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An uncommon location of rhabdomyosarcoma in a 2-year-old patient

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Abstract: Rhabdomyosarcoma is a malignant tumor: It is the commonest soft-tissue sarcoma of childhood. It can arise from several sites in the body including the ocular region. The prognosis depend on the earliness of the diagnosis as well as the management. The case presented here was located in the lower eyeli which is an unusual area of this tumor.
Keywords: Unusual location, rhabdomyosarcoma

INTRODUCTION

Rhabdomyosarcoma (RMS) is the commonest soft-tissue sarcoma of childhood. Primary Orbital RMS is mainly a disease of young children, where 90% of cases present before the age of 16 years old. The mean age of onset is 5-7 years old [1]. It is considered to be the commonest malignant tumor of soft tissue in children under 15 years of age, accounting for 5% to 10% of all childhood malignancies [2]. It can occur in several sites in the body including the ocular region. Orbital rhabdomyosarcoma accounts for about 25-35 % of head and neck rhabdomyosarcoma [3]. Classic clinical picture is sudden onset and rapid evolution of proptosis without history of previous trauma or sign of upper respiratory tract infections [4]. Ocular RMS comprises tumors that occur in the orbit, or rarely in other ocular adnexal structures or within the eye [5].

CASE REPORT

A 2-year-old female child was reffered to pediatric ophthalmology service due to right lower lid swelling since three months before visit. There was no history of trauma or family malignancy. On external examination, the rigth lower lid had a huge swelling. On palpation, the swelling was firm and nonmobile. The eyeball was almost hidden by the swelling. The left eye was normal. Orbital computed tomography (CT) scan with axial and coronal view was requested and revealed an homogenous well-defined mass. A biopsy was performed under general anesthesia and the histopathologic exam confirm the diagnosis of embryonal rhabdomyosarcoma. The patient was subjected to chemotherapy prior to total excision.

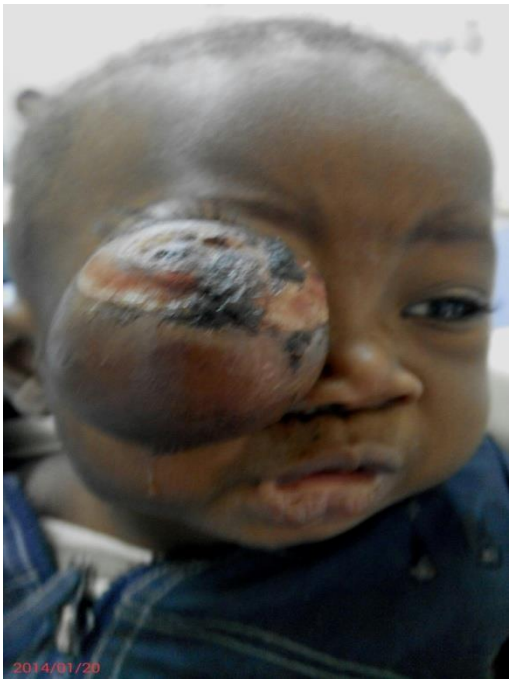

Fig.1: Photograph of the child with lower eyelid rhabdomyosarcoma

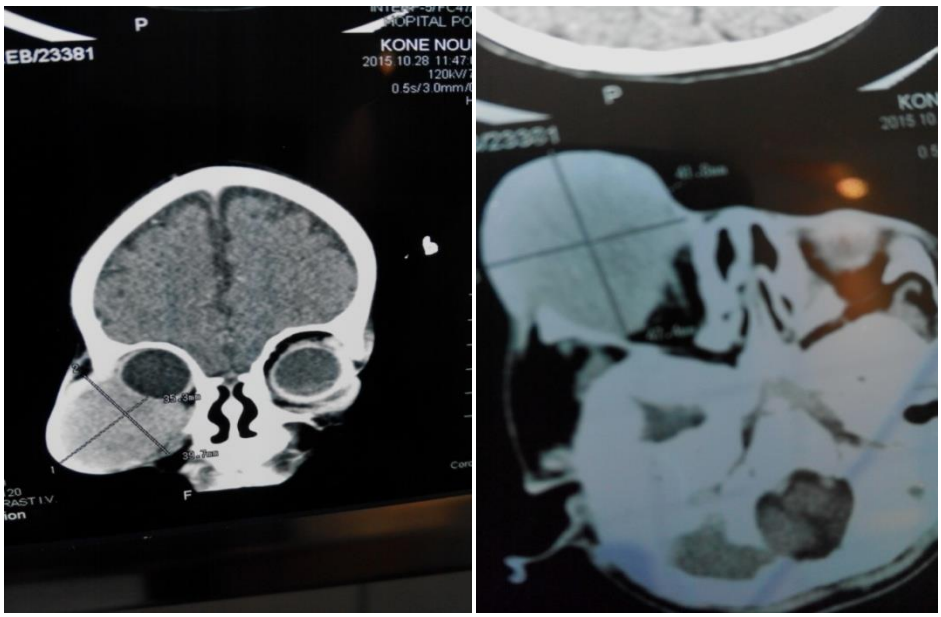

Fig.2: CT Scan showing the right eye rhabdomyosarcoma

DISCUSSION

The typical presentation for primary orbital RMS is the rapid onset of unilateral proptosis and inferior or inferiotemporal displacement of the globe. Otherwise, patients may have a history of worsening eyelid edema and erythema, chemosis, ophthalmoplegia, blepharoptosis, or a palpable mass [1]. In the current case, the onset was very fast with a rapid increase of the mass looking like a giant cyst of the lower lid; only the histopathology could confirm the diagnosis.

Although RMS was once believed to arise from extraocular muscles, it is now accepted that orbital it develops from undifferentiated mesenchymal cells that have the capacity to differentiate into striated muscle.The histopathologic types of RMS include embryonal, alveolar, and pleomorphic. The embryonal form is the commonest; the alveolar variety is less common and carries the worst prognosis [6]. The case presented here is rare because this site is not the classic location of RMS. A review of clinical spectrum of 33 pediatric patients by shields and al. [5] showed that the orbit is the most commonly affected area (76%) followed by the conjunctiva (12%), uveal tract (9%), within the globe (9%) and eyelid (3%). [7, 8]. Early diagnosis, complete surgical resection followed by a combination of chemotherapy and irradiation offer approximately 70 to 90% 3-year survival.

CONCLUSION

Rhabdomyosarcoma is a highly malignant tumor; pure eyelid localization is rare and can be confused with other eyelid swellings. Only biopsy with histopathologic exam can confirm the diagnosis

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