

Case Report

Rhabdomyosarcoma in a Female Adult a Rare Case Report at BMC Mwanza

Olivia Michael Kimario^{1*}, Leonard Washington², Avelline Aloyce³, Fidelis Mbunda⁴, Alex Donasiano¹, Zephania Saitabau², James Komanya⁴

¹Department of ENT CUHAS P.O. Box 1464 Mwanza, Tanzania

²Department of General Surgery CUHAS p.o.box 1464 Mwanza, Tanzania

³Department of ORL MUHAS P.O. Box 65001 Dar es Saalam, Tanzania

⁴Department of Surgery University of Dodoma P.O. Box 259 Dodoma, Tanzania

Article History

Received: 23.06.2021

Accepted: 26.07.2021

Published: 03.08.2021

Journal homepage:

<https://www.easpublisher.com>

Quick Response Code

Abstract: Rhabdomyosarcoma (RMS), is a neoplasm of skeletal muscle in origin, is the second most common soft tissue sarcoma encountered in childhood after osteosarcoma. The common sites of involvement are the head and neck region, genitourinary tract, retroperitoneum, and to a lesser extent, the extremities. Site predilections is the oral cavity which are soft palate, maxillary sinus and alveolus, posterior mandibular region, cheek and lip, and sometimes tongue. RMS is highly malignant tumor with extensive local invasions and early hemorrhagic and lymphatic dissemination. We present a case of 25yrs old lady came to our emergency department with history of swelling of the right side of the face and protrusion of the right eye. Patient was also pregnant at 38 weeks of gestation. C/s was done and later on the lateral rhinotomy with extended weber fergusson was done followed by radiochemotherapy.

Keywords: Rhabdomyosarcoma, Female Adult, muscle.

Copyright © 2021 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution **4.0 International License (CC BY-NC 4.0)** which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

Rhabdomyosarcoma (RMS) is a rare, soft tissue malignancy. The aetiopathogenesis of RMS is uncertain, although it is thought to develop from immature cells of the skeletal muscle lineage [1]. RMS is typically found in children and in adulthood is very rare [2]. The presenting symptoms and signs of RMS are varied and largely depend on the site of origin of the primary tumour, patient age and the presence or absence of metastases [3].

Around 40-50% RMS involve head and neck region. In the head and neck region, it can either arise from parameningial sites (nasal fossa, nasopharynx, paranasal sinuses, middle ear) or non parameningial site (oral cavity, oropharynx, larynx and neck). RMS arising from parameningial sites have a propensity to spread intracranially and have the worst prognosis [4, 5]. There are four subtypes:- alveolar, embryonal (most common), pleomorphic and botryoid. In adults, the most common type seen is alveolar, which has the worst prognosis.

Clinically, the manifestations of RMS may vary from a small cutaneous nodule on the face to an

extensive fast-growing facial swelling, which may be painless or occasionally associated with pain, trismus, paresthesia, facial palsy, and nasal discharge [6].

The histogenesis of RMS is still unclear, but the most widely accepted hypothesis is that RMS arises due to the proliferation of embryonic mesenchymal tissue [7].

There is a slight predilection for disease in males have been reported.[7,8] The survival rate of patients with this tumor ranged from 20% to 35% in reported series.[9]

CASE REPORT

A 25yrs old lady presented at our ent clinic with main complains of swelling of the right side of the face, bilateral nasal obstruction and protrusion of the right eye for 3 months. Also reports of episodes of nasal bleeding and severe headache. This problems are associated with nasal speech. Patient was also pregnant at 38 weeks of GA. No history of such diseases in the family and also no history of smoking or taking alcohol nor been expose to radiation. Review of other systems were essentially normal.

General Examinations

Alert,conscious,cachexic,Protrusion of the right eye,not pale ,not cyanotic,not jaundice, no enlarged lymphadenopathy,slightly swelling of the right side of the face.

ENT examination

Anterior rhinoscopy- mass seen in the right nasal cavity,redish and gray in colour.It bled on touch.

Oral cavity examination

Bluish laceration on both palates,bulging of the right side of the palate.

Ear examination: Normal

Systemic examination: Normal

We had a working diagnosis of Right maxillary tumor with Pregnancy at term in GA of 38 weeks.

Patient was planned for C/S an the baby was delivered healthly then later on lateral rhinotomy with extended weber fergusson with orbital exenteration was done.The mass was taken for histopathology.

Histology Results Reveils

Alveolar Rhabdomyoma but lymphoma was not ruled out so the pathologist recommended; immuno-histochemistry with CD45 and DESMIN Immuno-histochemistry results: CD 45 Negative; Desmin Positive. So the mass was rhabdomyosarcoma alveolar type.

DISCUSSION

The incidence of RMS is the highest in children aged 1–4 years, lower in children aged 10–14 years, and lowest in those aged 15–19 years. These tumors exhibit a fast and aggressive growth, reaching large dimensions, and are generally painless associated with high rates of recurrence and generalized metastases through the hematogenic and/or lymphatic routes [10, 11].

RMS is a rare, soft tissue malignancy of a mesenchymal origin, suspected to arise from cells of the skeletal muscle lineage [12]. RMS is very uncommon in adults and constitutes only 3% of all soft tissue sarcomas [12]. The commonest site of metastasis is the lungs followed by bone marrow and lymph nodes, as demonstrated in our case. Most primary adult RMSs occur in the extremities, and the lower limb mass in our patient was highly suspicious of the primary tumour. Moreover, adult RMS is extremely aggressive with 5-year survival rates of approximately 27%, which is lower than survival rates in paediatric populations [12].

