International Journal of Research in Medical Sciences
Gupta BK et al. Int J Res Med Sci. 2020 Nov;8(12):4515-4517
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20205335

Case Report

A rare case in an 11-year old child with bilateral primary orbital rhabdomyosarcoma with rapid onset of bilateral proptosis and temporal lobe abscess secondary to chronic suppurative otitis media

Bikram K. Gupta[1]*, Shubham Tomar[2]

[1]Department of Anaesthesiology, [2]MBBS Student, Department of Medicine, Institute of Medical Sciences, Banaras Hindu University, Varanasi, Uttar Pradesh, India

Received: 07 October 2020
Accepted: 10 November 2020

***Correspondence:**
Dr. Bikram K. Gupta,
E-mail: bikramgupta03@gmail.com

ABSTRACT

A rare case in an 11-year old child with bilateral primary orbital rhabdomyosarcoma (RMS) with rapid onset of bilateral proptosis and temporal lobe abscess secondary to chronic suppurative otitis media (CSOM). This article describes the clinical, radiological, histopathological, and immunohistochemical findings. Orbital RMS is always unilateral and mainly presents as proptosis with lid edema and chemosis. The uniqueness of the present case is the simultaneous occurrence of bilateral RMS and temporal lobe abscess.

Keywords: Orbital rhabdomyosarcoma, Embryonal rhabdomyosarcoma, Bilateral proptosis, Temporal lobe abscess, Craniotomy

INTRODUCTION

Soft tissue sarcomas (STS) estimate about seven percent of malignant neoplasm in children.[1] Rhabdomyosarcomas (RMS) constitute fifty percent of all soft tissue sarcomas in children under the age of 15, with an incidence of 4.6 per million per year.[2,3] RMS first described by Weber in 1854, is a rare aggressive, rapidly growing tumor arising from undifferentiated skeletal myeloblast-like cells with head and neck involvement in approximately 40% of cases.[1,4,5]

RMS most commonly involves the retrobulbar region but can involve other areas, including eyelids, conjunctiva, and rarely, uveal tissue, often manifesting as rapidly progressive proptosis or globe displacement.[6,7] We are reporting this case because: rare bilateral orbital RMS with a very aggressive course in an uncommon age group; bilateral proptosis, and simultaneous confusion secondary to temporal lobe abscess in one diagnosis.

CASE REPORT

An 11-year old Hindu male presented at department of ophthalmology in Sir Sunderlal hospital, BHU, Varanasi with rapidly progressive bilateral proptosis with exposure keratitis and loss of vision along with right-sided temporal area swelling and bilateral ear discharge for 15 days (Figure 1). There was a history of loss of appetite and lethargy for two weeks. There was no history of trauma. Past, prenatal and birth history were uneventful. On examination, the general condition was poor, with no lymphadenopathy or hepatosplenomegaly. The chest was clear bilaterally.

Proptosis was non-axial, non-pulsatile with the right eye proptosed more than the left about 40 mm and 38 mm, respectively. The eyelids were edematous; conjunctiva was highly chemosed and congested. The patient had restricted eyeball movement in all directions. Intraocular pressure and fundus examination could not be determined. A soft swelling was present in the right temporal region

with bilateral ear discharge suggestive of bilateral otitis media.

Figure 1: Bilateral proptosis with corneal sloughing, conjunctival chemosis and lid edema.

During the investigation, complete hemogram, general blood picture, erythrocyte sedimentation rate (ESR), coagulation profile, urine, and stool examination was within normal limits. On imaging, x-ray of the orbit showed marked soft tissue density, without any bony involvement. X-ray of the skull, chest, abdomen, and lower extremities were normal. T1 and T2 weighted magnetic resonance imaging (MRI) of the orbit showed an isointense lesion in the right temporal lobe of 2.6×2.5 cm with mild perifocal edema. Sizeable soft tissue extending up to orbital apex is seen in the right superior and left superomedial extraconal spaces. Soft tissues were noted in the lateral wall of bilateral orbits, in the posterior fossa and bilateral petrous temporal bone, and sub-periosteal deposits along with the outer table of bilateral mastoids. MRI brain showed bilateral otitis media with a cerebral abscess in the right temporal lobe with sub-periosteal and intra-orbital collection (Figure 2).

