10.5005/jp-journals-10013-1168

CASE REPORT

Esthesioneuroblastoma in Young Boy: Masquerading as Invasive Aspergillosis

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ABSTRACT

We present the case of a 15-year-old boy with a swelling and pain in right eye. He was diagnosed as esthesioneuroblastoma on the basis of radiology and histopathology.

Keywords: Small round cell tumor, Esthesioneuroblastoma, Proptosis.

How to cite this article: Bakshi J, Dash AK, Gupta B, Panda NK. Esthesioneuroblastoma in Young Boy: Masquerading as Invasive Aspergillosis. Clin Rhinol An Int J 2013;6(3):118-120.

Source of support: Nil

Conflict of interest: None declared

INTRODUCTION

Esthesioneuroblastoma (ENB), also known as olfactory neuroblastoma, is a rare neoplasm originating from olfactory neuroepithelium. Approximately 1,000 cases have been identified since Berger and Luc described the first case in 1924. Due to the rare and complex nature of ENB, multiple opinions exist regarding the etiology, optimal staging system, and treatment modalities. These tumors often display varying biologic activity ranging from indolent growth, with patient survival exceeding 20 years, to a highly aggressive neoplasm capable of rapid widespread metastasis, with survival limited to a few months.[1]

CASE REPORT

A 15-year-old boy presented with protrusion of right eye of 1 month duration and patient underwent endoscopic clearance from ethmoids under general anesthesia (Fig. 1).

Fig. 1: Patient with right proptosis

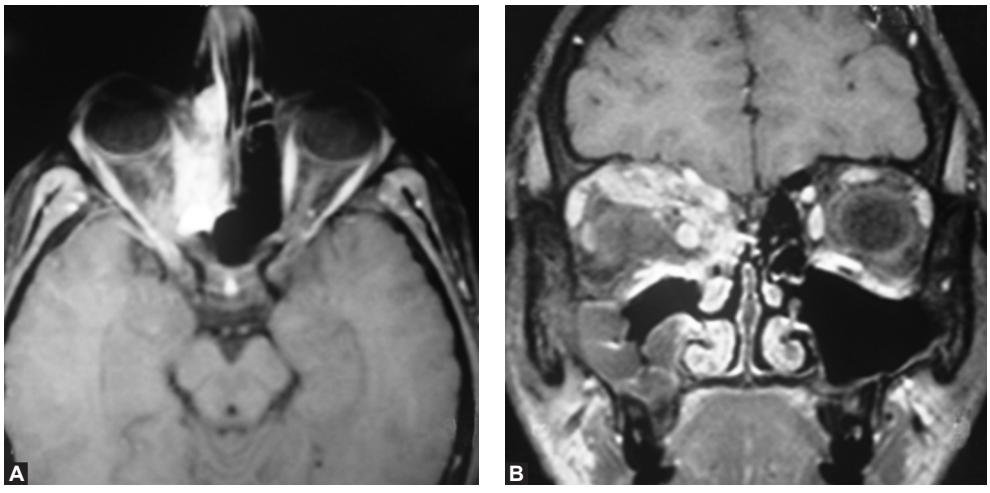
The biopsy report came out to be invasive aspergillosis. The fungal smear was negative. The patient was given oral voriconazol for 4 months. As the proptosis did not regress after medical treatment for 4 months, external ethmoidectomy was done under general anesthesia. There was a whitish grey mass in anterior ethmoid and frontal sinus, which was observed intraoperatively. The orbital periosteum was intact. The biopsy report came out to be small round cell tumor, suggestive of esthesionuroblastoma. Further immuostaining could not be done as patient was too young, in view of delaying management. The CT scan and MRI scan was done for this patient to note the extent of disease preoperatively. NCCT PNS and orbit showed heterogeneous extracranial soft tissue in right orbit and right ethmoid sinus with erosion of surrounding bone, intracranial extension and extraocular muscles involving medial and inferior rectus muscles (Figs 2 and 3). After biopsy report this patient was managed by chemotherapy and radiotherapy. Six months post-treatment patient is doing well without any local recurrence.

