Case Report

Indian Journal of Cancer Education and Research
Volume 12 Number 2, July - December 2024
DOI: http://dx.doi.org/10.21088/ijcer.2321.9815.12224.8

Orbital Rhabdomyosarcoma Masquerading as a Sinonasal Carcinoma: Case Report

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How to cite this article:

Neha Singh Rajput, Dipshi Agrawal, Ashar Iqbal Lodi, Virendra Bhandari, Orbital Rhabdomyosarcoma Masquerading as a Sinonasal Carcinoma: Case Report. Ind J Canc Educ Res 2024;12(2):92-95.

Abstract

Background: Rhabdomyosarcoma (RMS), the most common soft tissue sarcoma in children. While most children with localized RMS achieve long-term remission, the prognosis for those with metastatic RMS continues to be challenging, with limited improvements in survival outcomes.

Aim: This report aims to highlight a rare case of orbital rhabdomyosarcoma (RMS) initially misdiagnosed as a sinonasal carcinoma.

Objectives: Initially suspected to be sinonasal carcinoma, histopathological analysis revealed a malignant round cell tumor. Immunohistochemistry (IHC) further confirmed the diagnosis as embryonal rhabdomyosarcoma. This report studies the role of radiation in orbital Rhabdomyosarcoma.

Material: At the SAIMS institute's Radiation Oncology Department, pathology records were examined for case of Rhabdomyosarcoma. The treatment approach for this patient included a combination of radiation therapy and chemotherapy.

Result: We found that throughout the course of radiation therapy patient shows excellent tolerance and patient is on regular follow up in our department.

Conclusion: Orbital RMS has a relatively optimistic prognosis due to its favorable location and histology. In cases where surgery isn't possible, radiotherapy remains essential, offering strong response rates with manageable side effects.

Keywords: Orbital tumors, Embryonal rhabdomyosarcoma, Sinonasal carcinoma.

INTRODUCTION

Orbital rhabdomyosarcoma (RMS), the leading primary soft-tissue sarcoma in children, makes up about 4% of all childhood orbital cancers. This highly aggressive tumor arises from pluripotent embryonic mesoderm, showing a tendency to develop into cells that mimic fetal rhabdomyoblasts. Several distinct types of RMS have been identified: embryonal sarcoma, which originates from early-stage mesenchymal tissue that can develop into

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Received on: 14-11-2024

Accepted on: 10-12-2024

muscle; nonstriated embryonal RMS; and striated embryonal RMS. Diagnostic imaging, including CT and MRI, plays a key role in tumor assessment. Among children, the embryonal subtype is most frequently observed. The primary treatment strategy combines complete surgical removal of the tumor with chemotherapy and radiotherapy for the best outcomes.[1]

CASE REPORT

Patient of a 14-year-old male child presented to us with complaints of proptosis of right eye for the past 6 month and inability to open right eye with severe pain.There was progressive axial proptosis for the past 2 months. Vision was 6/24 in right eye with best pin hole correction of 6/12. Right eye vision was 6/6. There was no history of trauma or history of fever. On local examination, swelling was non-tender, approximately 4 cm × 3 cm in size, firm in consistency, and immobile (*Figure 9.1*).On examination neither pallor nor swollen lymph node observed. Routine blood investigations including complete blood counts, renal function test, and liver function test were within normal limits.

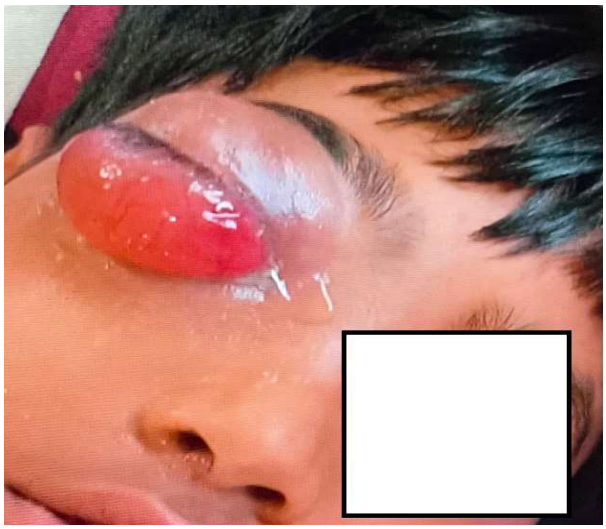

Fig. 9.1: Pretreatment appearance of the tumor

CEMRI shows a lesion of 5.5x5x5.3cm epicentred in right sinonasal region causing destruction of medical wall of right orbit with extension to medial extraconal compartment. It's extending into superior half of right maxillary sinus through through osteomeatal complex. Destruction seen in right lamina papyraea, floor of orbit and medial wall of maxillary sinus. (*Fig. 2*)

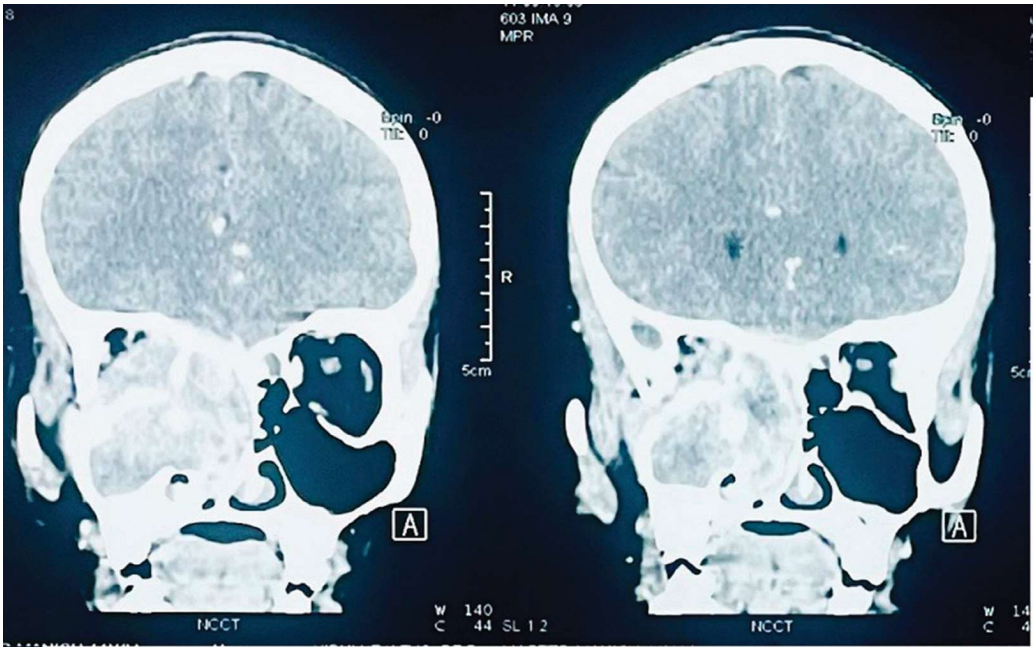

Fig. 2: MRI Brain + Orbit: Solid soft tissue lesion in right sinonasal region with partial destruction of medial wall of right orbit

Biopsy showed malignant round cell tumor and differential diagnosis were olfactory Neuroblastoma, Undifferentiated Carcinomaor Lymphoproliferative Disorder. Immunohis to chemistry was positive for Myogenin, MyoD1, Desmin, CD56 Synaptophysin and negative for CK, LCA, p40, Chromogranin, INSM1 and Calretinin (*Fig. 3*). It favored the diagnosis of embryonal Rhabdomyosarcoma.The patient was subjected tosystemic chemotherapy with VAC and IE alternative as per standard protocol (IRS-IV) followed

byexternal beam radiotherapy by IMRT. There was a good response to the chemotherapy notedwith rapid reduction of bulk of tumor in right eye with no toxicities (*Fig. 4*). MRI Brain and PNS was done for response assessment reveals soft tissue thickening (3-4 m) along floor and medial wall of right orbit in extraconal compartment, adherent to medial and inferior recuts extraocular muscle. As growth was inoperable the patient was planned for radical dose of 54Gy in 30 fractions (1.8Gy per fraction) using 6MV photons on Halcyon Elite. Patient tolerated the treatment well. Now he is on follow up for further evaluation and response to treatment.

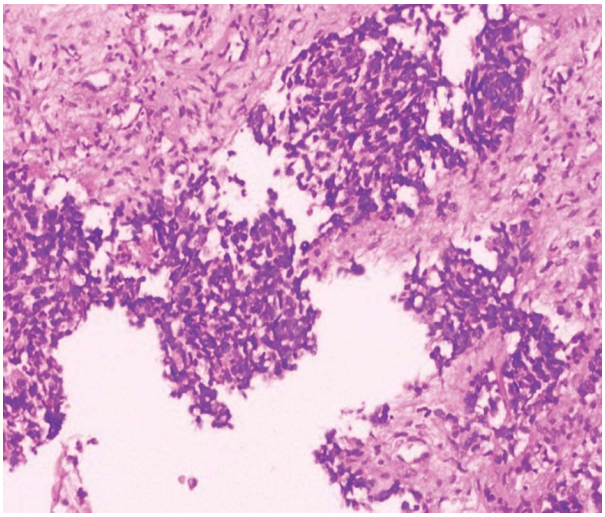

Fig. 3: IHC

Throughout the course of external beam radiotherapy, the patient exhibited excellent tolerance, maintaining normal vision acuity with no dryness. The sole adverse outcome noted was hyper pigmentation (*Fig. 4*).

