Solitary fibrofolliculoma of the eyelid
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Abstract

Purpose: A solitary fibrofolliculoma of the eyelid is a rarely reported entity. We report a case of a solitary fibrofolliculoma in the eyelid of 32 a year old man. Case: A 32 year old man presented with a 3 cm x 4 cm swelling in his right upper lid for the past year and a half. It was previously diagnosed as a chalazion and incision-drainage was performed. The lesion grew back in 6 months’ time and caused mechanical ptosis for which the patient underwent excision biopsy and histopathological examination (HPE). A diagnosis of fibrofolliculoma was made on HPE. Conclusion: A solitary fibrofolliculoma of the eyelid is rare and it is essential to consider fibrofolliculoma as a differential diagnosis when evaluating eyelid tumours.

Keywords: Fibrofolliculoma, Birt–Hogg–Dubé syndrome, Eyelid tumour

A fibrofolliculoma is a fibrotic hamartoma characterized by abnormal hair follicles with epithelial strands extending out from the infundibulum of the hair follicle into a hyperplastic mantle of specialized fibrous tissue¹. The lesions were described by Birt et al. in a case series of 15 patients who presented with fibrofolliculoma, trichodiscoma, acrochordons, and connective tissue nevus. These traits were inherited in an autosomal dominant pattern. These are clinically manifested as papules, 2 to 4mm in diameter, dome-shaped, yellowish or skin-coloured, usually located on the head, neck, and upper trunk. When multiple, they are often numerous and are a marker for Birt–Hogg–Dubé syndrome (BHD). BHD is caused by mutations affecting the highly conserved protein folliculin (FLCN), which probably has a role in intracellular transport²,³.

Various reports are available in reported literature as far as BHD and multiple fibrofolliculomas are concerned but solitary lesions though are extremely rare, only 7 case reports worldwide (Medline search) and only one such report is of an eyelid tumour⁴,⁵,⁶,⁷,⁸,⁹,¹⁰.

Case:
A 32 year old man presented with a 3 cm x 4 cm swelling in his right upper lid for the past year and a half (fig. 1A). The swelling was firm, mobile and painless. It was overhanging and encroached onto his right pupil. It was previously diagnosed as a chalazion and incision-drainage was performed. The lesion grew back in 6 months’ time and caused mechanical ptosis (2mm). The patient’s acuity was 20/20 in both eyes without aid. The anterior segment examination and fundus examination were quite unremarkable. There were no significant systemic anomalies. Magnetic resonance imaging revealed a soft tissue mass over the right upper lid without any intra orbital extension.

The patient was taken up for full thickness excision of the mass under local anaesthesia. The incision was made through the lid crease. A spindle shaped wedge resection...
with 3 mm margin was performed. The mass was pink in colour, firm in consistency and smooth in appearance. There was no involvement of the conjunctiva below or the upper eyelid margin. Wound was closed in layers with 6-0 Vicryl and 5-0 black silk. A follow up anterior segment analysis revealed no significant findings except a slight swelling of adnexa, which spontaneously resolved on sixth post operative day. Skin sutures were removed on that day. There was a ptosis of 1 mm on that day. On further follow up at one month, the ptosis had resolved completely with residual scarring on the upper eyelid crease (fig. 1B).

Discussion:

Birt–Hogg–Dubé syndrome (BHD) is an inherited disorder characterised by multiple fibrofolliculomas, pulmonary cysts, pneumothorax and renal cysts and tumours\(^2-3\). A fair amount of controversy exists on whether this syndrome should be named after Hornstein and Knickenberg as they had previously studied a woman and her brothers with similar skin lesions and intestinal polyps\(^{11}\). They has labelled it a cutaneo-intestinal syndrome \textit{sui generis}\(^{12}\). The most serious manifestation of BHD remains renal cell carcinoma (RCC) owing to its poor prognosis. RCC generally occurs in adult members of a BHD-affected family and rarely affects infants and teenagers. Skin tumours also develop around the age of 30 years, whereas RCC tends to affect middle-aged and elderly individuals with the syndrome. Pulmonary cysts and pneumothorax are found occasionally in young adult patients aged 20–30 years\(^{13}\).

Fibrofolliculoma demonstrates cords and strands of 2–4 cell epithelium emanating from a follicular structure. The epithelial elements may anastomose and sebaceous elements may be present. Interestingly though, reported cases of solitary fibrofolliculomas did not have any other skin lesions and were of early onset as opposed to multiple fibrofolliculomas\(^4,5\).

In recent times, dermatopathologists have concluded that the fibrofolliculomas, perifollicular fibromas, trichodiscomas and acrochordons seen in patients with BHD represent a spectrum of the same skin tumour. The spectrum of benign follicular neoplasms, fibrofolliculoma appears to be intermediate between perifollicular fibroma (purely of mesodermal origin) and trichofolliculoma (purely of epithelial origin). This creates some difficulty in determining a fibrofolliculoma, which is sometimes thought to be indistinguishable from these. However, a fibrofolliculoma frequently has visible central hair, keratotic plug, dell, or umbilication, whereas trichofolliculoma has features of telangiectasia and peripheral location of hair that may help distinguish it from fibrofolliculoma\(^{14}\). Microscopically, it shows a well-circumscribed dermal tumour with a central well-formed dilated hair follicle. The neoplastic epithelial stroma was arranged separately from the surrounding dermis by intervening clefts.

The patient was referred to a pulmonologist and an urologist to rule out involvement of the lungs or kidneys. There was no evidence of any renal or pulmonary involvement.

The histopathological report revealed a characteristic appearance of a dilated central follicular infundibulum with keratin within (fig. 2). There was perifollicular fibrosis. The fibrotic stroma around the infundibulum had basaloid cells arranged in a radiating pattern (fig. 3). Plenty of large fibroblasts were present in the fibrotic stroma. The fibrotic

\begin{figure}[h]
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\includegraphics[width=\textwidth]{figure2.png}
\caption{Figure 2: High power magnification photomicrograph of dilated infundibulum with lamellae of keratin inside the shaft (ARROW). (Hemotoxylin & Eosin Stain).}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure3.png}
\caption{Figure 3: Low power magnification image through a section demonstrates perifollicular fibrosis (SOLID ARROW), fibrotic stroma all around the infundibulum with basaloid cells in a radiating pattern (HOLLOW ARROW). (Hemotoxylin & Eosin Stain).}
\end{figure}
cells form thin anastamosing cords that radiate from the central hair follicle. The perifollicular stroma is accentuated and fibrotic. In conclusion it is to be said that many solitary fibrofolliculomas of the eyelid may go undetected after a simple excision.

References:


Cite this article as: