Case Report

A Case of Intraorbital Intracanal Lymphangioma with Post-traumatic Apoplexy

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Orbital lymphangioma is an infrequent benign cystic lesion manifesting in childhood and presenting with slowly progressive proptosis, and restriction of eye movements. Here we report a rare case of 8 year old male patient presenting with unilateral painful proptosis and subconjunctival hemorrhage with decrease of vision and restriction of eye movements. CT scan and MRI were done which revealed an intraorbital and intracanal cystic space occupying lesion. Fronto-orbito zygomatic craniotomy was done for orbital decompression and subtotal excision of tumor and blood evacuation. Histopathological findings were suggestive of lymphangioma.

Keywords: Intraconal, intraorbital, lymphangioma, orbital decompression, proptosis

INTRODUCTION

Orbital lymphangioma is a rare, benign, slow-growing, cystic, vascular malformation constituting up to 0.3%–1.5% of diagnosed orbital tumors. It occurs in children and teenagers, but more frequently during the first decade of life. Spontaneous hemorrhages can occur in 55% of the cases. These cases may often present with acute proptosis and loss of vision. Different modes of treatment for orbital lymphangioma are conservative management, partial resection of the cystic lesion, and local radiotherapy. Orbital decompression can also be performed occasionally to restore vision. Recurrences are quite common as complete surgical resection is very difficult.

CASE REPORT

An 8-year-old male patient was brought to outpatient department with complaints of bulging of left eye for 10 days. The patient was apparently asymptomatic before 10 days, following which he started developing bulging of the left eye, gradually progressive, associated with continuous pain and watery discharge from the left eye and continuous frontal headaches. The child's father reported a history of trauma 12 days back. The child denies the history of fever, a decrease of vision, or vomiting. The child did not have a history of surgery or upper respiratory tract infection.

Orbital lymphangioma is an infrequent benign cystic lesion manifesting in childhood and presenting with slowly progressive proptosis, and restriction of eye movements. Here we report a rare case of 8 year old male patient presenting with unilateral painful proptosis and subconjunctival hemorrhage with decrease of vision and restriction of eye movements. CT scan and MRI were done which revealed an intraorbital and intracanal cystic space occupying lesion. Fronto-orbito zygomatic craniotomy was done for orbital decompression and subtotal excision of tumor and blood evacuation. Histopathological findings were suggestive of lymphangioma.

KEYWORDS: Intraconal, intraorbital, lymphangioma, orbital decompression, proptosis

On examination, there was a unilateral proptosis of the left eye with subconjunctival hemorrhage and restriction of eye movements. There was a loss of vision in the affected eye (counting fingers up to 3 m). Computed tomography (CT) scan revealed multiple, hypodense, nonenhancing, intracanal lobulated lesions. Magnetic resonance imaging (MRI) revealed a heterogeneous cystic multiseptated soft-tissue lesion measuring 2.4 × 2.5 × 3 cm with intracanal and extraconal component noted in the left orbit [Figure 1]. The cyst contained fluid of varying intensity suggestive of left orbital lymphangioma with internal hemorrhage. Left orbitozygomatic craniotomy was performed, and the cystic space–occupying lesion was excised and hematoma was evacuated. The specimen was sent to histopathology examination, which suggested features of lymphangioma [Figure 2]. The postoperative course was uneventful with good aesthetic result, complete reduction of proptosis and pain, and improvement of vision [Figure 3]. The patient was followed up at the end of 1 and 3 months and had a good recovery with no recurrence.

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Orbital lymphangiomas are rare benign vascular tumors and constitute up to 0.3%–1.5% of histopathologically diagnosed orbital tumors. These tumors are usually congenital and slow growing, and may not become apparent for months or years. Lymphangiomas are more common in males. Histologically, lymphatic malformations consist of nonencapsulated channels lined by a single layer of endothelium. The International Orbital Society classified the orbital vascular malformations into three groups based on their clinical and hemodynamic relationships: (1) no-flow malformations, (2) venous-flow malformations, and (3) arterial-flow malformations. The lymphangioma or lymphatic malformation falls into the no-flow malformation category because it is hemodynamically isolated.

Most of the lesions are asymptomatic until hemorrhage or upper respiratory tract infection occurs, resulting in proptosis, decreased vision, decreased motility, and pain. CT and MRI show a cluster of nonenhancing “grape-like” cystic lesions with internal septations. Old bleeds within the lesion appear hyperintense on T1 imaging, whereas recent ones appear hypointense. Lymphangiomas can be classified radiographically as macrocystic (cystic structures >1 cm in size), microcystic (cystic structures <1 cm in size), or combined.

Treatment options for lymphangioma include conservative therapy, partial surgical excision of the cystic lesion, intralesional injection of the sclerosing agent, and local radiotherapy; in case of intralesional hemorrhage, orbital decompression can be performed. Recurrences are quite common as complete surgical excision is very difficult.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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