Case Study

Case report on Giant Solitary Trichoepithelioma

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Abstract

Giant solitary trichoepithelioma (GST) is a rare benign skin appendage tumor. An 80-year-old male presented with swelling right elbow, Fine Needle Aspiration Cytology (FNAC) of the swelling was suggestive of adnexal tumor. Histopathological examination of the excised lesion shows features of trichoepithelioma. About 12 cases of giant solitary trichoepithelioma have been reported in literature. The present case is the first one to present as swelling in the elbow. The diagnosis of GST is important because of its rarity, histopathological resemblance to keratotic basal cell carcinoma, recurrence and possibility of malignant transformation.

Introduction:

Trichoepithelioma is a commonly encountered benign appendageal tumor that shows differentiation towards mature hair structure and probably arises from pluripotential cell. Trichoepitheliomas can present as multiple trichoepitheliomas, solitary trichoepithelioma, Giant solitary trichoepithelioma & Desmoplastic trichoepithelioma. The common areas to be affected are face, head and neck, with rare occurrence at the perianal region. Genetic abnormality located on band 9p21 may result in either Brooke-Spiegler syndrome, familial cylindromatosis or multiple familial trichoepithelioma.

Case Report

A 80 year old male presented to the OPD of Government Vellore Medical College Hospital with a swelling on right elbow for around 50 years duration. Swelling is insidious onset, slow growing. Case was referred to Department of Pathology for FNAC (Fine Needle Aspiration Cytology). Aspiration
from the swelling yielded a gray white and blood mixed aspirate. Examination of the hematoxylin & eosin (H & E) stained smears show uniform basaloid cells with scant cytoplasm and darkly stained nucleus arranged in sheets and nests, in a background of spindle shaped cells admixed with blood cells. Based on these findings a cytological diagnosis of benign appendageal tumor was offered.

Wide local excision of the tumor was done and presented to Dept. of Pathology for histopathological examination.

Grossly, external surface of the specimen showed skin covered nodule of size 2.5 x 2.5cms. Cut surface showed a well circumscribed gray white tumor just beneath the skin of size 2.8 x 2.5 x 2.5 cms (Fig.1), with deep cut margin and cut margins all around are free of tumor.

Histopathological examination of paraffin embedded, hematoxylin and eosin stained multiple sections studied show attenuated epidermis with an underlying dermal tumor composed of basaloid cells arranged in solid aggregates and lace like pattern. The tumor cells are basophilic with round to oval hyperchromatic nuclei and scant cytoplasm. The tumor islands show peripheral palisading of cells surrounded by a fibroblast rich stroma (Fig.2). The stromal fibroblast tightly encircle the basaloid islands and lack the retraction artefact typical of basal cell carcinoma. Horn cyst in between showed trichilemmal type of abrupt keratinisation (Fig.3).

Discussion
Trichoepithelioma also known as Brooke’s tumor/ Epithelioma adenoids cysticum is a commonly encountered entity midway between trichofolliculoma and keratotic basal cell carcinoma in its degree of differentiation towards mature hair structure. It probably arises from a pluripotential cell. Trichoepitheliomas occur as multiple skin coloured papules of size 2-4 mm over the nasal cleft. Solitary lesions occur as subcutaneous nodules. Giant solitary trichoepithelioma of size >2cm occurs rarely. Mostly giant solitary trichoepithelioma occur around the perianal region. This case is presented for its rarity and location(Table.1).
Table 1: Previous case reports of Giant Solitary Trichoepithelioma:

<table>
<thead>
<tr>
<th>S.N</th>
<th>Age in Years</th>
<th>Sex</th>
<th>Site</th>
<th>Duration (Years)</th>
<th>Tumor size (cms)</th>
<th>Recurrence / Follow up.</th>
<th>Reported by</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>58</td>
<td>M</td>
<td>R Thigh</td>
<td>20</td>
<td>8</td>
<td>None/?</td>
<td>Czernobilsky et al. 1972</td>
</tr>
<tr>
<td>2.</td>
<td>70</td>
<td>M</td>
<td>Nose</td>
<td>?</td>
<td>2.5x1.5</td>
<td>None/1Yr</td>
<td>Dvir E et al. 1981</td>
</tr>
<tr>
<td>3.</td>
<td>53</td>
<td>M</td>
<td>R Thigh</td>
<td>3.5</td>
<td>6.5x4.5x3</td>
<td>None/9months.</td>
<td>Filo GB et al. 1984</td>
</tr>
<tr>
<td>4.</td>
<td>77</td>
<td>F</td>
<td>Natal cleft</td>
<td>7</td>
<td>3.5x3.5x2.5</td>
<td>None/18months.</td>
<td>Tatnall FM et al. 1986</td>
</tr>
<tr>
<td>5.</td>
<td>71</td>
<td>M</td>
<td>Buttock</td>
<td>Many</td>
<td>5x3.5x2.5</td>
<td>None/1 year</td>
<td>Tatnall FM et al. 1986</td>
</tr>
<tr>
<td>6.</td>
<td>70</td>
<td>F</td>
<td>Natal cleft</td>
<td>10</td>
<td>3.5x2.5x2</td>
<td>None/6months.</td>
<td>Tatnall FM et al. 1986</td>
</tr>
<tr>
<td>7.</td>
<td>31</td>
<td>M</td>
<td>Scrotum</td>
<td>-</td>
<td>2</td>
<td>Recurrence/17yrs</td>
<td>Beck S et al. 1988</td>
</tr>
<tr>
<td>8.</td>
<td>-</td>
<td>-</td>
<td>Scar</td>
<td>0.5</td>
<td>3</td>
<td>None/?</td>
<td>Beck S et al. 1988</td>
</tr>
<tr>
<td>9.</td>
<td>67</td>
<td>F</td>
<td>Abdomen</td>
<td>15-20</td>
<td>17x8</td>
<td>None/?</td>
<td>Oursin C et al. 1991</td>
</tr>
<tr>
<td>10.</td>
<td>48</td>
<td>M</td>
<td>L shoulder</td>
<td>?</td>
<td>4x2x1</td>
<td>None/3.5 Yrs</td>
<td>Jemec C et al. 1999</td>
</tr>
<tr>
<td>11.</td>
<td>80</td>
<td>M</td>
<td>R side of nose</td>
<td>1</td>
<td>3x2</td>
<td>None/?</td>
<td>Krishnamurthy J et al. 2010</td>
</tr>
<tr>
<td>12.</td>
<td>45</td>
<td>F</td>
<td>R Forearm</td>
<td>25</td>
<td>9.5x4x2.5</td>
<td>None/6months</td>
<td>Goyal et al. 2012</td>
</tr>
<tr>
<td>13.</td>
<td>80</td>
<td>M</td>
<td>R Elbow</td>
<td>50</td>
<td>2.8x2.5x2.5</td>
<td>?/?</td>
<td>Present case</td>
</tr>
</tbody>
</table>

Giant solitary trichoepithelioma needs to be differentiated from keratotic basal cell carcinoma.

Trichoepitheliomas present as dermal tumor composed of basophilic cells that have the same appearance as the cells in basal cell carcinoma, except that they tend to lack high grade atypia and mitosis. Horn cyst shows abrupt keratinisation called “trichilemmal”keratinisation. The tumor islands composed of basophilic cells that are arranged in lace like or adenoid and in solid aggregates. These tumor islands show peripheral palisading of their cells surrounded by dense fibroelastic stroma without retraction artefact typical of basal cell carcinoma. Both adenoid and solid aggregates show invaginations, which contain numerous fibroblasts and resemble follicular papillae, also known as papillary bodies.

Basal cell carcinomas show predominant basal cell type, peripheral palisading of lesional cell nuclei, specialized stroma and clefting artefact between the epithelium and the stroma. In more than 90% of basal cell carcinomas, a connection between...
tumor cell formations and the surface epidermis can be shown to exist\textsuperscript{15}. Basal cell carcinomas showing differentiation toward hair structures are called keratotic basal cell carcinoma. Keratotic basal cell carcinoma shows parakeratotic cells and horn cysts in addition to undifferentiated cells. The horn cysts, which are composed of fully keratinized cells, represent attempts at hair shaft formation\textsuperscript{16}.

**Conclusion:**
Trichoepitheliomas shares with basal cell carcinoma the presence of horn cysts, and it is sometimes difficult to decide whether a lesion represents keratotic basal cell carcinoma or a trichoepithelioma. In such a situation, clinical data may be necessary to reach a diagnosis. As trichoepitheliomas are benign lesions, surgical excision is curative and hence it needs to be differentiated from malignant keratotic basal cell carcinoma. Close follow up of the cases of giant solitary trichoepithelioma is required as there is possibility of recurrence\textsuperscript{9} and rarely malignant transformation.

**Figures:**

![Image](image_url)

**Fig.1:** Cut surface shows a gray white tumor of size 2.8x2.5x2.5cm beneath the skin.
**Fig. 2:** Low power showing - Islands and nests of uniform basaloid cells with Peripheral palisading of tumor cells surrounded by dense fibroblastic stroma.

**Fig. 3:** Low power showing - Horn cyst with abrupt trichilemmal type keratinisation.
References: