International Journal of Current Advanced Research
ISSN: O: 2319-6475, ISSN: P: 2319-6505, Impact Factor: 6.614
Available Online at www.journalijcar.org
Volume 7; Issue 9(C); September 2018; Page No. 15405-15407
DOI: http://dx.doi.org/10.24327/ijcar.2018.15407.2810

Case Report

RHABDOMYOSARCOMA OF ORBIT IN A YOUNG CHILD: A CASE REPORT

Shilpi Gangwar., Roop Ganguly., Shaheefa Begum and HariRam

Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences,
King George's Medical University, Lucknow. U.P

A R T I C L E I N F O

Article History:

Received 13th June, 2018
Received in revised form 11th July, 2018
Accepted 8th August, 2018
Published online 28th September, 2018

Key words:

Chemotherapy and radiotherapy, rhabdomyosarcoma

A B S T R A C T

A four and a half year old child had rapidly progressive proptosis of the left eye with associated chemosis over a period of several weeks. Computed tomography demonstrated a solid extraconal mass in the inferior anterior left orbit. Histopathologic examination revealed the lesion to be an embryonal rhabdomyosarcoma. Consistent with the diagnosis, immunohistochemical assays demonstrated positive staining with myoglobin, desmin, and vimentin. The lesion grew rapidly. Subsequently, treatment chemotherapy was initiated.

INTRODUCTION

Rhabdomyosarcoma (RMS) is the most common primary malignancy of the orbit in children, accounting for 4% of all malignant disease in children[1,2]. Horn and Enterline in mid 1900 first classified RMS histologically into 4 major categories: embryonal, alveolar, botryoid and pleomorphic [2]. Prognostically pleomorphic variety has best prognosis[2]. The average age of presentation is 4-7 years[3]. Most of the time the tumor is retrobulbar, but it may arise from any part of the orbit, even from the conjunctiva and anterior uveal tract[4]. The majority of tumors are localized to the orbit. In its highly malignant form, RMS grows rapidly and behaves aggressively, frequently invading adjacent bones and soft tissues. Extension of tumor outside the orbit is associated with worse prognosis particularly if there is skull base erosion. Though earlier orbital RMS was treated by exenteration, in 1979, Abraham *et al*. demonstrated irradiation alone or in combination with chemotherapy to be more effective than exenteration for both control and long term survival. Marked improvement in RMS survival over the past seen with the use of radiotherapy and chemotherapy [5].

Case report

A patient of 4 and a half years reported to the department of oral & maxillofacial surgery with the chief complain of swelling around his left eye since 4 days. As the case was not related to any odontogenic infection,tumor or trauma,so the case was referred to deptt. of ophthalmology.Where he was prescribed with some eye drops.But patient was not relieved with the eye drops and swelling gradually increased in size over a period of 15 days(fig.1).

Figure 1 Clinical photograph before chemotherapy

Patient again visited to the department of ophthalmology where he was prescribed MRI & CT Scan.

CT findings revealed well defined heterogeneously enhancing soft tissue attenuation lesion with non enhancing hypodense areas of necrosis is seen in left orbit. Anteriorly the lesion is seen having ill defined interface with posterior wall of the eye ball however no intraocular extension is seen. The lesion is causing anteroinferiorly displacement of left eyeball resulting in to extraaxial proptosis .The lesion is displacing and encasing of optic nerve up to 360 degrees. The lesion is abutting posterior part of inferior rectus muscle and causing its heterogeneously enhancing thickening. Rest extraoccular muscles and lacrimal gland are not visualized separately from

Corresponding author:* **Shilpi Gangwar
Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, King George's Medical University, Lucknow. U.P

the lesion involved. Lesion is seen abutting superior, medial and lateral walls of orbit however no osseous erosion is seen(fig.2&3)

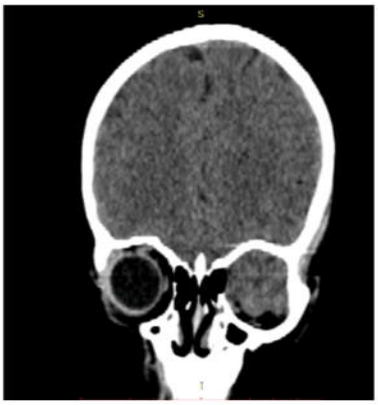

Figure 2 CT Scan – coronal view before chemotherapy

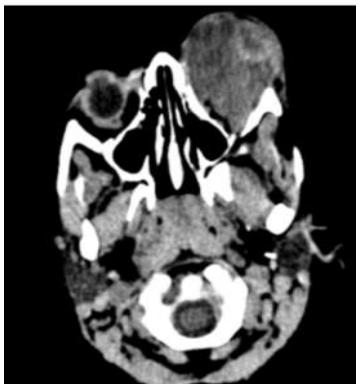

Figure 3 CT Scan – axial view before chemotherapy

MRI revealed a moderate size lobulated well defined inhomogeneous intraconal mass seen in the superomedial aspect of the left orbit. Left orbit is displaced antero-inferiorly with the resultant proptosis of the eyeball. Mass is seen compressing and displacing the superior rectus – LPS complex muscle. Optic nerve is displaced inferomedially. Posteriorly lesion is seen extending up to the orbital apex. However, no obvious optic nerve canal or intracranial extension seen (fig.4,5&6).

