similar to chronic suppurative otitis media and its complications, leading to delayed diagnosis [11]. Clinical features include purulent otorrhea, hypoacusis, otalgia, aural polyp, and granulations and, in advanced cases, neurologic symptoms such as facial palsy [8].

In case of a child presenting with an aural polyp, discharge with or without recent onset of facial nerve palsy, RMS should be suspected [12]. According to these scenarios, ME rhabdomyosarcoma usually is treated by antibiotics and only when this treatment fails, management strategy would be shifted to tissue biopsy and histopathological examination [13].

Pathological examination of rhabdomyosarcoma usually shows round cell neoplasm, and immunohistochemistry is required to differentially diagnose it from other small round cell neoplasms, mainly lymphoma (CD45-

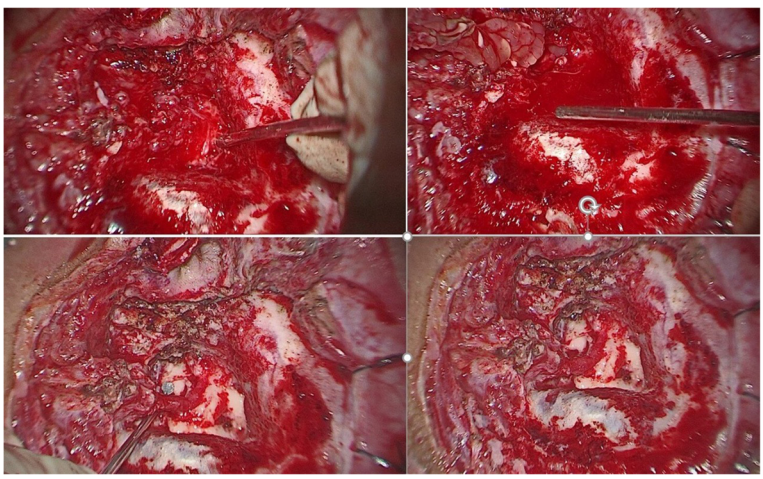

Fig. 3 Upper left: after partial mastoidectomy and debulking, tumor was bloody filling the middle ear. Upper right: tumor filling the eustachian tube being removed with cumbersome bleeding in the cavity. Lower left: after the completion of the debulking, showing instrument passing in the retro-facial tract where tumor most likely affected the facial nerve. Lower right: final cavity before closure with maximal debulking performed and bleeding stopped

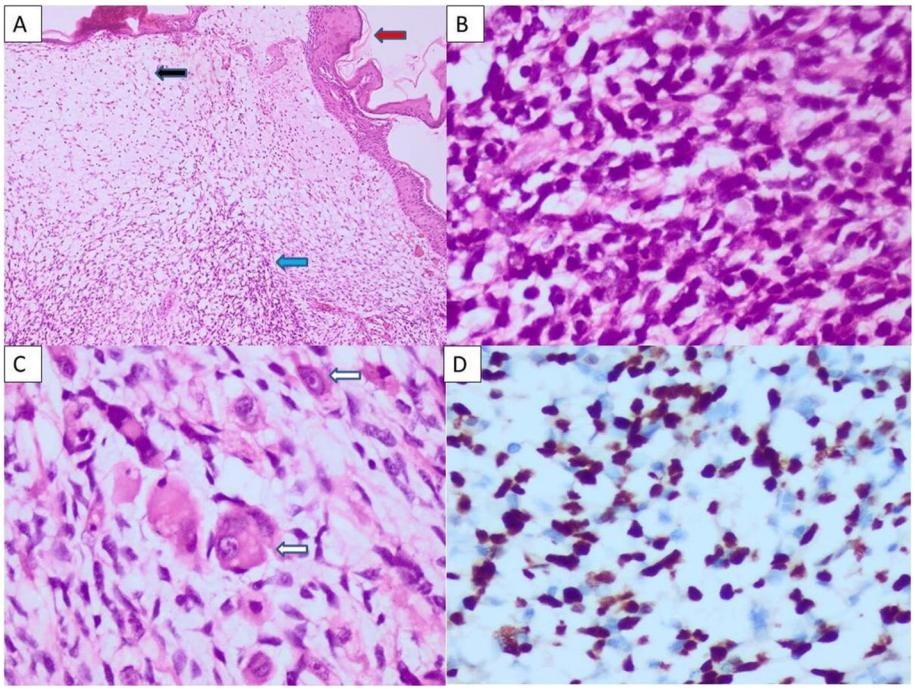

Fig. 4 The figure shows stratified squamous epithelium (red arrow) with an underlying neoplastic growth formed of alternating hypocellular myxoid areas (black arrow) and hypercellular areas (blue arrow) (**A**, H&E ×100). The neoplastic cells are predominantly primitive small hyperchromatic round and spindled cells (**B**, H&E, ×400) with few scattered rhabdomyoblasts (white arrow) (**C**, H&E, ×400). Immunohistochemistry for myogenin (L026, Leica biosystems, USA) shows strong and diffuse nuclear staining of the neoplastic cells (**D**, ×400)

positive) and Ewings sarcoma (CD99-positive). Rhabdomyosarcoma is diagnostically positive for myogenin myoD1 [13].

The treatment for rhabdomyosarcoma has been a controversial issue. Before 1972, the mainline of treatment was surgical extirpation followed by radiotherapy till the Intergroup Rhabdomyosarcoma Study (IRS) protocol was brought out, which advised the multi-agent chemoradiation as the first line of treatment and surgery only is preserved to provide tissue biopsy, stop discharge, or when FN decompression is required [14].

