

Case report

Cavernous lymphangioma of eyelid - a rare case report

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Abstract

Background: Lymphangioma is a lymphatic malformation, a benign proliferation of lymph vessels.

Case: We hereby present a case of eyelid lymphangioma of cavernous type in a twelve year old male patient. This is a very uncommon site for this type of lymphangioma.

Key words: lymphangioma, eyelid, cavernous type

Introduction

Lymphangioma is a lymphatic malformation, a benign proliferation of lymph vessels forming a yellowish tumor on the skin. It is composed of a mass of dilated lymph vessels. It is a rare tumor, incidence being estimated at around one per 10,000 live births (Ng et al 2001). It may occur anywhere, but around 75% occurs in the head & neck region. Most (around 90%) are either evident at birth or become evident before age of two years (Schwartz et al 2008). Ocular lymphangiomas can occur in orbit, conjunctiva and eyelids but eyelid lymphangiomas are very rare (Jones et al 1961). Most common location for ocular lymphangioma is orbit. Most of the eyelid lymphangiomas reported are of circumscriptum type (Jones et al 1961, Flanagan et al 1977, Goble et al 1990) although a few reports of cavernous lymphangioma involving the eyelid are also available in literature (Jones et al 1961, Pang et al 1984). We present a rare case of subcutaneous cavernous lymphangioma of eyelid.

Case report

A 12-year-old male child presented with a painless bulging mass under the skin of the left upper eyelid,

just below the eyebrow. The parents of the boy noticed the peanut size swelling when the boy was four years old. This swelling gradually increased in size since then but during last four years the size remained static. On examination, a 2 cm x 2cm soft, painless mass with a smooth surface was seen at the supero-nasal aspect of the left eyelid. CT scan of the orbit revealed a soft tissue mass lesion supero-medially (21.7mm x 11.5mm x 21mm) in the left eyelid, in the subcutaneous plane near the base of the nasal bone surrounded by the fatty tissue. A provisional diagnosis of dermoid was made.

The lesion was excised under general anaesthesia; the plane of incision was at the submuscular (below orbicularis oculi muscle) level. The lesion was not encapsulated but was found to be adherent to the surrounding structures.

Gross examination revealed a single grayish brown soft to firm tissue measuring 2 cm x 1.5 cm x 0.5 cm. Cut surface was variegated, bisected and both the halves passed in a single cassette. The microscopic features showed a circumscribed area containing many irregular, compressed and dilated spaces lined by simple squamous epithelium (fig-2). Large lymphatic spaces of varying size and shape, lined by simple squamous epithelium were present. These spaces were separated by collagen tissue. Some spaces were present in between muscle bundles also. There was no evidence of

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dermoid cyst. The histopathological examination was consistent with that of cavernous type of Lymphangioma. No recurrence was seen after 6 months of follow up.

Figures



Fig.1 A 2x2 cm swelling in superior-nasal aspect of left eye



Fig. 2 CT showing soft tissue mass in subcutaneous plane



Fig-3: Cavernous lymphangioma of eyelid showing many irregular, compressed and dilated spaces lined by simple squamous epithelium (H&E 40x) There are large lymphatic spaces of varying size and shape lined by simple squamous epithelium

Discussion

Lymphangiomas are rare. They account for 4 % of all the vascular tumors and approximately 25 % of all the benign vascular tumors in children. There is no sign of racial predominance. Also equal sex incidences are reported in most studies although some groups have reported that lymphangioma circumscriptum is more common in females while some report a 3:1 male-to-female ratio.

Lymphangioma is a hamartoma, or vascular developmental abnormality arising from lymphatic vessels, appearing as a raised, soft, shaggy, bubbly, pinkish-white lesion. Cosmetic considerations may warrant attempted removal of lymphangiomas.

Lymphangioma are of three types - cystic lymphangioma (or cystic hygroma), lymphangioma cavernosum and lymphangioma circumscriptum. Lymphangioma circumscriptum is a type of birthmark that generally occurs in clusters. They look a lot like small blisters and range in color from pink to dark red. They are benign and do not really require any medical treatment. This is also a type of tumor that is commonly seen in younger children (Schwartz et al 2008). Lymphangioma circumscriptum is thought to represent a developmental abnormality. The subepidermal lymphatic 'vesicles' are endothelium-lined spaces which are completely separate from the normal lymphatic system (Whimster et al 1976). Cavernous lymphangioma is an uncommon form, generally arising during infancy. This occurs deep under the skin and the external skin is not involved. Because they are deep seated, they form a bulging mass that is usually painless. This is also a tumor filled with lymph that is often mixed with blood. It usually affects the tongue and lips (Peachey et al 1970).

Cystic lymphangioma or Cystic hygroma are soft lymph filled masses within a thin-walled "sac" generally appearing on the neck (75 %), arm pit or groin areas. They often look like swollen bulges underneath the skin. While they are generally present at birth, children up to the age of three years may experience them. There is also disagreement on whether this type is distinctly different enough from



the cavernous lymphangioma to be classified separately.

Lymphangiomas that are deep in the dermis show no evidence of communication with the regular lymphatics. The cause for the failure of lymph sacs to connect with the lymphatic system is not known.

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