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Multiple Trichoepitheliomas of Pinna of Ear- A rare case

KEYWORDS: Trichoepithelioma, Ear, Pinna

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ABSTRACT *Trichoepitheliomas are benign hamartomatous cutaneous neoplasms that mostly occur as multiple lesions that are dominantly inherited. They usually appear in early childhood or in young age. In 1892, Brooke originally described it as epithelioma adenoids cysticum. They usually occur as multiple papulonodular lesions on face or upper trunk. It is a rare neoplasm and there is no case reported of trichoepitheliomas involving ear pinna in the literature we surveyed. We report rare case of multiple trichoepitheliomas involving ear pinna.*

Introduction-
Trichoepithelioma is a benign cutaneous neoplasms of pilosebaceous apparatus occurring mostly on face. They occur as nonhereditary solitary lesions or autosomal dominant multiple lesions. They can occur in any part of body but mostly involves face of an adult. The diameter is usually 2-7 cms. They can produce significant cosmetic disfigurement. Their association with basal cell carcinoma (BCC) is well documented. We present a case of multiple trichoepitheliomas of ear pinna which is a rare location.

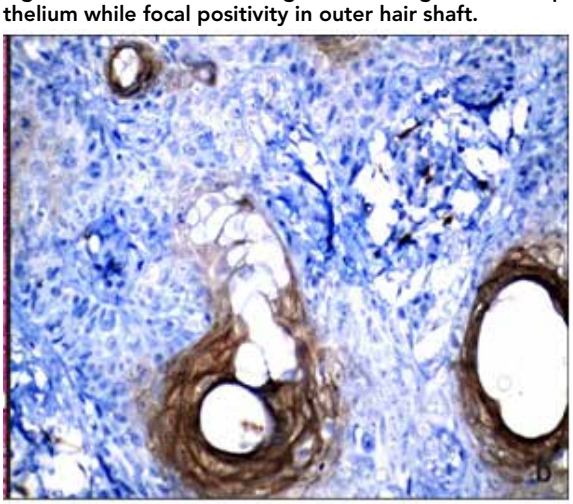
Case report-
20 year old male patient came with the complaints of slowly growing lesions on pinna of the ear since past 2 years. On examination, there were soft whitish brown nodules measuring about 0.5 to 1 cm in diameter on the pinna of the left ear. The centers were slightly depressed. The clinical diagnosis of pinna granuloma was made and biopsy was taken and sent to the pathology department.

On gross examination, there were 2 nodules which measured 0.5 cm and 0.7 cm in diameter. They were shiny, brownish coloured nodules. Microscopic examination showed well circumscribed two nodular lesions overlined by epidermis with surface keratosis. From the basal cell layer there were down growing cords and strands of the cells forming islands of basaloid cells. The basaloid cells were oval with vesicular or slightly hyperchromatic nuclei. In the center horn cysts with abrupt keratinisation reminiscent of hair shaft formation were noted. The stroma in between was fibroblastic and showed infiltration by plasma cells and few lymphocytes. A focus of giant cell reaction was found which was due to reaction to the ruptured horn cyst.(fig.1.)

Fig.1a. Downgrowing cords and strands of basal cells forming islands. In the center horn cysts with abrupt keratinisation reminisant of hair shaft formation were noted.

Immunohistochemical staining by calretinin showed negative staining in tumor cells with focal staining in outer hair shaft. (fig.2)

Fig.1b. Calretinin Stain- Negative staining in tumor epithelium while focal positivity in outer hair shaft.



Discussion-
Multiple trichoepitheliomas (MTs) usually appear in early childhood or young adults. They gradually increase in size and number sometimes leading to significant cosmetic disfigurement. They are located mainly on face but also can be seen on the scalp, neck and upper trunk.

Multiple trichoepitheliomas are transmitted as an autosomal dominant trait.[1]The gene for MTs has been mapped to a locus on chromosome 9p21[2].

They appear as multiple skin coloured papules or nodules and usually are firm, rounded, translucent, shiny and well demarcated. The center may be umbilicated.

Transformation to BCC is rare, though its association with BCC is reported.[3] Some authors consider it as an instance of nevoid basal cell carcinoma syndrome.[4]

The differential diagnosis includes keratotic BCC, syringoma and other appendegeal tumors. Diagnosis is based on history, clinical data such as numbers and distribution, family history and skin biopsy.

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In one study immunohistochemical marker CD34 was found to be useful in differentiating BCC from MT. In trichoepitheliomas focal positive CD34 staining was found in fibroblastic stroma, however in BCC this pattern was not seen.[5]

In the study done by E.Gonzalez-Guerra et al, they analyzed nine trichoepitheliomas and found that most of the tumor epithelium was negative for calretinin, with limited focal staining.[6] In our case also tumor epithelium was negative for calretinin with focal staining.

Although trichoepithelioma shows solid strands of basophilic epithelial cells and horn cysts like syringoma it lacks ductal structures.[7]

Treatment is usually surgical excision, cryotherapy or dermabrasion. The important side effect of all these treatments is scarring. Recurrences are common. A long term follow up is necessary to rule out BCC development.

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