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Case Report

Pathology Section

Folliculosebaceous Cystic Hamartoma: A Rare Entity

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ABSTRACT

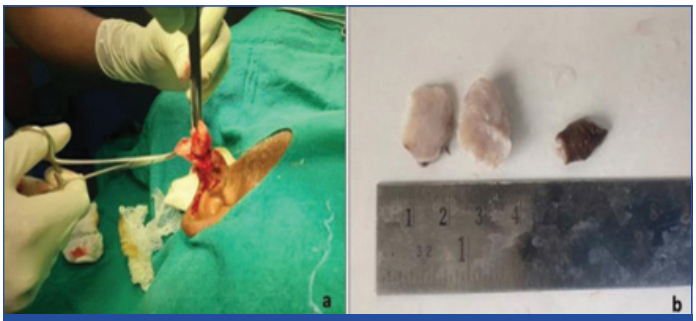
Folliculosebaceous Cystic Hamartoma (FSCH) is an uncommon cutaneous hamartomatous lesion. It is basically a tumour like malformations exhibiting abnormal overgrowth of biphasic elements i.e., epithelial and mesenchymal components that are normally found in the skin. Few other hamartomas of pilosebaceous origin are trichofolliculoma, trichodiscoma, fibrofolliculoma and pilar sheath acanthoma. Though distinct, this entity shares similar histological features to Sebaceous Trichofolliculoma (STF). Hence, a proper histological examination is must in differentiating it from STF and other clinical and histological entities. Authors hereby, describes a case of FSCH in a 37-year-old male who presented with a slow growing cystic mass on his left earlobe, an uncommon location for such lesion. The mass was excised, and a detailed histological evaluation showed a characteristic multinodular growth pattern with lobules of pilosebaceous glands forming nodules around cystically dilated follicular infundibular structures. No hair shafts were seen in the cystic cavity ruling out differential diagnosis of sebaceous hyperplasia which could have been a close mimic. The intervening stroma revealed dense collagen, sheets and lobules of adipocytes, many vascular channels filled with fibrinous deposits. Perivascular adipocytes were also noted along with few nerve tissues (perineurium). A detailed clinicopathological work-up helped in arriving at a final diagnosis of FSCH with neural component. The patient was doing well till last six months of follow-up. No recurrence of similar lesion was noted in the patient.

Keywords: Hamartomatous lesion, Histopathology, Pilosebaceous unit, Sebaceous trichofolliculoma

CASE REPORT

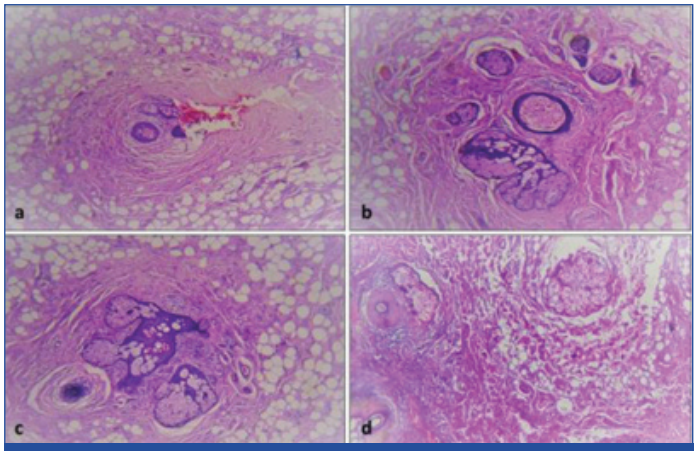
A 37-year-old male patient presented with a slowly progressive nodular swelling in his left earlobe for last five years. The lesion was solitary, measuring approximately 3×2 cm in size and was non tender, with no local rise in temperature. No similar swelling was noted elsewhere in the body of the patient. There was no history of tuberculosis, diabetes mellitus, hypertension or any other systemic illness in the past. Based on clinical examination, differential diagnosis considered were lipoma, sebaceous cyst, neurofibroma, etc.

