Case Report

Folliculosebaceous cystic hamartoma with neurofibroma: A rare association

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ABSTRACT

Folliculosebaceous cystic hamartoma (FSCH) is a rare cutaneous hamartoma composed of follicular, sebaceous, and mesenchymal elements. Especially interesting is its differential with another benign tumor sebaceous trichofolliculoma with which it shares several histologic features. This case report highlights the association of FSCH with neurofibroma and discusses its possible common pathogenesis.

Key words: Folliculosebaceous cystic hamartoma, nestin, neurofibroma

INTRODUCTION

Folliculosebaceous cystic hamartoma (FSCH) was first described by Kimura *et al*. in 1991.[1] FSCH generally occurs on the head and neck region as papule or nodule. Cases have also been reported on other areas such as nipple, genitalia, limbs, and upper back.[2,3] The age of the patients varies from 4 to 84 years The name "FSCH" refers to the main histopathological features of the lesion, which include a follicular cystic structure with multiple attached sebaceous lobules, stromal mesenchymal changes, and increased adipose and vascular tissue.[4] The usual histologic features include a cystic infundibular structure to which several lobules of sebaceous glands are attached along with several small venules, adipocytes, and bundles of collagen. Sebaceous trichofolliculoma (STF), which is a close histologic differential, generally presents as a depressed lesion whereas FSCH presents as a papule or nodule. STF usually has hair shafts within the follicular structures whereas FSCH does not.[1] FSCH does not show secondary follicles usually seen in conventional trichofolliculomas. The stroma of an FSCH contains thin bundles of collagen, adipocytes, spindle cells, an increased number of capillaries and venules, and occasionally, a proliferation of nerve fibers.[1] This mesenchymal component may form a considerable portion of the hamartoma. Such stroma is not found in STFs. Several authors have suggested that FSCH is a result of the secondary follicular regression of STF.[5]

Since they lack distinctive clinical features, they are often misdiagnosed as other lesions such as intradermal nevus, sebaceous hyperplasia, lipoma, and neurofibroma.[6] The differential diagnoses include STF, steatocystoma, and dermoid cyst.

CASE REPORT

A 62-year-old man presented to the dermatology outpatient department with a very slowly enlarging sessile soft papule on the right side of the face. There was no history of pain and no significant medical history. Physical examination revealed a 2.5 cm × 1 cm × 1 cm skin-colored smooth-surfaced papule. The overlying skin was unremarkable. A clinical diagnosis of skin tag (acrochordon) was made. He also had a 1 cm × 1 cm dome-shaped papule on the right side of the neck which was thought to be a dermal melanocytic

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nevus. Both were excised and sent for histopathological examination. The histopathology of the papule on the face showed a dilated follicular cystic structure with infundibular type keratinization and several lobules of sebaceous glands radiating from it [Figure 1]. There were no connections noted between the sebaceous lobules and the overlying epidermis. Lack of multiple hair shafts in the cystic structure, absence of secondary follicle formation, and prominence of stroma rich in fat [Figure 2] all favored a diagnosis of folliculosebaceous cystic hamartoma over STF. The histopathology of the other papule on the neck showed features of a cutaneous neurofibroma [Figure 3].

The patient did not have clinical features suggestive of neurofibromatosis Type I.

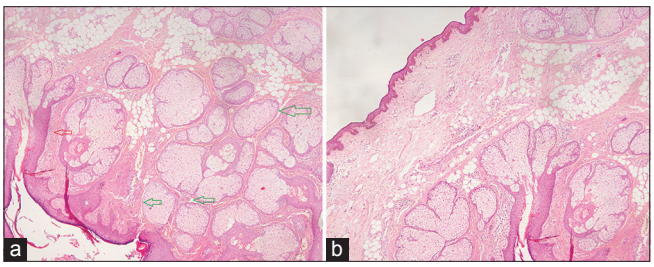

Figure 1: Papule on face: (a) An infundibular cystic structure with several sebaceous lobules attached to it via sebaceous ducts. The red arrow shows compactly laminated fibroplasia around the epithelial units. The green arrows show clefts between the fibroepithelial units and surrounding altered stroma (H and E, ×4). (b) The lesion is located in the dermis and subcutis (×4)

