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Clinical Challenge

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Multiple Firm Mobile Swellings Over the Scalp

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INTRODUCTION

A 30-year-old female presented with multiple, asymptomatic swellings over the scalp [Figure 1] since the past 1 year. Initially, patient noticed a small pea-sized swelling over the vertex, which gradually increased in size and similar lesions developed over different areas of the scalp. The lesions were mostly asymptomatic except for occasional pain due to trauma induced by combing. There was no history of preceding trauma and none of the other family members had a similar complaint. Clinical examination revealed eight firm, well-defined swellings located discretely over the vertex, parietal, and occipital regions of the scalp. They were freely mobile, non-tender with normal overlying skin and hair. Excision biopsy of one of the lesions was performed under local anesthesia and histopathological examination revealed a cystic lesion with central eosinophilic homogenous mass lined by an epithelium [Figure 2]. The lining epithelium showed 2-3-layered cells which were flatter and more eosinophilic at the base and, larger and paler toward the cyst cavity [Figure 3].

WHAT IS YOUR DIAGNOSIS?

Diagnosis

Trichilemmal cysts (pilar cysts)

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DISCUSSION

Trichilemmal cysts are benign cystic lesions that are filled with keratinous material and lined by epithelium derived from outer root sheath (ORS).[1,2] They affect 5 to 10% of the population with a female preponderance.[3] Familial cases with an autosomal dominant mode of inheritance have been seen with linkages to short arm of chromosome 3.[4,5] They have quite a resemblance to epidermoid cysts. However, these lesions are less common than epidermoid cysts and differ in their location. Trichilemmal cysts arise from the epithelium of follicular isthmus, where the inner root sheath is shed off and the ORS (trichilemma) undergoes a specific form of keratinization (trichilemmal keratinization) forming a cyst wall without a granular layer.[3] Ninety percent of the trichilemmal cysts occur over the scalp and rarely the face, trunk, and extremities may be involved. Majority of the patients have multiple cysts. Solitary lesions are seen only in 30% of cases.[6] They present as slow-growing, freely mobile, firm, non-tender nodules scattered over the scalp. Unlike epidermoid cysts, no punctum is seen. They may become inflamed and tender following trauma. Rapid growth and ulceration indicate malignant degeneration which although, is rare. Histopathologically, the lesion is comprised of a cyst wall containing homogenously eosinophilic material composed of keratin and cellular debris. Intervening basophilic areas may be seen in older lesions indicating calcification. Cholesterol clefts are often present. The hallmark diagnostic feature is in the lining epithelium. The cells of the lining epithelium gradually increase in size as they approach the cyst cavity appearing swollen with pale cytoplasm. At this point, they abruptly keratinize without forming a granular layer. Some lesions may show focal changes of an epidermoid cyst, with formation of a granular cell layer, known as hybrid cysts. Immunohistochemically, the central keratinous mass stain for anti-keratin antibodies

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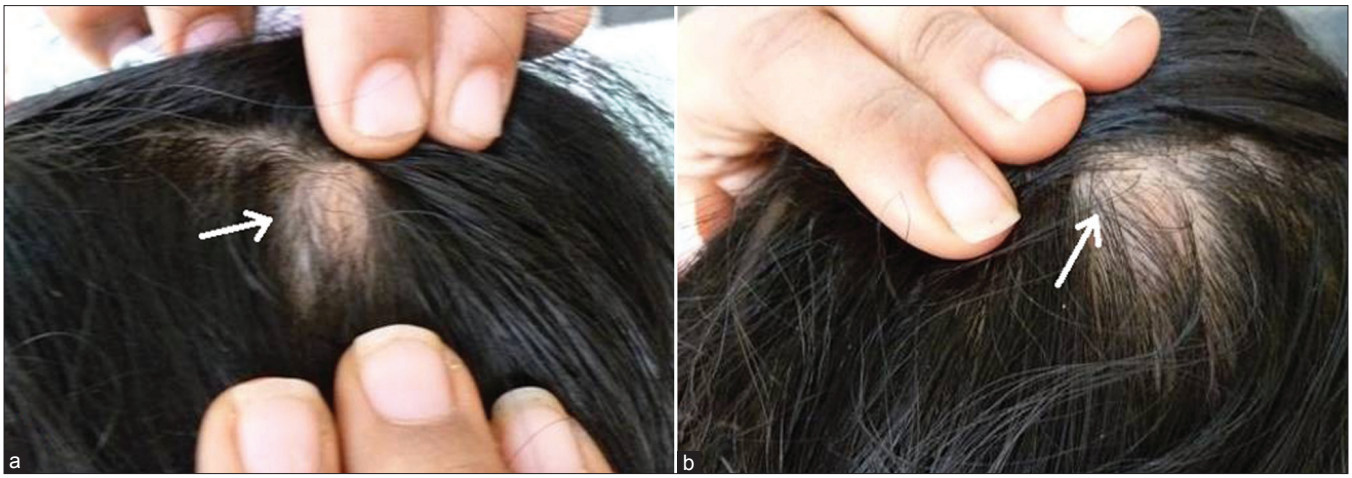