Conclusion

Rhabdomyosarcoma is a common pediatric neoplasm, however, rare in the middle ear and mastoid cavity. Its presentation is deceiving in this region, since it would mimic complicated otitis media presenting with polyps, discharge which is not responding to medical treatment, and occasionally facial palsy. This might lead to delayed diagnosis until metastasis has occurred which might worsen the prognosis. Rapid progression, neurological signs, and a firm postauricular swelling might be suggestive of a neoplasm requiring CT. In turn, if CT is suspicious, an MRI should be performed. The role of surgery in these neoplasms is only biopsy, debridement, stoppage of discharge, and decompression of the facial nerve when required and not radical excision with curative intent.

Abbreviations

EAC: External auditory canal; CT: Computed tomography; MRI: Magnetic resonance imaging; ME: Middle ear; HB: House-Brakmann facial nerve paralysis scale; RMS: Rabdomyosarcoma; TB: Temporal bone; PTA: Pure tone audiometry; CHL: Conductive hearing loss; ABG: Air bone gap; ET: Eustachian tube; FN: Facial nerve; IRS: Intergroup Rhabdomyosarcoma Study

Acknowledgements

Not applicable.

Authors' contributions

AG formulated the idea of reporting analyzed and interpreted the patient data, OA collected the operative data, followed-up the case, and was a major contributor in writing the manuscript; AR and HT performed and revised the histopathological diagnosis; and RG did the pathology figures and approved of the final version. The authors read and approved the final manuscript.

Funding

None.

Availability of data and materials

All data and material of this case are available.

Declarations

Ethics approval and consent to participate

The institutional ethical board review was obtained and approved for publication (Faculty of Medicine, Alexandria University, serial number 0304759).

Consent for publication
Informed written consent for surgery, for reporting the case, and for publication were obtained in native language from the guardian of the patient (the father).

Competing interests
The authors declare that they have no competing interests.

Author details
[1]Department of Otorhinolaryngology, Faculty of Medicine, Alexandria University, Alexandria, Egypt. [2]Pathology Department, Faculty of Medicine, Alexandria University, Alexandria, Egypt.

Received: 11 February 2021 Accepted: 10 June 2021
Published online: 13 July 2021

References

1. Ragab AA, Fakoury MMA, Kassouma J, Moustafa K, Al Salem FAM (2018) Botryoid rhabdomyosarcoma in mastoid and middle ear in a 4-year-old boy: A rare case report. Hamdan Med J. 11(3):130. https://doi.org/10.4103/HMJ.HMJ_6_18
2. Young J, Miller RW (1975) Incidence of malignant tumors in US children. J Pediatr. 86(2):254–258. https://doi.org/10.1016/S0022-3476(75)80484-7
3. Carli M, Guglielmi M, Sotti G (1997) Paediatric Oncology. Chapman and Hall, London
4. Durve D, Kanegaonkar R, Albert D, Levitt G (2004) Paediatric rhabdomyosarcoma of the ear and temporal bone. Clin Otolaryngol Allied Sci. 29(1):32–37. https://doi.org/10.1111/j.1365-2273.2004.00764.x
5. Rao BN, Santana VM, Fleming ID, Pratt CB, Shapiro D, Fontanesi J, Kumar APM, Austin BA (1989) Management and prognosis of head and neck sarcomas. Am J Surg. 158(4):373–377. https://doi.org/10.1016/0002-9610(89)90136-0
6. Wells S (1984) Embryonal rhabdomyosarcoma of the ear: a review of the literature and case history. J Laryngol Otol. 98(12):1261–1266. https://doi.org/10.1017/S0022215100148376
7. Pizzo PA, Poplack DG (2015) Principles and practice of pediatric oncology. Lippincott Williams & Wilkins, Stanford University
8. Vegari S, Hemati A, Baybordi H, Davarimajd L, Chatrbahr G (2012) Embryonal rhabdomyosarcoma in mastoid and middle ear in a 3-year-old girl: a rare case report. Case Rep Otolaryngol. 2012:871235
9. Hayes S, Biggs T, Bird J, Hellier W (2019) Rhabdomyosarcoma of the middle ear cavity presenting with Horner's syndrome. Ann R Coll Surg Engl. 101(1):e8–e10. https://doi.org/10.1308/rcsann.2018.0157
10. Abbas A, Awan S (2005) Rhabdomyosarcoma of the middle ear and mastoid: a case report and review of the literature. Ear, Nose Throat J. 84(12):780–784. https://doi.org/10.1177/014556130508401214
11. Shirani S, Alizadeh F, Sharif KS (2003) Rhabdomyosarcoma of the middle and inner ear
12. Vasiwala R, Burud I, Lum SK, Saren RS (2015) Embryonal rhabdomyosarcoma of the middle ear presenting with aural polyp and facial nerve palsy. Med J Malaysia. 70(5):314–315
13. Parham DM, Ellison DA (2006) Rhabdomyosarcomas in adults and children: an update. Arch Pathol Lab Med. 130(10):1454–1465. https://doi.org/10.5858/2006-130-1454-RIAACA
14. Viswanatha B (2007) Embryonal rhabdomyosarcoma of the temporal bone. Ear, Nose Throat J. 86(4):218–222. https://doi.org/10.1177/014556130708600413

Publisher's Note