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Case Report

Giant solitary trichoepithelioma: A rare trichogenic tumor

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ABSTRACT

Giant solitary trichoepithelioma (GST) is a rare benign skin adnexal tumor which arises from hair follicle. It represents more mature end of the spectrum of trichoblastoma, and a unique type of trichoepithelioma that is most common in elderly but can be present at any age. Only few cases are reported worldwide and some of which have showed malignant transformation in to basal cell carcinoma. This case is presented due to its scarce presentation over neck region and difficulty encountered in diagnosis of the case.

1. Introduction

Trichoepithelioma originates from benign proliferation of undifferentiated pluripotent cells of the follicular-sebaceous-apocrine unit.[1] Most of the adnexal tumors are of dermal or subcutaneous origin but epidermal connection has also been seen in some cases.[2] Different variants of trichoepithelioma include: Multiple Trichoepithelioma, Solitary, Giant solitary and Desmoplastic; out of which multiple trichoepitheliomas are most common. Histopathologically, these are divided into basaloid, clear or eosinophilic and tumors with glandular differentiation; although histopathologically all variants are similar but not identical.[3] Histopathology plays important role in diagnosis; to differentiate it from other tumors with same clinical presentation and to rule out malignant transformation. Only countable cases are reported in the world till date.[2–5]

2. Case Report

A forty-nine years old female presented with slow growing painless mass over right side of neck for six years. Initially lesion was pea sized, then gradually increase in size over a period of six years to tennis ball sized. Patient took multiple treatments from private practitioners for the same in the form of oral/topical antibiotics, various topical steroid combinations but lesion didn't show any improvement. Patient had no history of trauma preceding the lesion, no past and family history of tuberculosis and had no other comorbidities.

On examination Single, well defined hyperpigmented verrucous plaque with multiple studded skin colored papular lesions with two brownish crusted lesions with atrophic lesions in between measuring 5×3 cm^2 in size over right submandibular region (2 cm away from midline, 1.5 cm below mandibular margin and 4 cm above clavicle), with rim of hyperpigmentation and atrophy of surrounding skin. It was firm in consistency, fixed to the overlying skin but not to the underlying structures. It was non tender and didn't bleed on touch. No associated lymphadenopathy. [Figure 1]

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2.1. Differential diagnosis considered

Lupus vulgaris, Trichoblastoma, Basal cell carcinoma, Microcytic adnexal carcinoma, Sarcoidosis.

Mantoux test, chest x-ray, 2 sputum Acid fast bacilli test was done to rule out tuberculosis as lesion was mimicking lupus vulgaris but all investigations came out to be normal. Ultrasonography of local part showed minimal vascularity with few hypoechoic areas. Local part x-ray was normal. All routine investigations done including complete blood count, liver function test, Renal function test before taking lesional fine needle aspiration cytology (FNAC); were within normal limits. FNAC report showed basaloid cells arranged in nests with scanty cytoplasm and dark stained nuclei and papillary mesenchymal bodies which was suggestive of skin adnexal tumor. Punch biopsy report showed dermal tumor with nests of dark stained basaloid cells with abrupt keratinization at places-horn cyst [Figure 2] with trilamellar differentiation and differentiation towards hair structure suggestive of trichoepithelioma [Figure-3]. Despite immunohistochemical tests being confirmatory, their use in our case was limited due to unaffordability of patient.

Wide local excision with 5mm safe margin was done with flap surgery by General Surgery department. Patient was followed for 1 year without any evidence of recurrence or malignancy.

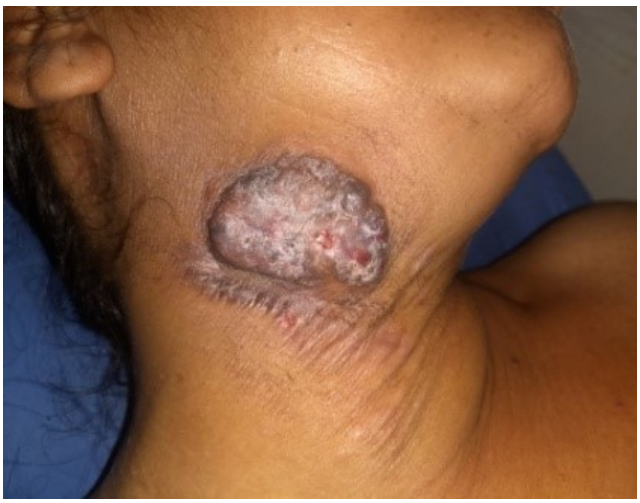

Figure 1: Single, well defined verrucous plaque over right submandibular region, atrophy of surrounding skin and firm in consistency, fixed to skin