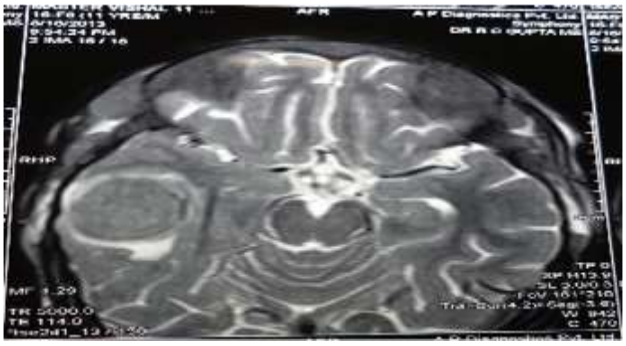

Figure 2: MRI showing right temporal lobe abscess and bilateral extra conal soft tissue mass displacing the eyeballs.

The pre-anesthetic evaluation revealed a mallampatti class II patient in altered sensorium with a pulse rate of 90 beats/min, blood pressure of 118/70 mmHg, mild grade fever, and normal labs. No other systemic complaint was present. A provisional diagnosis of temporal lobe abscess with intracranial and extraconal extension leading to proptosis was made. The decision was taken to drain the abscess and operate under general anesthesia.

A right-sided orbitozygomatic craniotomy was performed for decompression of space-occupying lesion and temporal lobe abscess drainage. The left-sided tumor was untouched. The patient also received bilateral tarsorrhaphy. During a craniotomy, the patient received injection mannitol 1 mg/kg and injection dexamethasone 0.5 mg/kg. The patient was extubated successfully with an uneventful intraoperative and postoperative course.

Based on histopathological examination of the tumor, sheets of monomorphic round to ovoid cells have small round eccentric nuclei and scanty eosinophilic cytoplasm with mitotic figures. The tumor involved adjacent adipose tissue and muscles with perivascular cellular arrangements. Myo D1 was positive on immunohistochemical analysis. These features favored embryonal RMS.

The patient received chemotherapy with vincristine, dactinomycin, with cyclophosphamide followed by adjuvant radiation therapy. At the 3-month follow-up, the patient was recovering well.

DISCUSSION

Retinoblastoma is the most common orbital tumor in children, followed by capillary hemangioma.[5,8] The most common causes of proptosis in children are orbital Rhabdomyosarcoma and retinoblastoma with an orbital spread.[9] RMS incidence is highest among children aged 1-4 years, further decreasing at age 10-14 years and lowest among age 15-19 years.[10] Pediatric RMS is mostly sporadic, but parental use of cocaine or marijuana and radiation therapy are some risk factors. Pediatric RMS is also associated with Li-Fraumeni syndrome, neurofibromatosis, Costello syndrome, Noonan syndrome, and Beckwith-Wiedemann syndrome.[11] RMS patients tend to delay consultation and generally present with advanced disease because of the tumor's rapid growth.[1]

Biopsy or fine needle aspiration cytology (FNAC) of RMS is problematic because these are vascular tumors, which can be associated with severe chemosis and lid edema.[7] Histologically, various RMS types are present, among which embryonal (ERMS) and alveolar (ARMS) subtypes are most common in the pediatric age group. Spindle cell, botryoid, and pleomorphic are amongst other minor variants.[12] The most differentiated pleomorphic RMS carries the best prognosis, followed by embryonal and botryoides RMS. Alveolar RMS has the worst prognosis and a greater frequency of disseminated metastases.[7] RMS appears on histology as small round blue cells with expression of desmin and MyoD muscle-specific antigens and eosinophilic rhabdomyoblasts on standard pathological staining.[3] Yes-associated protein (YAP) is associated with sarcoma genesis in RMS patients and is

more prevalent in ERMS than ARMS, thus differentiating the two.[13]

The differential diagnosis of orbital RMS includes proptosis secondary to inflammatory, infectious, vascular, and neoplastic conditions, and thus careful histological examination is required to differentiate such lesions.[14] In our case, the marked pleomorphism noted was critical for differentiating RMS from Ewing's sarcoma. The presence of an embryonal pattern, pleomorphism, cohesive nature of the cells, and the absence of lymphadenopathy ruled out lymphoma diagnosis. In this respect, the most critical differential diagnosis is neuroblastoma. A diffuse pattern of small round cells and the presence of rosettes/pseudorosettes with pale eosinophilic material seen in cases of neuroblastoma may suggest alveolar RMS. A high level of urinary catecholamines observed in neuroblastoma is critical to rule out this tumor. Following diagnosis, systemic evaluation is necessary for metastasis. The majority of RMS are not encapsulated and can involve the surrounding structures or can extend intracranially.[7]