His routine laboratory investigations were within normal limits. The patient was admitted under Ear, Nose, Throat (ENT) Department and underwent excision of the nodule [Table/Fig-1a]. The excised specimen was sent for Histopathological Examination (HPE). On gross examination, the received specimen was a single grey-white nodular soft tissue measuring 3×2 cm along with one flattened skin covered tissue measuring 1.5×1 cm [Table/Fig-1b].

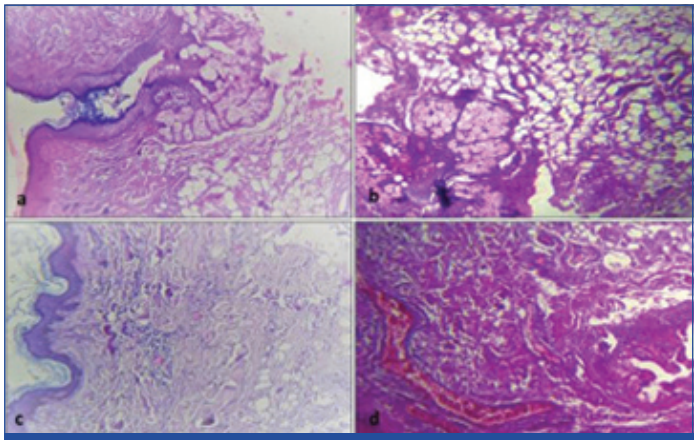


[Table/Fig-1]: a) Clinical image showing excision of single nodular, left lobe swelling, measuring approximately 3×2 cm in size. b) Gross image of single nodular soft tissue measuring 3×2 cm along with a flattened skin covered tissue measuring 1.5×1 cm.

Histopathology shows multiple sections with characteristic multinodular growth pattern with lobules of pilosebaceous glands forming nodules around cystically dilated follicular infundibular structures [Table/Fig-2a-d]. Infundibular type of keratinisation was noted. No hair shafts were seen in the cystic cavity. The intervening stroma revealed dense collagen, sheets and lobules of adipocytes, many vascular channels filled with fibrinous deposits [Table/Fig-3a-d]. Perivascular adipocytes were also noted [Table/Fig-4a]. Few nerve tissues (perineurium) were also identified in the section [Table/Fig-4b].

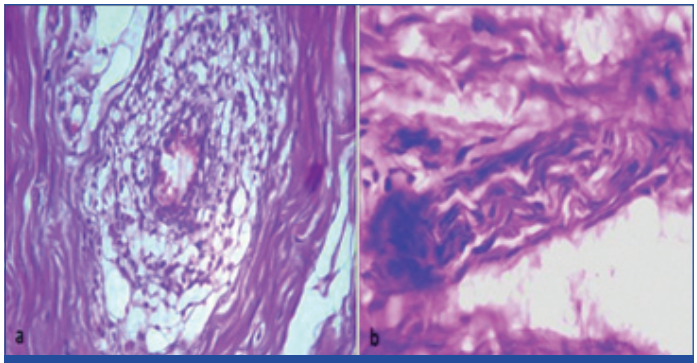


[Table/Fig-2]: a) Section showing a characteristic multinodular growth pattern. b-d) Sections show lobules of pilosebaceous glands forming nodules around cystically dilated follicular infundibular structures (H&E stain 40X).



[Table/Fig-3a-d]: Intervening stroma revealing dense collagen, sheets and lobules of adipocytes, many vascular channels filled with fibrinous deposits (H&E; 20X).

No evidence of primitive hair structures, vellus hair follicles, mammary ducts or sweat glands were seen in the sections examined. Based on histological evaluation, initially a differential diagnosis of sebaceous hyperplasia and FSCH were considered. However, on detailed clinical and histological evaluation (as described in the discussion section



[Table/Fig-4a-b]: Image showing perivascular adipocytes and few nerve tissues (perineurium) (H&E, 40X).

below), a diagnosis of FSCH was made. No complication was noted after excision of the nodule. The patient was doing well till last six months of follow-up and no evidence of recurrence were noted.

