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ORBITAL RHABDOMYOSARCOMA -A CASE REPORT

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ABSTRACT

Rhabdomyosarcoma is the most common soft tissue mesenchymal tumour in children. It accounts for 3.4% of all childhood malignancies. Orbital Rhabdomyosarcoma is one the few life threatening disease which, with prompt diagnosis and treatment can be life saving. It can involve the orbit, eye lid, conjunctiva and uveal tract. Following treatment the recurrence rate is reported to be 18 %, metastasis in 6% and death in 3%. A nine year old boy presented with painless progressive eccentric proptosis in Right eye of one month duration. On examination a non tender mass was noted in the inferotemporal region of right orbit, causing upward and inward displacement of the eye ball, suggestive of orbital Rhabdomyosarcoma. Inferior orbitotomy was done and mass was excised in total. Histopathological study confirmed the presence of Rhabdomyosarcoma. Most common site being supero-nasal quadrant, here we report this case due to its atypical site of presentation and under reporting in literature.

Keywords: Orbital Rhabdomyosarcoma, Eccentric proptosis, Infero-temporal quadrant

INTRODUCTION

Rhabdomyosarcoma arises from pleuripotent mesenchymal precursors that normally differentiate into striated muscles [1]. Orbital rhabdomyosarcoma accounts for about 25-35 % of head and neck Rhabdomyosarcoma and about 10-20% of all Rhabdomyosarcoma [2]. Weber was credited with providing the first acceptable description of Rhabdomyosarcoma, which was noted in tongue of a 21 year old man [3]. In 1882, Bayer was first to publish the description of Rhabdomyosarcoma [4]. It is primarily a disease of young children with 90% occurring before 16 years and mean age of onset of 5-7 years [5].Boys are more commonly affected than girls, at a ratio of 5:3 [2]. In the orbit most common histological variant is the embryonal type, followed by the alveolar type [1]. The most frequent clinical findings in patients with ocular Rhabdomyosarcoma is proptosis, globe displacement, lid oedema, and conjunctival congestion. Thus, the differential diagnosis includes inflammatory lesions such as orbital cellulitis, idiopathic inflammatory orbital pseudo tumour, conjunctivitis, allergic oedema, other tumours such as orbital capillary haemangioma, lymphangioma, and Langerhan's cell histiocyosis [6, 7]. The approach to treatment has undergone radical change over the last 20 years from primary exenteration to a more conservative multidisciplinary approach of excision of mass combined with systemic chemotherapy and local radiation therapy. The

children who survive need to be followed up for many years for development of late effects of local treatment [8]. Several investigators have focused on late effects of therapy in children with orbital Rhabdomyosarcoma which include cataract, decreased visual acuity, orbital hypoplasia, dry eye, chronic keratoconjunctivitis, and retinopathy [9].

Case report:

A 9 year old boy presented to of BLDEU's Department of Ophthalmology outpatient department with complaints of rapidly progressive, painless proptosis of right eye of one month and blurring of vision in right eye of 15 days duration. No complaints in left eye. No history of fever, sinusitis, epistaxis or trauma. On ocular examination right eye was displaced upwards and inwards, lids were swollen, conjunctiva chemosed, ocular movements were grossly restricted in all positions of gaze with fundus examination showing blurring of optic disc margins and venous dilatation. Visual acuity was 6/60 with pin hole improvement of 6/36. Colour vision was normal. On palpation a non tender mass of 2X3 cms, irregular surface, and firm in consistency was noted in the inferior quadrant of orbit. It was non pulsatile or non expansile. Left eye was normal. Pallor was noted in general examination and the systemic examination of all the organs were within normal limits. Blood, urine and stool examination was normal. Ultrasonography of orbit suggested 3.5x2.7x1.8 cms of heterogenous lesion, with solid and cystic areas with increased vascularity at the margin. Lesion appeared to be extra as well as intra orbital in location, seen infero lateral to right eye ball, insinuating between bony orbit and right eye ball. Fine needle aspiration cytology showed moderate cellular material with round ductal cells having round nuclei with pink cytoplasm. Background showed abundant mucoid material, RBC's with pink material with no atypia, suggestive of benign mucoid mass. Inferior orbitotomy was done and a 4x3 mass was found communicating with major vessels, which were ligated and mass, was separated from its posterior and inferior attachments and cut in toto. Mass was sent for histopathological examination which was reported as suggestive of embryonal Rhabdomyosarcoma of orbit.

DISCUSSION

Rhabdomyosarcoma, the most common primary orbital malignancy of childhood. The tumour can arise primarily in the orbit or it can arise in the sinuses or nasal cavity and secondarily extend to involve the orbit.

There was once a misconception that orbital Rhabdomyosarcoma arise from extraocular muscles, similar to pleomorphic Rhabdomyosarcoma of adulthood. In 1962, Porterfield and Zimmerman refuted the prevailing assumption that embryonal Rhabdomyosarcoma originates from preformed muscle [10]. Conceivably, Rhabdomyosarcoma could arise in any part of the body where such primitive pleuripotent cells exist, even if that area did not contain any skeletal muscle.

Orbital Rhabdomyosarcoma has occurred as a second tumour in patients who had ocular irradiation therapy for retinoblastoma [11] and after irradiation for squamous cell carcinoma of the eyelid [12] .There appears to be no recognizable environmental, viral, or chemical influence in the pathogenesis of Rhabdomyosarcoma.

The tumour has been seen in patients with certain familial syndromes like neurofibromatosis and Li-Fraumeni syndrome [13]. The latter has been associated with germ line mutation of the p53 tumour suppressor gene [14].

Embryonal Rhabdomyosarcoma is by far the most common variant found in the head and neck region, including the orbit. The alveolar

and botryoid types are less common, and the pleomorphic type is extremely rare in the orbit.

Embryonal Rhabdomyosarcoma is characterized histopathologically by spindle shaped to round cells that show features of skeletal muscle in various stages of embryogenesis. The alveolar type appears as loosely arranged, malignant cells with septae that reminiscent of the alveoli of the lung. The botryoid type may be a variant of the embryonal type that assumes a papillary configuration. Today, immunohistochemical studies have become the main approach to establishing the diagnosis. The most useful immunohistochemical marker includes antibodies against desmin, muscle-specific actin, and myoglobin, which typically show positive reaction in Rhabdomyosarcoma. Although vimentin shows a positive reaction in most cases, it is less specific and can be positive in a variety of other neoplasms.

Current management of orbital Rhabdomyosarcoma depends on the location and extent of disease and generally consists of combination chemotherapy and radiotherapy delivered to the orbit and all involved sites of tumour. Recurrent tumours in the orbit usually are treated with orbital exenteration, sometimes supplemented with chemotherapy and radiotherapy.

In each case of suspected Rhabdomyosarcoma, the surgeon should carefully review the axial and coronal CTs and MRIs to determine whether it is possible to attempt complete removal of the mass. If the tumour is very anterior in the orbit and presenting in the conjunctival fornix, a forniceal approach can be considered. However, most tumours are located more posteriorly or are palpable through the eyelid. In such instances, a cutaneous approach is preferred. Patients with alveolar cell type show 74 % 5-year survival whereas those with embryonal cell type demonstrated 94 % 5-year survival.

CONCLUSION

In conclusion, Orbital Rhabdomyosarcoma being the most common orbital malignancy of childhood, ophthalmologists should keep in mind the probability of atypical site of presentation i.e. infero- temporal as in our case as against the usual supero-temporal quadrant and plan a suitable line of management after histological confirmation.

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