Orbital lymphangioma: Considerable Shrinkage without Biopsy and Surgery

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Abstract

A 5-year-old girl presented with a sudden left proptosis just after being hit by her sister’s hand. The Hertel measurements were 8 mm OD and 12 mm OS. Computed tomography scans disclosed an extensive multi-cystic mass in the left orbit with niveau formation. Based on these findings, the tumor was diagnosed as “orbital lymphangioma”. Since the patient could close her eyes completely without corneal involvement and visual acuity loss, we did not perform a biopsy or surgery. Four months later, her left proptosis improved (Hertel: 10.5 mm). Five months after this examination, her left proptosis further improved (Hertel: 8 mm, OU). The proptosis showed a symmetrical appearance. Computed tomography scans showed considerable shrinkage of the tumor. This is the first case report of an orbital lymphangioma with detailed description of considerable spontaneous shrinkage without stimulation from a biopsy and surgery.

Keywords: Orbital lymphangioma; Multi-cystic mass; Shrinkage; Biopsy; Surgery

Introduction

Orbital lymphangiomas are diffusely infiltrating, benign vascular tumors that appear most commonly in the first 2 decades of life [1]. Acute hemorrhagic proptosis is one of the most frequent presentations, which is caused by either spontaneous or traumatic hemorrhage into the tumor spaces [2]. Although multiple subtotal excisions have long been recommended for management of lymphangioma [1], conservative management, including simple observation [1,3] and/or systemic corticosteroid therapy [4], has recently become popular with satisfactory outcomes. However, as almost all the conservative therapies are performed after biopsy, it is possible that tumor size is reduced by stimulation from the biopsy. Currently, however, a biopsy is not always required for diagnosing lymphangiomas because of an improvement in orbital imaging technology [5]. Three cases of orbital lymphangiomas have been previously reported showing considerable spontaneous tumor shrinkage without biopsy and surgery. However, a detailed clinical course of orbital lymphangioma has not been reported yet [1,5].

We report a case of orbital lymphangioma showing considerable spontaneous shrinkage without stimulation from a biopsy and surgery was not performed.

Case Report

A 5-year-old girl presented with a sudden left proptosis just after being hit by her sister’s hand (Figures 1A&1B). Two weeks later, she came to our outpatients’ clinic. Her mother said she had not shown proptosis before that incident. The corrected visual acuity was 1.2 OU. The Hertel measurements were 8 mm OD and 12 mm OS. The eye movements were full and smooth in both eyes. She could close her eyes completely. Although no corneal, lenticular or fundic lesions were demonstrated, she showed a small subconjunctival hemorrhage in her left eye. Axial computed tomography (CT) scans disclosed an extensive multi-cystic mass in the left orbit with niveau formation (Figure 1C). Coronal CT scans illustrated a mass occupying the temporal upper part of the left orbit pushing the optic nerve inferonasally (Figure 1D). Based on these findings, the tumor was diagnosed as “orbital lymphangioma”. Since she could close her eyes completely without corneal involvement and visual acuity loss, we did not perform surgery including a biopsy.

Four months after the first examination, her left proptosis improved and the Hertel measurements were 8 mm OD and 10.5 mm OS. Five months after the second examination, her left proptosis further improved. The Hertel measurement was 8 mm OU. At the latest examination, 9 months after the third examination, the proptosis had a symmetrical appearance (Figures 2A&2B) and the Hertel measurement...
was 8.5 mm OU. CT scans showed considerable shrinkage of the tumor without shifting of the optic nerve (Figures 2C&2D).

Discussion

We present a case of orbital lymphangioma showing considerable spontaneous shrinkage without stimulation from a biopsy and there was no surgery.

Management of patients with orbital lymphangioma has been controversial [1]. Multiple subtotal excisions, removing as much of the lymphangioma as possible, has long been recommended [1]. Complete excision is usually not possible because of the diffuse, noncapsulated growth pattern that allows these tumors to interdigitate with normal orbital structures [1]. Lately, conservative management has become an alternative choice for management of orbital lymphangioma [1,3,4]. Although conservative, most of them were performed after biopsy for diagnosis and there is the possibility of tumor size reduction by stimulation from the biopsy. However, the present study demonstrated considerable spontaneous shrinkage of an orbital lymphangioma without stimulation from biopsy.

Diagnosis of orbital lymphangioma has long depended on biopsies [1,6]. However, current imaging methods have enabled noninvasive diagnosis in almost all cases of orbital lymphangioma [5]. Orbital lymphangioma is diagnosed if there is a multi-lobulated pattern and a cystic internal structure [5]. The current orbital imaging technology permits simple observation of some patients and provides graphic postoperative follow-up data in others [5]. Imaging also broadens the topographic perspective of lymphangioma relative to normal structures, enhancing the perceptions gained from the surgical field and the pathologist’s microscope [5]. Based on these improvements, further observations of orbital lymphangiomas should now be possible.

This is the first case report of an orbital lymphangioma with detailed description of considerable spontaneous shrinkage without stimulation from a biopsy and surgery. Although surgery needs to be performed for patients with visually threatening or cosmetically disfiguring disease [5], conservative management is recommended as a first-line management for orbital lymphangioma.

References

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