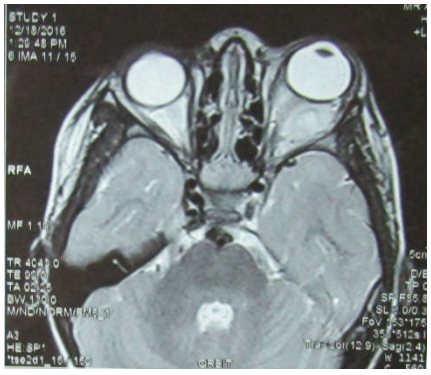

Figure 4 MRI Scan – coronal view before chemotherapy

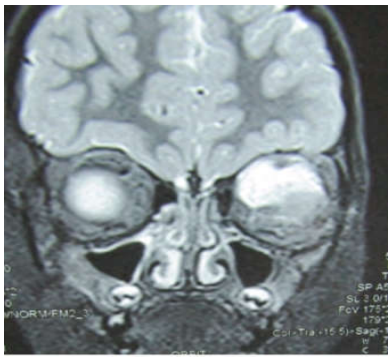

Figure 5 MRI Scan – axial view before chemotherapy

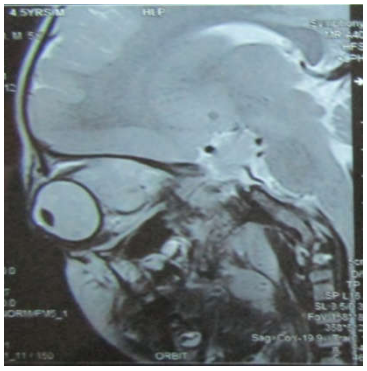

Figure 6 MRI Scan – sagittal view before chemotherapy

MRI and CT suspected the diagnosis of Rhabdomyosarcoma. Regional lymph nodes were not palpable and there was no distant metastasis observed on detailed work-up which include complete blood examination, liver function test, chest X-ray, ultrasonography of whole abdomen and bone marrow aspiration. Patient was then admitted there and the biopsy was done.

Section from biopsy tissue shows a malignant mesenchymal neoplasm disposed as diffuse sheets as well as loose fascicles at places along with dilated blood vessels in the substance of tumor. Individual tumor cells are pleomorphic, round to oval to spindle shaped have coarse granular chromatin and scant amount of cytoplasm. The histopathological diagnosis was confirmed by immuno-histochemical study.

Immunohistochemistry from incisional biopsy confirmed the diagnosis of Embryonal rhabdomyosarcoma (Vimentin – positive in tumor cells,Desmin – positive in tumor cells,Myogenin – positive in tumor cells,S100 – Negative in tumor cells,MPO – Negative in tumor cells) (fig.7,8,9 respectively).

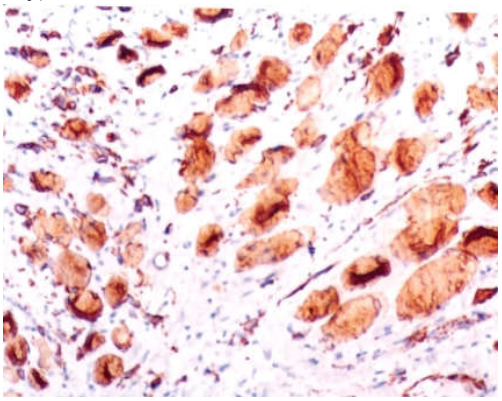

Figure 7 Histopathologic image-Tumor cells showing positive staining with vimentin

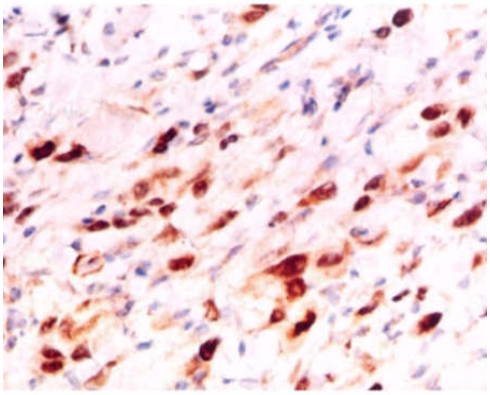

Figure 8 Histopathologic image- Tumor cells showing positive staining with desmin