DISCUSSION

The FSCH is a relatively rare biphasic hamartomatous adnexal lesion. It is composed of dilated folliculosebaceous units along with prominent mesenchymal elements. FSCH is an uncommon benign lesion of pilosebaceous origin and was first described by Kimura T et al., in 1991 in a lesion located on face [1]. The lesion usually presents as a solitary papule or nodule with face and scalp being a common location.

Various papulonodular or cyst like cutaneous lesions need to be considered in its evaluation due to lack of distinctive clinical features [2-5]. A careful HPE aids in the definitive diagnosis of such hamartomatous lesion. This entity was described for the first time by Kimura T et al., in 1991 with usual location on face [1]. Clinically, it is an asymptomatic, slow growing, small papule or nodule usually noted on the head and neck region. Few cases of FSCH have been reported in uncommon location like upper back, forearm, genital area, scalp [2-6]. Previously, a case of FSCH has been reported on earlobe, similar to the index case; however, the patient was diagnosed with neurofibromatosis type I [4].

The age of the patients with FSCH ranges from 4-84 years, as described in literature [5-9]. The present case was of a 37-year-old male patient. The size of the tumour, as reported in literature, ranges from 0.5-1.5 cm in diameter, but few cases of giant variant of FSCH (with size 7.5 cm and 15 cm in diameter) have also been described in literature. The size of the lesion in present case was 3 cm in diameter.

The term hamartoma as used by Kimura T et al., suggests the fact that a jumble of tissue elements is present at the site of lesion [1]. The tendency of this lesion to increase in size depends on the location like back, labia and extremities and can be sometimes seen as polypoid, plaque like or multinodular growth [5-9]. Though the index case did not show giant variant, but it has increased to about 3 cm in diameter by the time excision was done.

Due to lack of distinctive clinical features for this lesion, various differentials which can be considered clinically are sebaceous cyst, sebaceous hypeplasia, lipoma, intradermal nevus, neurofibroma, etc., [7-9]. Hence, the role of excision biopsy becomes important in making a definitive diagnosis.

The term "FSCH" correlates with the presence of histological features like follicular cystic structures, adjacent multiple sebaceous lobules and prominent mesenchymal changes in the stroma. The diagnosis in the present case was made as FSCH with considering dermoid cyst, steatocystoma, STF as differential diagnosis.

Kimura T et al., have suggested few criteria for diagnosis of FSCH, which are described as: a) an infundibular cystic structure to which sebaceous lobules were attached; b) surrounding bundles of collagen; c) adipocytes; and d) an increased number of small vessels [1,4-9]. The index case showed presence of all above mentioned features. The present case had neural component which makes it even rarer as very few cases of FSCH with neural component have been reported in literature till date [4].

However, the closest differential is STF. Clinically, STF presents as a depressed lesion, whereas FSCH shows a papular or nodular lesion, as in this case. Histologically, STF show hair shafts within follicular structures while FSCH does not. It is stated in the literature that FSCH is a late stage of STF and shows secondary follicular regression. However, few congenital cases reported cannot explain this theory, hence accepting the fact that FSCH is a distinct entity.

Though surgical excision of entire lesion is the management of choice, few case reports have described the successful role of CO_2 laser and acitretin therapy [6-9]. No recurrence after excision of the lesion has been reported in any case published in the literature.

CONCLUSION(S)

The present case is reported as an unusual case of FSCH with distinct histological findings in an adult male. To conclude, as FSCH has no distinctive and specific clinical features, various differentials were being considered. Hence, to achieve at a correct diagnosis in such case, histopathological evaluation of the lesion helps in arriving at a definitive diagnosis of FSCH.

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