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A rare presentation of proliferative trichilemmal cyst in an ear lobule Ruta Shanmugam, VellaveduUmapathy Shanmugam, Rajagopalan Mariappan, Balaji Swaminathan, Srikanth Nandipatti, P.Viswanathan, Amrutha Sathappan

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Abstract: Trichilemmal cysts, also called pilar cysts, are known skin lesions arising from the outer root sheath of the hair follicle. Proliferating trichilemmal cysts or pilar tumours, thought to arise from these lesions, are much less common. They are generally present on the Scalp. Other locations, although much less common, have been reported. To our best knowledge proliferating trichilemmal cysts involving a non hairy area like an ear lobule have not been reported so far. Hence we report this rare case of a cyst involving the ear lobule for which excision biopsy was performed and histopathological findings were consistent with those of proliferative trichilemmal cyst. Hence it should be considered as a differential diagnosis in case of swelling in a non hairy region.

Keywords: Ear lobule, non hairy region, nodular swelling, Proliferative trichilemmal Cyst

INTRODUCTION

A proliferating trichilemmal cyst, also known as proliferating pilar tumour of the scalp, is a rare benign neoplasm arising from the isthmus region of the outer root sheath of the hair follicle. Most proliferating trichilemmalcysts arise from within a pre existing trichilemmal or pilar cyst. While trichilemmal cysts are common intradermal or subcutaneous cysts occurring in 5-10% of the population, 2% of these cases will develop into proliferating trichilemmal cyst. They may be inherited in an autosomal-dominant mode, linked to chromosome 3. They are generally present on the Scalp. Other locations, although much less common, have been reported and these include the neck, trunk, groin, mons pubis, vulva, and gluteal region; the upper and lower extremities, including the elbow, the dorsum of the hand, and the index finger; the face, including the forehead. nose, evelid, lip, and intraorally. Trichilemmal cyst involving a non hairy area like an ear lobule has not been reported so far.

CASE REPORT

A 46-year-old female presented to our ENT outpatient department with a history of gradually progressive, painless swelling on the right ear lobule involving both the lateral and medial surface of 2 years duration. It was not associated with redness, discharge or pain. There was no history of trauma or history of any surgery performed in the past. There was no significant family history. On clinical examination, there was a single spherical firm nodular mass with well defined margins occupying the right ear lobule measuring 2 cm \times 1 cm, non tender, skin overlying the swelling was not pinchable and fixed to the underlying tissue. Examination of the rest of the ear and tympanic membrane was normal. The left ear was normal. A clinical diagnosis of sebaceous cyst was made and excision biopsy was performed under local anaesthesia. The diagnosis of proliferative trichilemmal cyst was a histopathological surprise. FIG (1 and 2) Nodular mass at the medial and lateral aspect of ear lobule.

HISTOPATHOLOGY

Low power view of tumor in dermis (H&E $\times 100$) shows well-circumscribed lesions with islands of squamous epithelium displaying trichilemmalkeratinization. (Figure 6 & 7)

Scanning power view shows a epithelial lined cyst filled with brightly eosinophilickeratinaceous debris (Figure 8). Focal rupture of the cyst with an associated giant cell reaction (Figure 8). Closer inspection identifiestrichilemmal differentiation(Figure 9).

Follow up of the patient after five months revealed no recurrence.(fig 10 & 11)





Fig-3

Fig-4



Fig-5

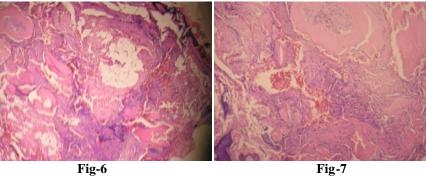


Fig-7

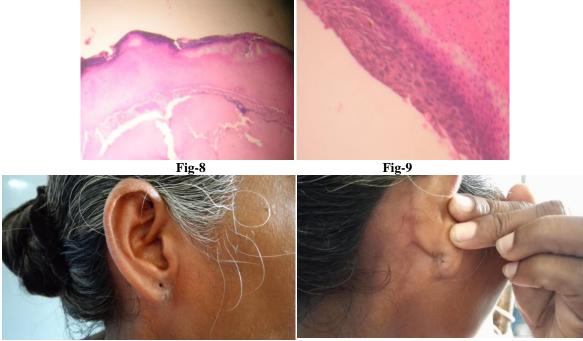


Fig-10

DISCUSSION

Trichilemmal cysts originate from the external hair sheath and may lead to so-called proliferative cysts, initially called proliferating epidermoidcysts [1]. It is also known as proliferating trichilemmaltumoror pilar tumour of the scalp. Ninety percent of Proliferative type are characterized by trichilemmalkeratinization, i.e., the sudden change from nucleated epithelial cells to anucleated cells in the absence of a granular layer [1,2]. These masses are localized in dermis or subcutaneous tissue, may become exophytic, sometimes exhibits ulceration, and are solid or partially cystic [3]. They are more common in women over the age of 60 years, and in 90% of the cases they occur as a solitary lesion on the scalp[1,4]. These tumours may also affect the neck, trunk, groin, pubis, vulva, upper and lower extremities, shoulder, back of the hand, index finger, face, forehead, nose, eyelids, lips, oral cavity, jaw skull base, and gluteal region[2,4]. The Proliferating trichilemmal tumours generally have a benign clinical course, and a histological differentiation from squamous cell carcinoma is often difficult. Trauma or infection is believed to stimulate a malignant transformation[1]. Trichilemmal cyst formation, trichilemmal type hyaline keratinization, eosinophilic membrane, calcification are features favouring the diagnosis of benign lesion.Sudden rapid growth, severe nuclear atypia, marked cellular pleomorphism with atypical mitoses, dyskeratotic cells, and infiltrating margins are the diagnosis of necessary for malignant transformation[5,6]. The differential diagnosis should include epidermoid cyst, keratoacanthoma, squamous cell carcinoma, pilomatrixoma, sweat gland tumour, cylindroma, basal carcinoma, cell and

Fig-11

angiosarcoma[1,2]. Trichilemmal cysts comprise about 20% of all cysts, the remaining are epidermoid cysts. If these unfavourable probabilities are kept in view; as far as we are concerned, wide surgical excision with long-term surveillance may be the best choice for both diagnosis and treatment still today. However to the best of our knowledge, no cases of trichilemmal cysts involving the ear lobule have been reported. This case report highlights the need for considering trichilemmal cyst as differential diagnosis of the swelling in a non hairy area (ear lobule).

CONCLUSION

We present this case to highlight the need for considering proliferative trichilemmal cyst as a differential diagnosis of swelling in a non hairy area (ear lobule) and to emphasize the need for wide excision biopsy and regular follow up in such cases.

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