For our case this was adult woman and involvement was the orbit with the right maxillary.This

is abit different from the literature as this tumor has predilation in man.Interms of occurance and presentation in adult is the same as literature points out even the histopathology is the same as the literature states.

RMS invading orbit can present in the form of proptosis, lid mass or an orbital mass mimicking orbital cellulitis, lymphangioma, hemangioma, metastasis to orbit, lymphoma, dermoid cyst, or chalazion [13, 14]. In adults, RMS shows increased tendency to invade cranial cavity even after treatment with radiotherapy and chemotherapy [13] and therefore the unfavourable prognosis.

In this case we managed by using the approach of lateral rhinotomy with extended weber fergusson with orbital exenteration.Later on the patient received radiochemotherapy.Followed up the patient and the prognosis was not good.Died at within the six month on treatment .This tallys with the literatures as this tumors has worst prognosis.

CONCLUSION

Patients should have high index of suspicious whenever see any thing not normal to there body should come to the hospital immediatelly regardless is painless or not.

RECOMMENDATION

Public health should offer knowledge to the community to relief this gap of visiting to the hospital late for the patient having tumours.

Abbreviations

RMS-rhabdomyosarcoma
C/S-cesarian section
GA-Gestation age
CD45-Marker of all hematopoietic cells

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

Authors' Contributions

AD:Discussed the case together and assisted in the review of literature
LW:Assisted in the preparation of the manuscript
JK:Involved in the management of the patient
AV-Review of literature
ZS-Review of literature
FM-Management of the patient

Consent: Written informed consent was obtained from the patient for publication of this case report.

Acknowledgements: Department of pathology, Bugando medical centre.

REFERENCES

1. Hettmer, S., Li, Z., Billin, A. N., Barr, F. G., Cornelison, D. D. W., Ehrlich, A. R., ... & Keller, C. (2014). Rhabdomyosarcoma: current challenges and their implications for developing therapies. *Cold Spring Harbor Perspectives in Medicine*, 4(11), a025650.
2. Egas-Bejar, D., & Huh, W. W. (2014). Rhabdomyosarcoma in adolescent and young adult patients: Current perspectives. *Adolesc Health Med Ther*, 5, 115-125.
3. Saboo, S. S., Krajewski, K. M., Zukotynski, K., Howard, S., Jagannathan, J. P., Hornick, J. L., & Ramaiya, N. (2012). Imaging features of primary and secondary adult rhabdomyosarcoma. *American Journal of Roentgenology*, 199(6), W694-W703.
4. Watkinson, J., & Gilbert, R. (2011). *Stell & Maran's textbook of head and neck surgery and oncology*. CRC Press.
5. Macgregor, F., Gleeson, M., & Clarke, R. (2008). 7th ed: *Scott-Brown's Otorhinolaryngology: Head and Neck Surgery*. CRC Press, 1, 1255-1258.
6. Wiss, K., Solomon, A. R., Raimer, S. S., Lobe, T. E., Gourley, W., & Headington, J. T. (1988). Rhabdomyosarcoma presenting as a cutaneous nodule. *Arch Dermatol*, 124, 1687-1690.
7. Loducca, S. V., Mantesso, A., de Oliveira, E. M., & de Araújo, V. C. (2003). Intraosseous rhabdomyosarcoma of the mandible: A case report. *Int J Surg Pathol*, 11, 57-60.
8. Wexler, L., & Helman, L. (1997). Rhabdomyosarcoma and the undifferentiated sarcomas. In: Pizzo, P., Poplack, D., editors. *Principles and Practice of Pediatric Oncology*. 3rd ed. Philadelphia: Lippincott-Raven; pp. 799-829.
9. Crist, W., Gehan, E. A., Ragab, A. H., Dickman, P. S., Donaldson, S. S., Fryer, C., ... & Heyn, R. (1995). The third intergroup rhabdomyosarcoma study. *Journal of Clinical Oncology*, 13(3), 610-630.
10. Ghavimi, F., Exelby, P. R., d'Angio, G. J., Cham, W., Lieberman, P. H., Tan, C., ... & Murphy, M. L. (1975). Multidisciplinary treatment of embryonal rhabdomyosarcoma in children. *Cancer*, 35(3), 677-686.
11. França, C. M., Caran, E. M., Alves, M. T. S., Barreto, A. D., & Lopes, N. N. (2006). Rhabdomyosarcoma of the oral tissues: two new cases and literature review. *Medicina Oral, Patología Oral y Cirugía Bucal (Internet)*, 11(2), 136-140.
12. Gordón Nuñez, M. A., Piva, M. R., Dos Anjos, E. D., & Freitas, R. A. (2008). Orofacial rhabdomyosarcoma: report of a case and review of the literature. *Med Oral Patol Oral Cir Bucal*, 13, 765-769.
13. Turner, J. H., & Richmon, J. D. (2011). Head and neck rhabdomyosarcoma: a critical analysis of population-based incidence and survival data. *Otolaryngology--Head and Neck Surgery*, 145(6), 967-973.
14. Kaliaperumal, S., Tiroumal, S., & Rao, V. A. (2007). Orbital rhabdomyosarcoma: a case series. *Indian journal of cancer*, 44(3), 104.

Cite This Article: Olivia Michael Kimario *et al* (2021). Rhabdomyosarcoma in a Female Adult a Rare Case Report at BMC Mwanza. *East African Scholars J Med Surg*, 3(8), 136-138