SALVAGING VISION IN AN EXTENSIVE PARAMENINGEAL RHABDOMYOSARCOMA : A CASE REPORT.

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Abstract: This communication presents a case report of a parameningeal rhabdomyosarcoma in a young male leading to vision loss which was effectively salvaged using prompt intervention in the form of surgery and high dose systemic steroids. Early debulking was the key in the prevention of complete blindness in our patient.

Keywords: Rhabdomyosarcoma, Vision Loss, Vision Salvage.

INTRODUCTION:

Soft-tissue sarcomas in the paediatric population account for 6% of all paediatric cancers.^[1] Rhabdomyosarcoma is the most common soft-tissue sarcoma in infants and children. The Intergroup Rhabdomyosarcoma Studies (IRS) have established that the ideal management of this disease is to use multimodality treatment, using a combination of surgery, chemotherapy, and radiotherapy.^[2] We present a case of a parameningeal rhabdomyosarcoma leading to unilateral visual loss and restriction of extra ocular movements. The visual loss was effectively managed by surgical excision of the tumor and high dose systemic steroids. The vision improved significantly following treatment.

CASE REPORT:

A ten year old boy, presented to our Out patient department with chief complaints of decreased vision in the left eye since three days, left sided nasal mass noticed since one month and intermittent episodes of left sided nasal bleed since 1 month. On examination the patient had a reddish mass protruding out of the left nasal cavity with external nasal, maxillary region swelling and a left sided soft palatal bulge.

Examination of the eye revealed that there was perception of light (PL) and projection of rays (PR) in the left eye. This progressed to absence of PR in the next 48 hours despite the administration of intravenous steroids. The left eye extra ocular movements were restricted in the left lateral quadrant. Relative afferent papillary defect (RAPD) was observed in the left eye.

Contrast enhanced CT PNS of the patient showed a left sided enhancing soft tissue lesion in the left nasal cavity, nasopharynx, maxillary sinus extending into the pterygopalatine fossa, infratemporal fossa, sphenoid sinus with intracranial extension into the parasellar location. No metastatic lymph nodes were seen. MRI with angiography showed a multilobulated enhancing soft tissue mass with epicentre in the region of the sphenopalatine foramen. Anteriorly there was extension in the superior orbital fissure till the orbital apex where it appeared to be abutting the rectus muscle. There was intracranial but no intradural extension with involvement of the left cavernous sinus with most of the mass lateral to the sinus (Fig. 1).

A high clinical suspicion of juvenile nasopharyngeal angiofibroma was kept in mind and preoperative embolization was performed 72 hours prior to planned surgical excision of the tumor by Weber-Fergusson approach. Intra operatively, the tumor was excised from the nose, nasopharynx, infratemporal fossa, oropharynx, parapharyngeal space and it was cut in the sphenoid. Blood loss was around 170 cc.

The patient was given injectable Methyl Prednisolone in the dose of 500 mg once a day for three consecutive days starting from the day of surgery. The patient's vision improved to finger counting at one foot in the superior and inferior temporal quadrants, 72 hours after surgery. Subsequently, there was improvement in vision to FC more than five feet in the superior and inferior temporal quadrants in the next three days. However, the restriction in left sided extra ocular movements in the left lateral quadrant persisted.

Histopathology revealed a high grade rhabdomyosarcoma of the embryonal type and Masson's trichome stain was positive for muscle (Fig. 2). The patient was referred to a medical oncologist in view of further management in the form of chemotherapy.

Fig 1: MRI of patient showing left sided nasopharyngeal mass with extension into the cavernous sinus abutting against the left optic nerve.

Fig II: Histological section showing round to oval tumor cells containing nuclei with open chromatin and prominent nucleoli (hematoxylin and eosin, original magnification x 400).

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DISCUSSION:

Rhabdomyosarcoma accounts for approximately 3.5% of the cases of cancer among children 0 to 14 years and 2% of the cases among adolescents and young adults 15 to 19 years of age.^[3] The most common primary sites for rhabdomyosarcoma are the head and neck, the genitourinary tract, and the extremities.^[2] In the head and neck region, three broad anatomic regions can be identified: orbital, parameningeal, and nonparameningeal. Parameningeal sites include the nasopharynx and nasal cavity, paranasal sinuses, infratemporal and pterygopalatine fossae, and middle ear.

It is usually a curable disease in most children with localized disease who receive combined modality therapy, with more than 70% surviving five years after diagnosis.^[2,4] Histologically, there are four subtypes: embryonal, alveolar, botryoid and pleomorphic. The embryonal subtype is the most frequently observed, accounting for approximately 60% to 70% of rhabdomyosarcomas of childhood.^[5] The prognosis for a patient with rhabdomyosarcoma is related to the age, site of origin, resectability, presence of metastases, number of metastatic sites, histopathology,^[2,6,7] and unique biological characteristics of rhabdomyosarcoma tumor cells.

In our patient, a prompt decision towards debulking of the tumor was taken considering the progressive visual loss and an attempt to salvage his vision was made. Reports of visual recovery following surgical excision of malignant tumors are rare. A 67% overall improvement in visual acuity following transthemoid optic nerve decompression in cases of compressive optic neuropathy secondary to neoplasia has been reported by Li et al.^[8] Patrocínio et al have reported vision recovery following excision of an angiofibroma.^[9]

Healey et al have recommended radical tumor resection whenever complete locoregional removal appears possible subsequently followed by chemotherapy.^[10] The administration of intravenous steroids was the first step on admission. While the patient received intravenous steroids, embolisation was achieved in view of the highly enhancing nature of the tumor on CT scan. The patient was given high dose systemic steroids postoperatively in order to give him the best chance of vision salvage. Li et al recommend a trial of corticosteroids as an initial step in the treatment of compressive optic neuropathy and surgical decompression only if there is no improvement after 48 hours.^[8] Thus, in our patient early surgical intervention combined with judicious systemic steroid therapy were a key to vision salvage however a long term follow up is required and a larger series of patients managed in a similar way is essential to substantiate the results.

KEY MESSAGES:

Prompt surgical intervention combined with judicious systemic steroid therapy were important factors in the improvement of visual function in this patient of embryonal rhabdomyosarcoma, however a larger series is required to arrive at a conclusion.

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