3. Discussion

Trichoepithelioma was first described by Brook in 1892 as "Epithelioma Adenoides Cysticum",so it is also known as Brooke's tumor.[2]It is a hamartoma of hair germ cells and genodermatosis.[6]

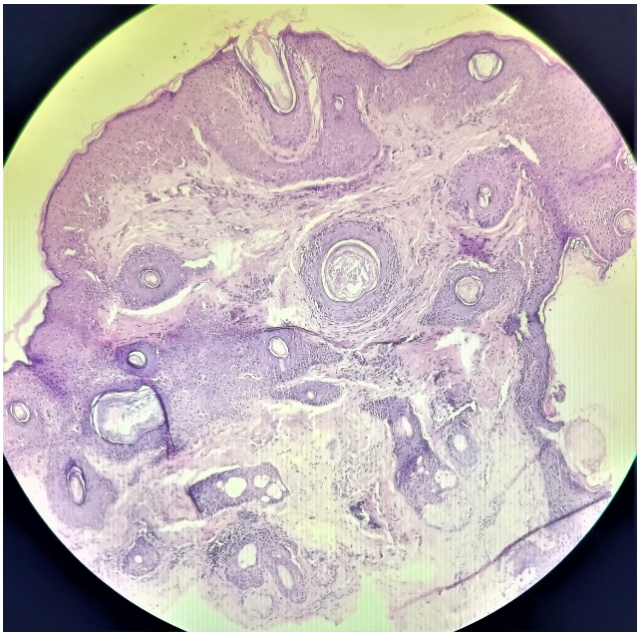

Figure 2: Dense cellular stroma and focal stromal cracking (between collagen bundles) with keratin horn cysts surrounded by nests of basaloid cells and abortive hair follicle (H&E, 10×)

Giant solitary trichoepithelioma is a rare form of solitary Trichoepithelioma measuring more than 2 cm in diameter. It is more common in older age (> 60 years) but can be present at any age with male predominance. In most of the case reports of GST, most common sites involved are perianal, groin and thighs.[4] In our case lesion was on right submandibular region, in a female patient and no case report has been reported describing giant solitary trichoepithelioma at this unusual site.

Giant melano-trichoblastoma is a variant of giant solitary trichoepithelioma, demonstrating pigmented colonization by dendritic melanocytes.[7]

Multiple variant is more common than the solitary variant.[8] Multiple trichoepitheliomas are more common in young females involving head and neck region. It may be associated with Spiegler-Brookler syndrome and Brooke-Fordyce syndrome.[7]

An inheritance pattern is not seen in solitary trichoepithelioma and is usually <2 cm in diameter, smooth, firm, nonulcerated nodule. It occurs most commonly on face in middle-aged person.[7]

A report of simultaneous presentation of multiple trichoepithelioma and giant solitary trichoepithelioma with malignant change has also been reported.[1]

Changes of basal cell carcinoma has been noted in few case reports[5,6] and it can only be differentiated based on biopsy and immunohistochemical markers. Immunohistochemical staining is crucial in differentiating basal cell carcinoma from Giant solitary trichoepithelioma;

CD34, CD10, PLHDA 1 positivity and p75NTR negativity[4] of the stromal cells around the nest suggests trichoepithelioma but negative in basal cell carcinoma which is an important differentiating feature. Histopathologically, trichoepithelioma shows papillary-mesenchymal bodies, granulomas, and calcification, while basal cell carcinoma is more likely to show inflammation, retraction, a higher frequency of apoptotic cells, and mitotic figures and trichilemmal keratinization.[4,9]

Surgical excision with 5 mm margin with or without flap is the treatment of choice. Other treatment option includes radio surgical ablation, cryotherapy, dermabrasion and curettage. Adjuvant radiotherapy is indicated in cases of malignant transformation to BCC after surgical excision. Although recurrences are rare, all such patient should be kept on a yearly follow-up.

4. Conclusion

Though Giant solitary trichoepithelioma is a rare appendageal tumor, it is of a great concern because of potency of local recurrence, conversion into basal cell carcinoma. It may mimic other cutaneous disorders leading to confusion in clinical diagnosis and may be the reason for the scarce case reports.

5. Source of Funding

None.

6. Conflict of Interest

None.

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