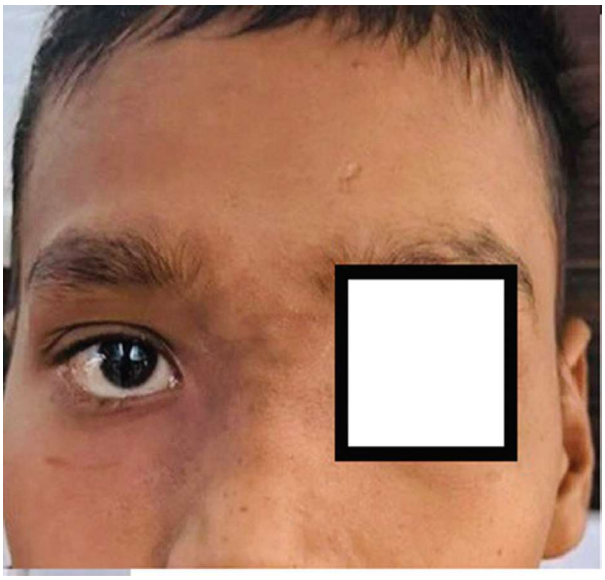

Fig. 4: Post Radiotherapy response of tumor

DISCUSSION

Rhabdomyosarcoma stands as the leading orbital cancer in children, affecting boys more frequently than girls. It often manifests as a fast-growing, one-sided eye protrusion, sometimes accompanied by globe displacement. Diagnosis is confirmed through specific immunohistochemical markers, including desmin, myogenin, MyoD1, MSA, and myoglobin. While desmin and vimentin may appear in various muscle related tumors, they're less specific markers. In contrast, myogenin and MyoD1 key transcription factors in early skeletal muscle development offer greater accuracy, particularly for the embryonal type of RMS. Treatment typically involves surgery, followed by chemotherapy and radiation for optimal results.[2]

Puri DR, Wexler LH, in their study stated that radiation therapy for RMS is generally reserved for cases where surgery is incomplete, lymph nodes are involved, or chemotherapy shows limited effectiveness. International RMS research groups also take a conservative approach, particularly for children under three, to limit the side effects of external beam radiation therapy (EBRT). On average, EBRT is started 18 weeks after the beginning of chemotherapy, with a median dose of 36Gy. For young children, a reduced 36Gy dose after delayed gross total resection (GTR) can balance local control and long-term safety, although unresectable tumors, like those in parameningeal regions, still require higher doses.[3]

Cameron AL, Elze MC in their study stated that radiation therapy was associated with significantly better overall survival (OS) compared to cases without radiation, with the best outcomes seen in patients who received radical rather than partial radiation. The three-year OS rates were 84% for those receiving radical irradiation, 54% for partial irradiation, and only 23% for those who received no radiation. Furthermore, aggressive treatment of the primary tumor whether by surgery, radiation, or a combination markedly improved both event-free survival (EFS) and OS compared to cases without treatment.[4]

Oberlin O, Rey A, Sanchez de Toledo J, and colleagues conducted a study in which patients received an initial course of ifosfamide, vincristine, and dactinomycin (IVA). Following this, patients were randomly assigned to either continue with IVA alone or switch to a more intensive six-drug regimen that alternated IVA with carboplatin, etoposide, and vincristine (CEV) and ifosfamide,

vincristine, and etoposide (IVE). The findings revealed no notable difference in overall survival (OS) or event-free survival (EFS) between the groups. Three-year OS rates were 82% for IVA and 80% for the six-drug combination. However, the six drug regimen resulted in higher toxicity, with patients experiencing more infections, myelosuppression, and mucositis.[5]

CONCLUSION

Rhabdomyosarcoma is the most common primary malignant orbital tumor in children and should be considered when a child presents with rapidly worsening, one-sided eye protrusion. Though highly aggressive and life-threatening, advances in multimodal therapies have significantly improved patient outcomes. Immunohistochemical staining using myogenin antibodies provides precise diagnostic insights, enhancing early detection. Orbital RMS has a relatively optimistic prognosis due to its favorable location and histology. In cases where surgery isn't possible, radiotherapy remains essential, offering strong response rates with manageable side effects.

Conflict of Interest: No conflict of interest

Funding: Nil

Ethics Declaration: No ethical issues involved

REFERENCES

1. Singh, Aditya Pratap; Gupta, Arun Kumar; Mathur, Vinay; Barolia, Dinesh Kumar. Embryonal Rhabdomyosarcoma of the Orbit in a Child. Medical Journal of Dr. D.Y. Patil Vidyapeeth 11(3):p 276-278, May–Jun 2018.
2. Karakosta C, Liaskou M, Kattamis A, Rigatou E, Paraskevopoulos K. Orbital Rhabdomyosarcoma Masquerading as a Dermoid Cyst: A Case Report and Review of the Literature. Cureus. 2023 Dec 11;15(12).
3. Puri DR, Wexler LH, Meyers PA, La Quaglia MP, Healey JH, Wolden SL. The challenging role of radiation therapy for very young children with rhabdomyosarcoma. International Journal of Radiation Oncology* Biology* Physics. 2006 Jul 15;65(4):1177-84.
4. Cameron AL, Elze MC, Casanova M, Geoerger B, Gaze MN, Minard-Colin V,et al. The impact of radiation therapy in children and adolescents with metastatic rhabdomyosarcoma. International Journal of Radiation Oncology* Biology* Physics. 2021 Nov 15;111(4):968-78.
5. Cameron AL, Elze MC, Casanova M, Geoerger B, Gaze MN, Minard-Colin V, et al. The impact of radiation therapy in children and adolescents with metastatic rhabdomyosarcoma. International Journal of Radiation Oncology* Biology* Physics. 2021 Nov 15;111(4):968-78.