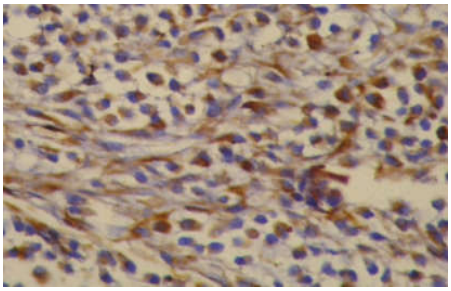

Figure 9 Histopathologic image -Tumor cells showing positive staining with myogenin

Patient was then referred to department of paediatric oncology where he was given 4 cycles of chemotherapy (VAC regimen) after 3 weeks duration . Patient is in follow up since 4 months and his swelling reduced in size up to normal but patient has no vision in his eyes after the course of chemotherapy (fig.10).

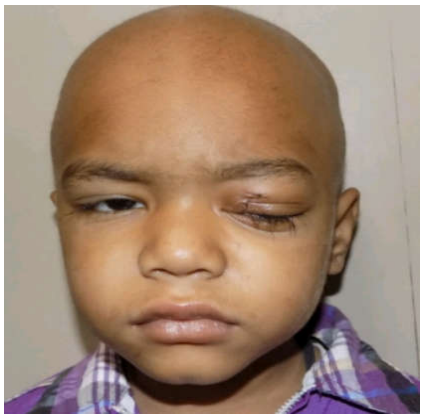

Figure 10 Clinical photograph after chemotherapy

DISCUSSION

The orbital rhabdomyosarcoma mostly presents as painless expansile lesion which may mimic various benign and malignant pathologies occurring in orbit. Metastasis in orbital rhabdomyosarcoma is mainly hematogenous and may involve distant bones and organs with rare regional lymph node metastasis[3,5]. Recurrence in rhabdomyosarcoma usually appears with in three years of treatment [6] but in our case no recurrence was observed uptill four months of follow up. Intergroup Rhabdomyosarcoma Study Group (IRSG) whose protocols are followed now a days for treatment of rhabdomyosarcoma.[7]

As per IRSG staging our case was instage III. Recently excision biopsy is preferred more than incisional biopsy as it confirms diagnosis as well as it removes the tumor too, which is significantly helpful for better outcome.[7] Exenteration is usually done for recurrence cases or the lesions which are resistant to chemotherapy and radiotherapy[8].

References

1. Bajaj MS, Pushker N, Chaturvedi A, Betharia SM, Kashyap S, Balasubramanya R, *et al*. Orbital space occupying lesion in Indian children. *J Pediatr Ophthalmol Strabismus* 2007;44:106-11.
2. Volpe NJ, Jakobiec FA. Pediatric Orbital tumors. *Int Ophthalmol Clin* 1992;32:201-21.
3. Shields JA, Shields CL. Rhabdomyosarcoma: review for the ophthalmologist. *Surv Ophthalmol* 2003;48:39-57.
4. Shields CL, Shields JA, Honavar SG, Demirci H. Clinical spectrum of primary ophthalmic rhabdomyosarcoma. *Ophthalmology* 2001;108:2284-92.
5. Shields JA, Shields CL, Scartozzi R. Survey of 1264 patients with orbital tumors and simulating lesions. *Ophthalmology* 2004;111:997-1008.
6. Mannor GE, Rose GE, Plowman PN, Kingston J, Wright JE, Vardy SJ. Multidisciplinary management of orbital rhabdomyosarcoma. *Ophthalmology* 1997;104:1198-201.
7. Pappo AS, Shapiro DN, Crist WM, Maurer HM. Biology and therapy of pediatric rhabdomyosarcoma. *J Clin Oncol* 1995;13:2123-39.
8. Omezzine N, Khouatra C, Larivé S, Freyer G, Isaac-Pinet S, Gérinière L, *et al*. Rhabdomyosarcoma arising in mediastinal teratoma in an adult man: a case report. *Ann Oncol* 2002;3:323-6.

How to cite this article:

Shilpi Gangwar., Roop Ganguly., Shaheefa Begum and HariRam (2018) 'Rhabdomyosarcoma of Orbit in a Young Child: A Case Report', *International Journal of Current Advanced Research,* 07(9), pp. 15405-15407.
DOI: http://dx.doi.org/10.24327/ijcar.2018.15407.2810

****