CASE REPORT

ORBITAL LYMPHANGIOMA

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ABSTRACT:

The objective of this case report is to emphasize that in any case of proptosis in childhood orbital lymphangioma should be considered. Tissue diagnosis, along with imaging studies, is the best and most consistent confirmation of lymphangioma. The treatment of orbital lymphangioma has to be as conservative as possible if vision is not at risk and cosmesis is acceptable.

Key words: Lymphangioma, Proptosis, Sclerotherapy.

INTRODUCTION

Lymphangiomas are localized malformations in the development of the lymphatic system that most frequently affect the head and neck 1,2 followed by the proximal extremities, the buttocks, and the trunk. Incidence has been reported to be less than 2.8 per 1000 population. No sex preponderance or site predilection has been reported. Although 20% of lymphangiomas involve the orbit and ocular adnexa,3,4 they account for nearly 2% of orbital biopsies.3 Patients with orbital lymphangiomas may be asymptomatic, or they may experience symptoms such as pain, blurred vision, or double vision. Presenting signs include slowly progressive proptosis, vertical globe displacement, restriction of eye movements, and ptosis.4,5 In addition, spontaneous hemorrhage into vascular channels can result in acute proptosis, compressive optic neuropathy, and loss of vision.5,6

The clinical characteristics of the lesions, and their anatomic features (particularly their relationship to the vascular supply of the orbit) may overlap those of the other orbital vascular lesions.5-7 Tissue diagnosis, along with imaging studies, is the best and most consistent confirmation of lymphangioma.

The treatment of choice for lymphangiomas is surgery. The primary intention is to accomplish total resection. However, because of lesion size and location, total resection is not always possible. If significant cosmetic or functional deficits are probable, consider partial staged reduction or alternative therapy. Alternate therapy has been proposed as the primary treatment for lymphangiomas, particularly for sensitive (eg the orbit) and, more commonly, for recurrent disease, after surgical therapy. Radiation therapy has been effective but abandoned because of later malignant transformation or retardation of growth. Intralesional sclerotherapy with group A streptococcus pyogenes of human origin (OK 432) has had some success controlling lymphangiomas.

CASE REPORT

10 months old male child presented with history of fever for 1 month and right orbital swelling since birth.

The orbital swelling had variable increase in size but for the last 2 weeks the swelling had increased significantly, and also developed signs of inflammation with pus discharge from right eye. For these complaints he was treated by various doctors, but there was no improvement and finally he was admitted in Children Hospital, Baqai Medical University.

On examination, child was febrile with temperature of 100°F. His anthropometric measurement were on 25th percentile. Child had right orbital swelling with signs of inflammation. Vision was intact. No ear discharge, lymphadenopathy, hepatosplenomegaly and no petechiae or bleeding from any site.
On the basis of history and clinical examination the differential diagnosis was Retinoblastoma, Neuroblastoma, Rhabdomyosarcoma, Ruptured dermoid cyst.

Complete blood count revealed Hb 10.0g/dl, Total leucocyte count 24,500/cumm, Neutrophils 80%, Lymphocytes 14%, Eosinophils 2%, Monocytes 4%. No blast cell seen on peripheral smear. His serum electrolytes, urea & creatinine were normal, X-ray chest revealed no mediastinal mass.

MRI of the brain revealed evidence of large abnormal signal intensity mass lesion in right retrobulbar region involving the extraocular muscles causing proptosis of right eye. The mass was encasing the optic nerve. Eye globe appeared normal. The mass was causing expansion of right orbit. Soft tissue density area was seen in right frontal region. No intra cranial extension of the mass was found. Appearance suggestive of malignant lesion most likely Rhabdomyosarcoma. The left eye appeared normal.

Right orbital biopsy was done, which revealed a benign vascular lesion composed of variable sized vascular channels lined by endothelial cells. Thick layer of fibroadipose tissue was also identified around the vessels. Mild focal chronic non specific inflammatory cells also identified. On immunohistochemistry the endothelial cells show positivity for CD31. No evidence of granuloma or malignancy seen.

