A Case of Rhabdomyosarcoma in Adolescents Mimicking Sinus Abscess Causing Rapid Bilateral Blindness

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Abstract

Rhabdomyosarcoma is a very bad type of cancer that develops on skeletal muscles like muscles in the eye. It usually affects children and in less commonly affects adult. Its diagnosis depend on tissue biopsy and clinical presentation which sometimes is very subtle and can mimic infectious diseases.

Keywords: Rhabdomyosarcoma; Acute Pyogenic Sinusitis; Sub-Periosteal Abscess

Introduction

Orbital masses have a long list of differential diagnosis. On the top list is infectious disease and malignant masses. Rhabdomyosarcoma is a very malignant tumor. It is the most common soft-tissue sarcoma of the head and neck in childhood. Orbital Rhabdomyosarcoma is one of the few life-threatening diseases that present first to the ophthalmologist; therefore prompt diagnosis and treatment is a life-saving issue. In this case report we present a case of young patient presented with a subtle symptoms of an orbital mass.

Case Report

16-year-old female patient presented to ophthalmology clinic at Al-Mowasat University hospital complaining of proptosis (Figure 1), lid swelling and pain in her right eye that had persisted for 2 weeks. The patient had a significant past history of chronic sinusitis. There was no other past medical history or family history associated with ocular disease, trauma, or tumors. Exophthalmometer measurements were 25 mm on the right, and 18 mm on left. Eye movement was slightly limited in adduction in the right eye with diplopia presented when the patient looks at the left. The rest of ophthalmologic examination including visual acuity, color vision, and fundoscopy were normal. Full panel tests and thyroid function test were also normal. Laboratory test revealed a high white blood cells count which indicates infectious pathology. A computed tomography (CT) scan revealed a bilateral opacities of the frontal, sphenoidal and ethmoidal sinuses with extension to the right orbit. There was also a reactive thickening of the surrounding meninges in the base of the skull and anterior cranial fossa (Figure 2). The initial diagnosis was acute pyogenic sinusitis with sub-periosteal abscess in the right orbit. The initial management for this abscess was drainage with parenteral broad spectrum antibiotics. Two days later the patient developed a seizure. She transferred to ICU. Lumber puncture analysis was within normal limits. The patient was continued on antibiotics with addition of anticonvulsant drugs. After two weeks of antibiotic treatment, the CT scan revealed no improvement and the visual acuity in the right eye started to deteriorate to reach count finger from 3 meters. Optic disc edema and RAPD (relative afferent papillary defect) developed in the right eye. Magnetic resonance imaging (MRI) showed a mass that its signal intensity was low on a T1-weighted image with contrast slightly enhancement and isointense to muscles on a T2-weighted image (Figure 3). An emergent intervention and fiberoptic endoscopic biopsy of the nasal cavity to reduce the compression was done. Biopsies were taken. Microscopic examination revealed diffuse small round cells with abundant eosinophilic cytoplasm. Immunohistochemical analysis was positive for DES (desmin) and negative for CD20, CD03, NSE

(neuron specific enolase), LCA (leukocyte common antigen), Synaptophysin. The diagnosis was alveolar rhabdomyosarcoma. Metastatic work-up revealed right cervical lymph nodes involvement, indicating a stage IV RMS based on the Intergroup Rhabdomyosarcoma Study (IRS) TNM Pretreatment Staging System. The treatment was radiation therapy and chemotherapy. The chemotherapy included Vincristine, Doxorubicin, Cyclophosphamide, combined with radiation of the orbit and ethmoid sinuses. Unfortunately, during that period the patient’s visual acuity in the left eye started to deteriorate to reach count finger from 5 meter; so the patient was continued on chemotherapy, after 4 week the deterioration in visual acuity stopped and the patient’s proptosis started to reduce in the right eye.

Figure 1

Figure 2

Discussion

Orbital rhabdomyosarcoma usually affects young children with a mean age at diagnosis of 8 years, but it can occur at any age and relatively uncommon in adult. The most characteristic presentations are rapid onset and progression of proptosis, and displacement of the globe. Because Rhabdomyosarcoma are very rapidly growing with all inflammatory signs, it may be misdiagnosed as orbital cellulitis. If rhabdomyosarcoma is suspected, CT and MRI can be used to define the location and extent of the tumor. CT is particularly helpful if tumor has caused bony destruction, although the orbital walls remain intact in most cases [1,2].

Presence of an orbital tumor should be suspected if there is no response or worsening despite adequate intravenous antibiotic therapy, and even if there was a pus in the area of the mass. The only way to make the diagnosis definite is by doing biopsy.

Conclusion

The ophthalmologists should be very careful in approaching orbital mass that may have an infectious symptoms to save patients early and in an efficient way.

Bibliography