Figure 1: Firm mobile nodules (arrows pointing) over vertex (a) and parietal (b) regions

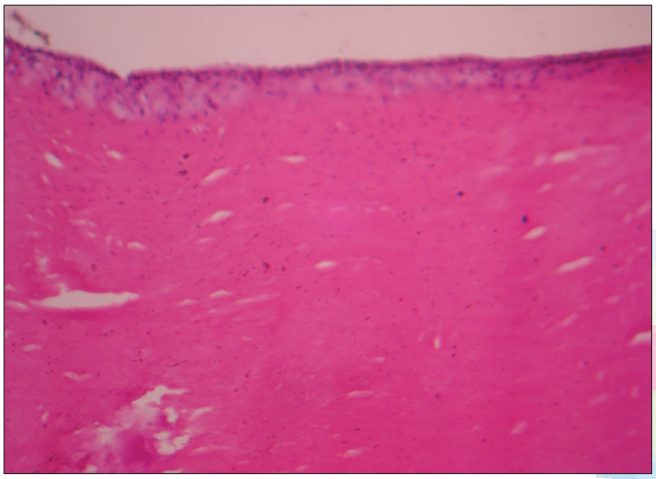

Figure 2: Central homogenous mass with lining epithelium (H and E, 10×)

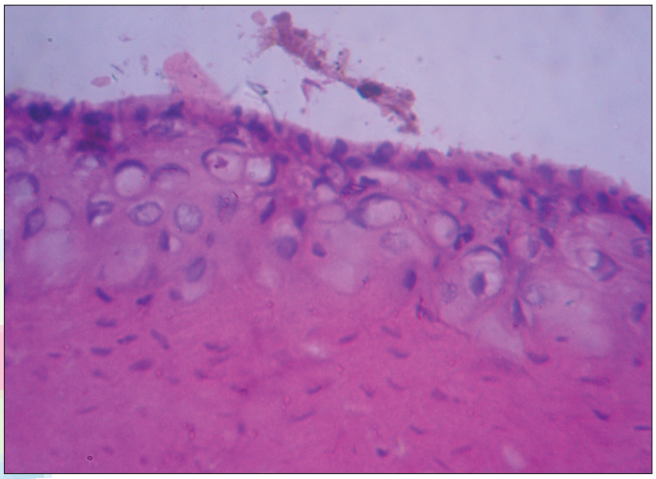

Figure 3: Lining epithelium with smaller, deeply staining cells at the base and larger, paler cells nearing the cyst cavity (H and E, 40×)

derived from human hair, in contrast to epidermoid cysts, which stain with anti-keratin antibodies obtained from human callus. Electron microscopy of the lining of trichilemmal cysts shows that on their way from the periphery toward the cavity, the epithelial cells have an increasing number of intracellular filaments. There is an abrupt transition from nucleate to anucleate cells with the loss of all cytoplasmic organelles. The junction between the keratinizing and keratinized cells shows interdigitations. The keratinized cells are filled with tonofilaments and, unlike those in epidermoid cysts, retain their desmosomal connections.[7] Simple excision is the treatment of choice. As the cysts are firmer than epidermoid cysts, the lesions can be enucleated with smaller incisions.[3] Excised lesions may recur.[6]

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