Embryonal Rhabdomyosarcoma of External Ear - A Rare Case Report

Ananth Nag Jakkula, Tarun Gogineni[1], Srikanth Damera, P. S. S. Meghana Sri Ravali

Department of Oral and Maxillo-Facial Surgery, G.S.L Dental College and Hospital, Affiliated to G.S.L Cancer Trust, [1]Surgical Oncologist, G.S.L Cancer Trust, Andhra Pradesh, India

Abstract

The Rationale: Rhabdomyosarcoma (RMS) is an uncommon tumour which presents at a wide variety of different head and neck sites. They are highly aggressive malignant mesenchymatous soft tissue sarcomas. Their occurrence in the outer ear is extremely rare. **Patient Concerns:** A 17-year-old female patient reported with a chief complaint of steady increase in growth over the left ear since 6 months, which is not associated with pain and ulceration. **Diagnosis:** Edge biopsy sections of the swelling over the left pinna and immunohistochemistry records of cells show MYODI positive confirming as Embryonal RMS. **Treatment:** Left auriculectomy and supra-omohyoid neck dissection with level V lymph node was performed. **Outcomes:** Uneventful postoperative healing in the resected site was elicited in the follow-up period of 1 year with no recurrence. **Take-away Lessons:** External ear RMS is extremely rare. Careful clinical, radiological and histopathological evaluation are essential for prompt definitive diagnosis and for successful management.

Keywords: Adolescent, ear, neck dissection, rhabdomyosarcoma, sarcoma

Introduction

Sarcomas represent only 1% of primary tumours within the head and neck region.[1] Rhabdomyosarcoma (RMS) is a highly aggressive malignant mesenchymatous soft tissue tumour. It arises mainly from the immature striated skeletal muscle cells and sometimes from nonstriated regions.[2] Common sites include genitourinary tract, retroperitoneum, head and neck, and extremities.[3]

The incidence of Head and Neck RMS (HNRMS) is very rare with a range of 4.6 cases/1 million children.[4] It accounts for about 5% of all paediatric malignant neoplasms. The main locations for HNRMS were orbit (33.3%), oral cavity and pharynx (29%) neck (24%).[5] Involvement of external ear, mastoid, and temporal regions is extremely rare.[6] If it involves the ear, most commonly noted location was the middle ear cavity.

HNRMS pathologic subtypes include embryonal (classic embryonal, botryoid, and spindle cell/sclerosing variants), alveolar, and pleomorphic/anaplastic. Frequent histopathological forms of the paediatric group are embryonal and alveolar with pleomorphic variant in adults.

Access this article online

Quick Response Code:

Website: www.amsjournal.com

DOI: 10.4103/ams.ams_85_21

This is a rare case report of an embryonal RMS of the left external ear in a 17-year-old female patient.

Case Report

This 17-year-old female patient reported to the Department of Oral and Maxillofacial Surgery with a chief complaint of a growth over the left ear since 6 months. There was a steady increase in the size of the lesion on the left pinna with no associated pain or ulceration.

There was also no history of fever, discharge, otalgia, otorrhoea, auditory disturbances, tinnitus, vertigo, facial weakness, dysphagia, odynophagia, easy bleeding or weight loss. There

Address for correspondence: Dr. P. S. S. Meghana Sri Ravali, Department of Oral and Maxillo-Facial Surgery, G.S.L Dental College and Hospital, (Affiliated with G.S.L Cancer Trust), Rajamahendravaram - 533 296, Andhra Pradesh, India.
E-mail: meghana.puranapanda@gmail.com

Received: 28-03-2021 **Last Revised:** 26-10-2021
Accepted: 09-11-2021 **Published:** 01-02-2022

How to cite this article: Jakkula AN, Gogineni T, Damera S, Sri Ravali PS. Embryonal rhabdomyosarcoma of external ear - A rare case report. Ann Maxillofac Surg 2021;11:317-20.

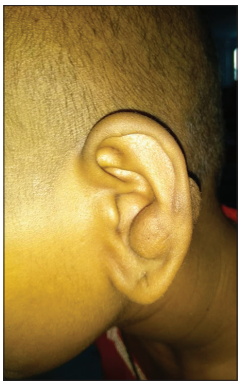

Figure 1: Extraoral swelling extending onto cavum, anti-tragus and inter tragic notch of left auricle

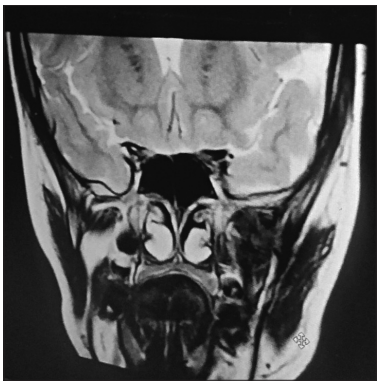

Figure 2: Coronal section of contrast enhanced computed tomography

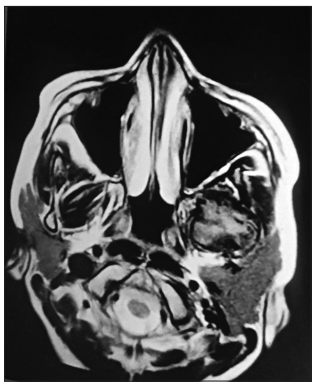

Figure 3: Axial section of contrast enhanced computed tomography

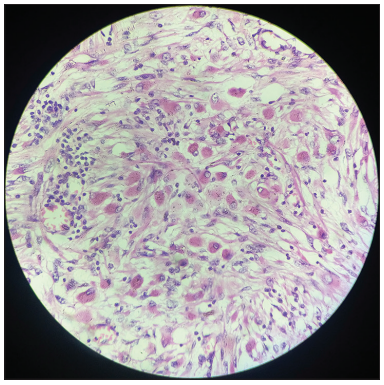

Figure 4: Histopathological specimen revealed bundles of fibro-collagenous tissue with an invasive tumour composed of epithelioid/spindle shaped cells arranged in the form of compact interlacing bundles with intervening desmoplastic stroma, thin-walled blood vessels and large lymphoid aggregates

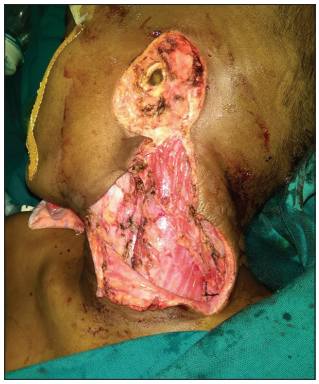

Figure 5: Auriculectomy with supraomohyoid neck dissection along with level V lymph node on left side

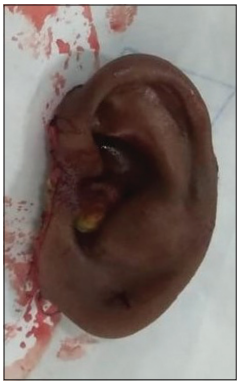

Figure 6: The excised auricle

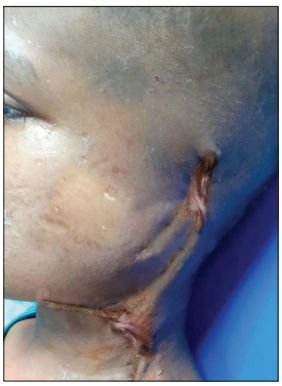

Figure 7: Postoperative healing in the operated site after 1 year

was no relevant medical or personal history. The patient gives a history of five cycles of chemotherapy associated with vincristine, actinomycin, and cyclophosphamide.

On presenting to OMFS Department, a swelling of size 3 cm × 3 cm was seen extending onto the cavum, antitragus, and intertragic notch. It was soft in consistency with a smooth surface [Figure 1]. Central induration and indentations were present. There was no tenderness or local rise in temperature.

