Raised Superior Bilateral Conjunctival Lesions: An Exploration of Conjunctival Lymphangioma and Haemangioma

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Case Report

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Abstract

This report is based on the case of a young 7-year-old child who presented with a four-month history of raised superior conjunctival lesions in both eyes and subsequently underwent a conjunctival biopsy in the right eye. He was not known to have any underlying congenital disorders and subsequent biopsy revealed an epithelium consisting of irregular, ectatic thin-walled channels lined by a bland single layered epithelium which expressed both CD31 and D240. No Cytokeratin (Cam 5.2) or EMA was expressed in the subsequent report. It was concluded that the features highlighted were likely due to an underlying conjunctival lymphangioma or haemangioma - which we will explore further.

Keywords: Bilateral Conjunctival lesions, epithelium, lymphangioma, haemangioma

Abbreviations

Cam 5.2	: Cytokeratin
CD31	: Platelet endothelial cell adhesion molecule-1
D240	: Podoplanin
EMA	: Epithelial Membrane Antigen
1A-4	: Pericyte/smooth muscle cell markers
VEGFR-3	: Vascular Endothelial Growth Factor Receptor 3

Conjunctival Lymphangioma

A Conjunctival lymphangioma is a rare disorder of the eye which appears as a multi-loculated cyst-like lesion on the surface of the conjunctiva. It is characterised by dilation of the lymphatic vessels and is also known to have the potential for recurrence [1]. They are benign and slow-growing; usually presenting as a mass - which generally does not affect the vision of the sufferer [2].

Usually, it is noted to be a unilateral condition - and it is somewhat rare to find patients with a bilateral manifestation. If found to be bilateral then it is generally known to be associated with Milroy's disease or Turner syndrome.

Milroy's Disease, also known as Nonne–Milroy–Meige disease, is a familial condition leading to lymphoedema due to abnormalities of congenital origin (inherited in an autosomal dominant manner) in the lymphatic system [3]. There have been reports that mutations in VEGFR-3 is associated with this condition from studies involving the phenotyping of patients. This growth factor is one of the most important associated with lymphangiogenesis. Other manifestations include oedema of the lower limbs, respiratory papillomatosis, varicose veins as well as upwards slanting of toenails. In males with the condition, an association with hydrocoeles has also been noted [4].

Conservative management is noted to be successful for many patients. Routine measures such as elevation of the limbs combined with adequate skin care and prolonged standing suffices for milder cases. Movement advice in the form of manoeuvres that facilitate manual lymphatic drainage have also been found helpful alongside compression and a myriad of other techniques. These include decongestive physiotherapy, bandaging, breathing exercises and dietary measures [5, 6]. If these measures are not helpful then surgical options are available and these revolve around the removal of lymphoedematous tissue and improving the functionality of the lymphatic system [7].

Turners syndrome condition which affected females. It is characterised by short stature and premature ovarian failure

amongst a myriad of signs such amenorrhoea, webbed neck and heart problems - including but not limited to aortic valve stenosis, coarctation of the aorta and bicuspid aortic valve (most common manifestation). It is caused by a partially or completely missing X-chromosome [8].

Treatment revolves around the use of Oestrogen and Growth Hormone replacement. Growth hormone, when given during maturation and development, has been shown to increase a patient's final adult height. Additionally, it has also been studied to reduce adipose tissue and increase lean body mass [9]. Oestrogen therapy is given around the time of pubertal development and this enables the development of the secondary sexual characteristics. In addition to this, it has also been shown to improve a patients psychosocial functioning whilst also increasing their bone mineral density [10].

Of note, in the pathology report, D240 was expressed and this has previously been reported to be a highly specific and sensitive marker for lymphatic endothelium in normal tissues and a subset of lesions - including lymphangiomas [11].

Conjunctival Haemangioma

A Conjunctival haemangioma on the other hand is also an uncommon and somewhat rare occurrence [12]. They are known to account for approximately 2% of neoplasms involving the conjunctiva [13]. Additionally, they tend to affect younger individuals [14]. Generally, in the second to third decade of life. It can present with bleeding of the conjunctiva [15]. Most of the cases reported in the literature are known to be congenital in origin, asymptomatic and usually involute by the age of seven [16].

It is pertinent to note that eyelid capillary haemangiomas can be a cause of cosmetic as well as functional concern. It had been noted that it can cause amblyopia and the underlying reason for this is the ptosis causing visual deprivation in this stage - or as a result of astigmatism. Contrastingly, in conjunctival haemangiomas this is not as much of a concern due to location – however the emphasis for careful observation remains [17].

On Pathological examination, in this condition it is common to find a surface epithelium which is intact with positive markers for CD31, CD34 alongside other markers such as IA-4 [18]. In this case CD31 was noted to be positive.

Management and treatment of conjunctival haemangioma involves taking into consideration the extent and size of the lesion. Frequent observation is therefore an integral part of the management. There are a multitude of treatment options which must be tailored to the individual patient. These include cryotherapy, radiotherapy, chemotherapy as well as modified enucleation and exenteration in more pressing cases [19]. It has previously been shown that treatment with beta-blockers in infants have also been of positive effect – however caution should be taken in patients with co-existing cardiovascular or pulmonary conditions [20, 21].

Conclusion

In conclusion it's important to review the histopathological marker findings alongside monitoring observations and clinical manifestations. At the same time it is essential to keep in mind an individuals medical history (including congenital abnormalities as discussed) to elucidate between underlying pathologies behind conjunctival lesions.

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