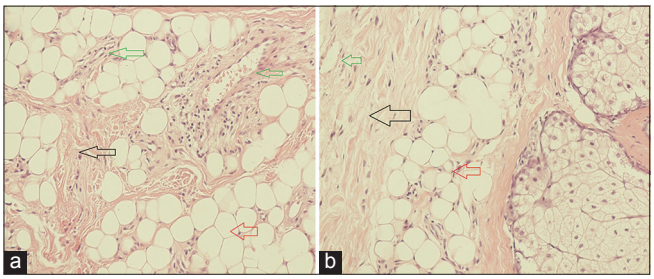

Figure 2: Papule on face: (a and b) The mesenchymal component around the epithelial units consists of fibrillary bundles of collagen (black arrows), immature appearing adipocytes (red arrows), and many small venules (green arrows) (H and E, ×20)

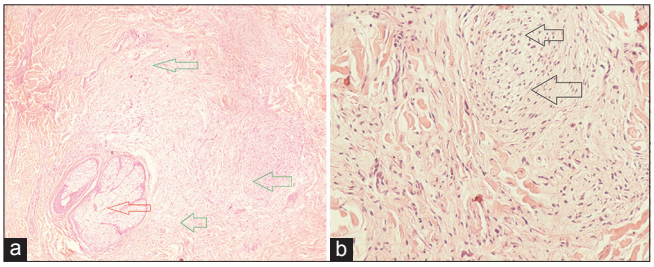

Figure 3: Dome-shaped papule on the neck. (a) Cutaneous neurofibroma (green arrow) closely associated with skin adnexal structures (red arrow) (H and E, ×10). (b) Typical wavy nuclei with pointed ends of neurofibroma (black arrows) (×40)

DISCUSSION

This case fulfilled the histological criteria laid down by Kimura *et al*.[1] for the diagnosis of FSCH. The criteria include:

- An infundibular cystic structure with several sebaceous lobules attached to it via sebaceous ducts [Figure 1a]
- Compactly laminated fibroplasia around the epithelial units [Figure 1a]
- Mesenchymal component around the epithelial units comprising fibrillary bundles of collagen, adipocytes, and many small venules [Figure 2]
- Clefts between the fibroepithelial units and surrounding altered stroma [Figure 1a]
- Dermal and subcutaneous location of the tumor [Figure 1b].

The association of FSCH and neurofibroma has been previously reported in a case of neurofibromatosis Type I.[7] However, the authors of this case report considered this to be an incidental finding since there were no previous reports of FSCH in association with neurofibroma.

At the histogenetic level, both cutaneous neurofibromas and FSCH are closely associated with hair follicular structures. A recently described case of neurofolliculosebaceous hamartoma[8] further supports the hypothesis that FSCH and cutaneous neurofibromas may have common histogenesis. Nestin is an intermediate filament protein expressed in the follicular stem cells in the bulge region of the hair follicle and its actions may result in the formation of various mesenchymal and neural tissues. Both cutaneous neurofibromas and FSCH show nestin immunopositivity.[9,10] The development of cutaneous neurofibromas includes the recruitment of multipotent precursor cells. These cells may be derived from the multipotent cells of the hair roots, which are often intimately associated with microscopic neurofibromas.[11]

CONCLUSION

FSCH is probably not as rare as once thought and it should be kept in the differential diagnosis of papules and nodules in the head and neck region. Its association with sporadic cutaneous neurofibroma, as seen in this case, needs to be looked into as and when further cases are reported.

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Conflicts of interest
There are no conflicts of interest.

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