Contrast-enhanced computed tomography (CT) of head and neck illustrated a heterogeneously enhancing lesion measuring 2.6 cm × 1.6 cm × 4.7 cm (Antero - posterior × Transverse

or Medio - lateral × Superio - inferior) noted in the left external ear involving the auricle (helix, antihelix, concha, and lobule). Posteriorly, it was obliterating the retro-auricular groove. Anteriorly, it was abutting the external auditory canal. However, there was no intraluminal extension. Inferiorly, it was extending along the subcutaneous plane and abutting the superficial lobe of parotid gland medially with ill-defined intervening fat planes. There was no involvement of middle and inner ear. There was also no significant bony erosion [Figures 2 and 3]. There were however homogenously enhancing subcentimetric short-axis bilateral level IIA nodes.

Edge biopsy sections of swelling over left pinna showed bundles of fibrocollagenous tissue with an invasive tumour composed of epithelioid/spindle-shaped cells displaying round to oval/spindle-shaped vesicular nuclei and moderate eosinophilic cytoplasm. These cells were arranged in the form of compact interlacing bundles with intervening desmoplastic stroma, thin-walled blood vessels, and large lymphoid aggregate. Mitotic activity was brisk and atypical. There was involvement of the margins. Immunohistochemistry tests revealed that tumour cells were CK negative, HMB 45 negative, and MYODI positive with intense nuclear staining confirming as embryonal RMS [Figure 4].

Treatment plan included wide surgical excision under general anaesthesia. Left supraomohyoid modified functional neck dissection was done along with level V lymph node. Left auriculectomy was done [Figure 5]. Specimens were then sent for histopathological examination [Figure 6].

Excisional biopsy revealed spindle cell variant of embryonal RMS. All surgical resection margins were free from tumour invasion.

The postoperative healing in the resected site was uneventful in the follow-up period of 1 year with no recurrence and the patient has been scheduled for prosthetic reconstruction of ear [Figure 7].

Discussion

Sarcomas are a heterogeneous group of malignancies. Osteosarcoma is the most common sarcoma in the head and neck region among adults. Weber first described RMS in 1854 and Stout described its morphology in 1946 based on its histology.[6] Paediatric RMS occurs with a predilection in the head and neck sites, whereas adult RMS occurs predominantly in the extremities.[7] Barnes noted that the tongue and palate are the most commonly affected intraoral sites of RMS. The clinico-epidemiological analysis on the Nigerian population by OA Fatusi, identified the palate and cheek as the most common intraoral sites.[8]

RMS have a bimodal pattern of age distribution. Their incidence peaks between the ages of 2 and 5 years and spikes again in late adolescence. Overall, 63% of all cases occur in patients younger than 10 years. Caucasians and males have slightly more predilection. Embryonal RMS is the third most common neoplasm in childhood after neuroblastoma and nephroblastoma in clinical sites of nasopharyngeal mucosa, inner ear, urogenital tract, and digestive apparatus. The alveolar type has consistently been marked by a translocation of chromosomes (2; 13) (q35qI4); in the embryonal type, the maternal allele is lost.[6]

RMS arises because of regulatory disruption of skeletal muscle progenitor cell growth and differentiation. Studies have suggested that distinct genetic factors are involved in the development and tumour progression of RMS, such as loss of heterozygosity, specific chromosomal translocations, and abnormal gene alterations.[9]

Histopathological tests, immunohistochemical staining, Computed tomography (CT), magnetic resonance image, positron emission tomography-CT, electron microscopy, and molecular genetic studies are major avenues of the diagnosis of HNRMS.[7] Immunohistochemistry is required to differentially diagnose it from other small round cell neoplasms, namely lymphoma (CD20, CD3 positive) and Ewing's sarcoma/primitive neuroectodermal tumour (CD99 positive) as RMS is negative for these markers.[9] The RMS tumour cells express Desmin, Myogenin, CD56, muscle-specific actin, Myoglobin, Vimentin, and MyoD1. The demonstration of foetal haemoglobin in tumour tissue by immunoperoxidase staining has been shown to be reliably accurate in identifying RMS.