On the basis of orbital biopsy, features favour lymphangioma.

After the consultation with Ophthalmology department and Paediatric surgery, the child was put on conservative management because spontaneous resolution has been reported in number of cases.

**DISCUSSION**

Lymphangiomas in general are uncommon congenital malformations of the lymphatic system that mostly involve the head and neck, skin and subcutaneous tissues. However sometimes they can be found in the intestine, pancreas, and mesentery. Deeper cystic lesions usually occur in areas of loose and areolar tissue, typically the neck, axilla, and groin. Children or, less commonly, adults usually present with a mass in the head and neck area. Approximately 90% cases of cervicofacial lymphangioma become clinically apparent by the third year of life. Common head and neck sites in childhood are the cervical area, floor of the mouth, and tongue. Orofacial manifestations include mandibular and maxillary deformation. In a study by Orvidas et al, the most common location involved was the submandibular region followed by the parotid and cheek. In approximately half of these patients, the lesion is present at birth, and in most cases the diagnosis is made before 2 years of age. In imaging lymphangiomas, plain skull radiographs and venograms were routinely obtained before 1974, however, CT and MRI imaging have largely replaced venography because they provide superior information regarding vascular flow and tissue details, without the potential complications of venography, such as vascular stasis, hemorrhage, and loss of vision. On histologic examination, these masses are composed of dilated cystic spaces containing blood or lymph lined by endothelial cells with a scant-to-dense fibrous stroma. Cells in the stroma include lymphocytes, lymphatic tissue, fat, or muscle. Spaces vary in size, from capillary to cavernous channels often resembling a Swiss cheese pattern. Because of the variable anatomic features of orbital lymphangiomas, their management is problematic. Observation is the preferred method of management for many of these lesions. Spontaneous resolution is seen and in some studies it is reported to be as high as 41%. Complete surgical resection of diffuse lesions is impossible, and even attempts at partial resection can result in significant bleeding and tissue injury with loss of vision or double vision. With incomplete excision, disease recurrence is extremely high. Cranial nerve injury exceeds the rate of 20%; the facial nerve is the most common neural deficit reported. Treatment of lymphangioma is typically indicated when it is associated with rapid growth in size, optic nerve compression, corneal exposure problems (Keratitis sicca), glaucoma or evidence of vision loss. When treatment of lymphangioma is considered, the goal is rarely complete removal. This is because the edges of most orbital lymphangiomas are poorly defined. Most patients undergo several debulking surgeries to relieve acute optic nerve compression or corneal exposure.
In rare cases orbital lymphangioma patients may require exenteration of the orbit, radiation therapy for the relief of pain. To reduce surgical morbidity and to decrease recurrence rates a variety of nonsurgical treatment have been proposed, including radiation, diathermy, and injection of a variety of agents such as bleo sulfate, triamcinolone acetonide, interferon alfa, fibrin fibronectin sealing systems, and alcoholic of Zein (corn protien). Because these treatment options have met with limited success and cause considerable local or systemic adverse effects, recent interest has focused on the sclerosant OK-432, as an alternative therapy. In the past 14 years, a number of reports have described the successful use of OK-432 for the treatment of lymphangiomas of the head and neck in children. This potent immunostimulant is a lyophilized mixture of a low-virulence strain (Su) of group A Streptococcus pyogenes incubated with benzylpenicillin. Intravascular sclerosis therapy with group A streptococcus pyogenes of human origin (OK-432) has had some success controlling lymphangiomas. The mechanism suggested is the stimulation of increased permeability of the endothelium, accelerating lymphatic fluid drainage and size reduction of the lymphangioma.

Many authors, predominantly from Japan, have recommended its use as therapy in the treatment of lymphangiomas.

CONCLUSION

Orbital lymphangioma should be considered in any case of proptosis in childhood. Associated extra orbital localization, particularly intracranial vascular anomaly, must be ruled out. A multidisciplinary approach is needed, and the treatment has to be as conservative as possible if vision is not at risk and cosmesis is acceptable.

REFERENCES