DISCUSSION

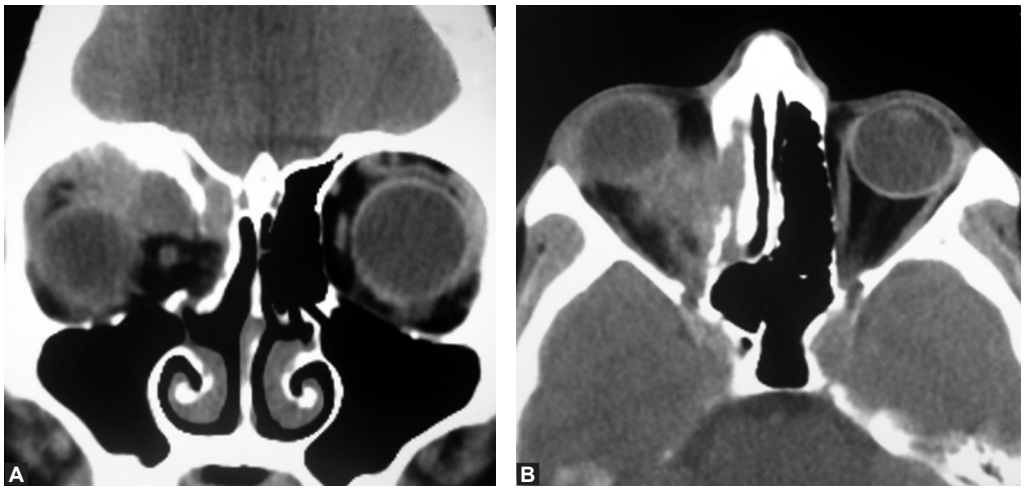
The diagnosis of esthesioneuroblastoma is based on radiology and histopathology. The radiological investigation includes CT scan and MRI. The characteristic MRI finding is the presence of cyst at brain-tumor interface. Using MRI, ENB appears as hypointense to gray matter on T1-weighted images and isointense or hyperintense to gray matter on T2-weighted images.[2]

Esthesioneuroblastomas (ENBs) can display various histologic presentations. The hallmark of well-differentiated ENBs is arrangements of cells into rosettes or pseudorosettes (sheets and clusters). True rosettes (Flexner-Wintersteiner rosettes) refer to a ring of columnar cells circumscribing a central oval-to-round space, which appears clear on traditional pathologic sections. Pseudorosettes (Homer-Wright rosettes) are characterized by a looser arrangement and the presence of fibrillary material within the lumen.[3]

Due to the rarity and complexity of esthesioneuroblastoma (ENB), there exists considerable heterogeneity in treatment. Complete surgical resection of the tumor followed by radiation therapy is recognized by most studies as the optimal treatment. However, some institutions report success with alternative treatment sequences, including surgery without radiation. More recently, chemotherapy has been introduced



Figs 2A and B: T1 weighted MRI images (axial and coronal) showing right medial and inferior rectus involvement



Figs 3A and B: NCCT PNS depicting intraorbital disease

in the therapeutic armamentarium. The literature gives little support to single-modality treatments; few studies advocate either surgery or radiation alone. Dulguerov's 2001 meta-analysis clearly showed lower recurrence rates for the combination of surgery and radiotherapy.[4]

Most institutions favor surgery as the first treatment modality, followed by postoperative irradiation. Preoperative radiation results in the usual loss of definable tumor borders, which makes an en-bloc resection problematic. However, it has been noted that a theoretical advantage to preoperative radiation therapy is to convert an inoperable tumor to one that is amenable to resection. This theory is not widely supported.

Standard techniques include external megavoltage beam and a 3-field technique; an anterior port is combined with wedged lateral fields to provide a homogeneous dose distribution. The radiation portals are nowadays planned by integrating pretreatment CT or MRI imaging within the radiotherapy software. The dose varies from 5500 to 6500cGy. The majority of patients receive <6000 cGy. These doses are close to or do not exceed the maximum radiation dose recommended for sensitive structures, such as the optic nerve, optic chiasma, brainstem, retina and lens. Therefore, these patients are susceptible to cataract and glaucoma formation. A possible role of proton beam radiotherapy, intensity-modulated radiotherapy, and stereotactic radiation has been suggested. Several institutions have reported that intensity-modulated radiotherapy can provide good tumor control with low rates of radiation-induced toxicity, in children as well as in adults. There are case reports describing the use of CT-guided interstitial high-dose-rate brachytherapy. However, prospective clinical trials confirming the efficacy of these modalities have not yet been completed.[5-9]

The use of chemotherapy has been advocated by authors from the University of Virginia In their protocol, patients with advanced disease (e.g. Kadish stage C) are treated first with 2 cycles of cyclophosphamide (300-650 mg/m^2) and vincristine (1-2 mg) with or without doxorubicin, followed

by 50 Gy of radiotherapy, which then is followed by a craniofacial resection. With this regimen, the 5-year and 10-year actuarial survival rates are 72 and 60% respectively. Similar results have been obtained without chemotherapy, and how much chemotherapy contributed to the cure rates is unclear.[10]

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