The standard treatment for RMS is multimodal including excisional surgery, multidrug chemotherapy, and external beam radiation therapy. External beam radiation therapy has many side effects due to the collateral damage to surrounding anatomical structures. In the head and neck region, it can result in hearing loss, facial hypoplasia, developmental delay, xerostomia, dental problems, problems with the pituitary/hypothalamic hormones, and secondary malignancies.[8]

Unless diagnosed after the cancer has spread to distant parts of the body (metastases), surgery is usually the first step in treating RMS. Complete resection of the primary tumour along with some surrounding normal tissue is the goal for excision. Removal of the cervical lymph nodes through neck dissection should be tailored with primary tumour excision to reduce risk and improve prognosis through prevention of nodal spread.

The chemotherapeutic elements of the First Intergroup RMS Study IV protocol are significantly successful with etoposide, ifosfamide, and melphalan which have been added to the standard treatment with vincristine, actinomycin, and cyclophosphamide. They eradicate metastasized tumour cells by travelling throughout the body.[7]

RMS can spread locally, regionally or distantly. The common site of metastatic spread are the lungs, followed by bone marrow, lymph nodes, and bones. At least 15% of children and adolescents with RMS present with distant metastases. Thirty-one percent of the embryonal subtype had metastasis. Based on HNRMS in children, metastatic disease at diagnosis (33% of all cases) occurred in the bone

marrow (11%), cerebrospinal fluid (6%), peritoneal fluid (6%), lungs (4%), parietal pleura (2%), pleural fluid (2%), and pericardial fluid (2%). For HNRMS in adults, the rate of cervical lymph node involvement was 28%.[7]

Newton *et al*. proposed a classification based on the prognostic value of histologically recognizable forms of RMS. Botryoid and spindle cell RMS are associated with a superior prognosis, conventional embryonal RMS with an intermediate prognosis, and alveolar RMS (all types) with a poorer prognosis. For HNRMS, the overall survival rate was 28.7% in 5–10 years. The recurrence rate was 18.9%.[10]

Conclusion

This case demonstrates the presentation of embryonal RMS at an unusual location of the external auricle which was managed successfully.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Patra S, Trivedi P, Shah A. Primary sarcoma of the head and neck: Three years' experience in a tertiary care cancer center. Asian J Oncol 2021. Vol. 00 No. 0/2021 10.1055/s-0041-1728219.
2. Chirat M, Dainese L, Fasola S, Couloigner V, Denoyelle F, Garabedian EN, *et al.* Unusual outer ear swelling: Childhood auricular rhabdomyosarcoma. Eur Ann Otorhinolaryngol Head Neck Dis 2016;133:23-6.
3. Ragab AA, Abdulber Fakoury MM, Kassouma J, Moustafa K, Al Salem FA. Botryoid rhabdomyosarcoma in mastoid and middle ear in a 4-year-old boy: A rare case report. Hamdan Med J 2018;11:130-3.
4. Crozier E, Rihani J, Koral K, Cope-Yokoyama S, Rakheja D, Ulualp SO. Embryonal rhabdomyosarcoma of the auricle in a child. Pediatr Int 2012;54:945-7.
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;2012:871235.
6. Alex RA, Nayak DR, Devaraja K, Reddy NA, Singh R, Shetty S. Head-and-neck rhabdomyosarcoma: Our experience. Ann Indian Acad Otorhinolaryngol Head Neck Surg 2018;2:1-4.
7. Chen E, Ricciotti R, Futran N, Oda D. Head and neck rhabdomyosarcoma: Clinical and pathologic characterization of seven cases. Head Neck Pathol 2017;11:321-6.
8. Barnes L.Tumours and tumor like lesions of the Soft Tissues In Surgical Pathology of the Head and Neck, Vol l(Barnes Led), Marcel Dekker Inc, New York 1985. p.725-880.
9. Bhargava S, Grover M, Mehta J, Maheshwari V. Rhabdomyosarcoma in middle ear of an adult: A rare presentation. J Surg Case Rep 2012;2012:9.
10. Newton WA Jr., Gehan EA, Webber BL, Marsden HB, van Unnik AJ, Hamoudi AB, *et al.* Classification of rhabdomyosarcomas and related sarcomas. Pathologic aspects and proposal for a new classification – An intergroup rhabdomyosarcoma study. Cancer 1995;76:1073-85.