Abstract: Rhabdomyosarcoma (RMS) is predominantly a childhood malignancy, seen rarely in adults. This is a case report of alveolar RMS involving the nasal cavity with intraorbital extension. A 28 year old gentleman presented with proptosis of the left eye and nasal block for 1 month. Vision was 6/6 J1 in both eyes. There was proptosis of 5mm in the left eye with restricted adduction and elevation and choroidal folds suggestive of external compression of the globe. MR CT imaging showed a well-defined polypoidal soft tissue mass in the left nasal cavity extending supero-medially into the left orbit. A diagnostic biopsy was done endoscopically which was reported as alveolar rhabdomyosarcoma. After undergoing three cycles of chemotherapy, there was significant reduction in size of the nasal mass. The orbital part of the mass completely disappeared after chemotherapy on repeat MR CT imaging. Adult RMS is a rare malignant tumor that calls for expert management. It is of paramount importance to formulate a treatment plan on an individual basis after considering the patients age, the stage of disease and the tumor size. Although it is found rarely in an adult, RMS should figure in the differential diagnosis of orbital tumors irrespective of age. Chemotherapy itself can bring down the size of the tumor drastically as in this case. Complete surgical removal of the tumor can be ensured in large tumors if surgery is performed after chemoreduction.

Keyword: adult alveolar rhabdomyosarcoma proptosis orbital mass chemotherapy

Introduction
Rhabdomyosarcoma (RMS) is predominantly a childhood malignancy, seen rarely in adults. It is characterized histologically by striated muscle fibres in varying stages of embryogenesis. With newer chemotherapy and radiotherapy, the mortality due to this malignancy has substantially dropped. (1,2) Four types of RMS have been described microscopically: embryonal, alveolar, botryoid and pleomorphic. (3) We report a case of alveolar RMS involving the nasal cavity with intraorbital extension.

Case report
A 28 year old gentleman presented to us with outward protrusion of the left eye and nasal block for 1 month. The protrusion gradually increased over time. However, there was no decrease in vision, pain or history of trauma. On examination, vision was 6/6 J1 in both eyes. There was proptosis of the left eye. Ocular movements were full in the right eye; adduction and elevation were restricted in the left eye as shown in Figure 1. Anterior chamber was quiet and lenses were clear in both eyes. There was no relative afferent pupillary defect. IOP was 12 mm of Hg in both eyes. Right eye fundus examination was unremarkable. Left eye showed choroidal folds suggestive of external compression of the globe. On Hertel’s exophthalmometry, there was a 5mm proptosis of the left eye with respect to the right eye. Colour vision was trichromatic in both eyes. Automated perimetry was normal in both eyes. MR CT imaging showed a well-defined polypoidal soft tissue mass in the left nasal cavity measuring 36x45x46 mm in the widest dimension. It was hyperintense on T2 and FLAIR images, hypointense on T1 images and showed heterogenous contrast enhancement. The mass was seen to extend supero-medially into the left orbit, destroying the medial wall of the orbit and displacing the ocular muscles. Both intra and extracranal extension of the mass was noted. The optic nerve was seen separate from the mass (Figure 2). Systemic evaluation did not show any metastases.
Lymph nodes, bone and lungs. The alveolar subtype is considered to be the most notorious for lymphatic and hematogenous spread. The most common route of spread is through the blood to the cervical group of lymph nodes. Alveolar RMS is the second most common subtype of the tumor. Embryonal RMS is the most common subtype found in the head and neck. Alveolar RMS is rarely found in the orbit. (3) However, alveolar RMS originating in the sinuses with orbital extension have been reported in adults. (5,6) RMS tumors generally display high chances of distant metastases. Head and neck RMS on CT imaging usually show inhomogeneous, poorly defined masses with destruction of adjacent bones. MRI picture is that of an isointense or minimally hypointense mass relative to muscle on the T1-weighted images and hyperintense appearance in relation to fat and muscle on the T2-weighted images. (7) The survival rate of patients with RMS depends on several prognostic factors, including extent of disease at the time of diagnosis, cytologic and histopathologic tumor type, tumor burden at diagnosis, site of primary tumor, age of the patient, cellular ploidy, and response to treatment. (8) The good prognostic factors are age less than or equal to 20 years, size of the tumor 5 cm, absence of any regional or distant disease, negative margins reported after surgical resection and pleomorphic subtype of RMS. (9) Treatment comprises a combination of chemotherapy, surgery and radiotherapy depending on various factors like primary tumor site, clinical stage and findings of the tumor on pathology. (10) Complete surgical removal of the tumor can be ensured in large tumors if surgery is performed after chemoreduction. Complete surgical removal of the tumor can be ensured in large tumors if surgery is performed after chemoreduction.


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