Survival of patients with RMS generally depends on several factors, including histopathologic and cytological tumor type, the extent of disease at diagnosis, tumor burden at diagnosis, primary tumor site, patient age, cellular ploidy, and therapeutic response. The favorable prognostic characteristics for disease-specific survival are age ≤20 years, tumor size ≤5 cm, absence of regional or distant disease, surgical resection with negative margins, and pleomorphic type.[11]

The overall five-year survival rate was about seventy percent in all pediatric age groups with combined chemotherapy, radiation therapy, and surgery. In pediatric RMS, the need for surgery depends on the anatomical site of the tumor. Administered cyclophosphamide dose has both acute and long-term side effects and is an essential factor in the treatment plan. Various clinical trials were conducted with a reduced dose of cyclophosphamide and newer drugs like irinotecan to prevent a negative effect on treatment outcomes.[15]

CONCLUSION

As per the author's knowledge, this is a rare case of biopsy-proven bilateral RMS. RMS's precise diagnosis and a thorough pre-operative evaluation can primarily minimize the difficulties and the various potential challenges associated with managing such cases.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Koscielniak E, Morgan M, Treuner J. Soft Tissue Sarcoma in Children. Pediatr Drugs. 2002;4(1):21-8.
2. Young J, Smith M, Roffers S, Liff J. Institute GB-NC, SEER undefined. Cancer incidence and survival among children and adolescents: United States SEER Program. 1975-1995.
3. Loeb DM, Thornton K, Shokek O. Pediatric Soft Tissue Sarcomas. Surgical Clinics of North America. NIH Public Access. 2008;88:615-27.
4. Skapek SX, Ferrari A, Gupta AA, Lupo PJ, Butler E, Shipley J. Rhabdomyosarcoma. Nature Reviews Disease Primers. Nature Publishing Group. 2019;5:1-19.
5. Hicks J, Flaitz C. Rhabdomyosarcoma of the head and neck in children. In: Oral Oncology. Pergamon. 2002;450-9.
6. Tang LY, Zhang MX, Lu DH, Chen YX, Liu ZG, Wu SG. The prognosis and effects of local treatment strategies for orbital embryonal Rhabdomyosarcoma: a population-based study. Cancer Manag Res. 2018;10:1727-34.
7. Kaliaperumal S, Tiroumal S, Rao V. Orbital rhabdomyosarcoma: A case series. Indian J Cancer.2007;44(3):104.
8. Castillo BV, Kaufman L. Pediatric tumors of the eye and orbit. Pediatric Clinics of North America. W. B. Saunders. 2003;50:149-72.
9. Bajaj MS, Pushker N, Chaturvedi A, Betharia SM, Kashyap S, Balasubramanya R, et al. Orbital space-occupying lesions in Indian children. J Pediatr Ophthalmol Strabismus. 2007;44(2):106-11.
10. McDowell HP. Update on childhood rhabdomyosarcoma. Arch Dis Child. 2003;88(4):354-7.
11. Yee NWK. Embryonal Rhabdomyosarcoma in a young boy. Mcgill J Med. 2007;10(1):16-9.
12. Pizzo P, Poplack D, Adamson P, Blaney S. Principles and Practice of Pediatric Oncology. Wolters Kluwer: Philadelphia, PA. 2015.
13. Ahmed AA, Habeebu SS, Sherman AK, Ye SQ, Wood N, Chastain KM, et al. Potential Value of YAP Staining in Rhabdomyosarcoma. J Histochem Cytochem. 2018;66(8):577-84.
14. Bilateral Intraocular Rhabdomyosarcoma: A Case Report. Available at: https://www.researchgate.net/publication/301608488_Bilateral_Intraocular_Rhabdomyosarcoma_A_Case_Report. Accessed on: 24 September 2020.
15. Miyachi M, Tsuchiya K, Hosono A, Ogawa A, Koh K, Kikuta A, et al. Phase II study of vincristine, actinomycin-D, cyclophosphamide and irinotecan for patients with newly diagnosed low-risk subset B rhabdomyosarcoma: A study protocol. Med (United States). 2019;98(52).

Cite this article as: Gupta BK, Tomar S. A rare case in an 11-year old child with bilateral primary orbital rhabdomyosarcoma with rapid onset of bilateral proptosis and temporal lobe abscess secondary to chronic suppurative otitis media. Int J Res Med Sci 2020;8:4515-7.