



FUNGATING ORBITAL MASS

Gursharn Singh Narang, Jivtesh Singh Pahwa.

Department of Pediatrics, Sri Guru Ram Das Institute of Medical Sciences and Research, Vallah, Amritsar, India.

Address for Correspondence: Dr Gursharn Singh Narang, Associate Professor, Department of Pediatrics, SGRDMISAR, Vallah, Amritsar. Email: jivteshpahwa@rediffmail.com.

Four year old boy, presented with painless, rapidly progressing protrusion of right eye ball since last 8 months. Later on, proptosis progressed and a mass appeared in between eyelids. This mass slowly enlarged in size and presented as fungating mass since

SPOT DIAGNOSIS (IMAGE GALLERY)

last 4 weeks with well defined margins and was firm on palpation. Patient at present had complete loss of vision. On examination right eye showed fungating mass measuring 12cm ×8 cm× 5.5 cm, with superficial necrosis. Left eye was apparently normal. No family member or relatives have been diagnosed to have eye tumors in the past. It was associated with ipsilateral cervical lymphadenopathy. Mild pallor was present. Systemic examination was normal.

What is the diagnosis?

It is a Rhabdomyosarcoma of the orbit. Histopathology of the mass showed embryonal rhabdomyosarcoma. According to the Intergroup Rhabdomyosarcoma Study {IRS} the tumor was staged in stageI, groupIII. Patient underwent surgery for debulking of the tumor followed by radiation therapy.

Rhabdomyosarcoma {RMS} is a rare tumor, with an annual incidence of 4.3 cases per million children. {1} The orbit is the primary site in approximately 10 percent of these tumors. {2,3} RMS is the most common malignant orbital tumor of childhood. RMS should be suspected whenever the clinical presentation of a rapidly progressive unilateral exophthalmos is observed in a child. Although the superior nasal quadrant of the orbit is the most common location of this tumor, it may also present as a palpable subconjunctival or eyelid nodule with edema of the lids and, or chemosis. Following the biopsy, the tumor should be staged according to the classification systems of the Intergroup Rhabdomyosarcoma Study {IRS} and American Joint Commission on Cancer manual. {4} CT and MR images are fundamental in the preoperative evaluation to determine location and size, but they are also important in evaluating residual or recurrent disease. RMS is best managed with combination treatment of chemotherapy, external-beam radiation therapy, and surgery. {5} Exenteration of the orbit is now confined to cases with recurrent disease. {6}

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