

# Congenital folliculosebaceous cystic hamartoma: A case report

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Conflict of interest : none to declare

Received: April 10, 2011

Accepted: May 18, 2011

Folliculosebaceous Cystic Hamartoma (FSCH) is a cutaneous hamartoma comprised of follicular, sebaceous and mesenchymal elements, and usually occurring during adulthood. Congenital and childhood presentations of this lesion are exceedingly rare. We describe herein a case of congenital FSCH on the midline and posterior region of the neck in a 1-year-old male infant and highlight the clinicopathological features of this rare lesion.

**Keywords:** congenital, folliculosebaceous cystic hamartoma, hamartoma

Iran J Dermatol 2011; 14: 109-112

## INTRODUCTION

Folliculosebaceous Cystic Hamartoma (FSCH), first described by Kimura et al, in 1991, is a rare cutaneous hamartoma with fewer than 50 reported cases to date. It consists of follicular, sebaceous and mesenchymal elements<sup>1,2</sup> and usually presents during adulthood as a dome-shaped or pedunculated nodule on the center of the face or scalp<sup>2</sup>. Congenital and childhood presentations of this lesion are exceedingly rare<sup>3-5</sup>. This report presents a case of congenital FSCH in a 1-year-old boy and highlights the clinicopathological features of this rare tumor.

## CASE REPORT

A 1-year-old male infant was visited with a soft

tissue mass on the midline and posterior region of his neck that was present since birth. It first appeared as a 1×1cm nodule with continuous growth until his first presentation to our department. No other skin lesions were found and no family history of similar lesions was mentioned. Clinical examination revealed a multinodular and exophytic cutaneous mass with intact overlying skin, measuring 4.5×2.5cm in diameter (Figure 1). Due to midline localization of the lesion, a Computed Tomography (CT) scan was performed in order to exclude any possible intracranial connection. Then, the patient underwent complete surgical resection of the lesion under general anesthesia.

Macroscopically, the incised surface of the lesion revealed several nodules and cystic spaces like keratinous cysts nested in a white fibrous tissue



**Figure 1.** Exophytic cutaneous mass in the midline and posterior region of the neck

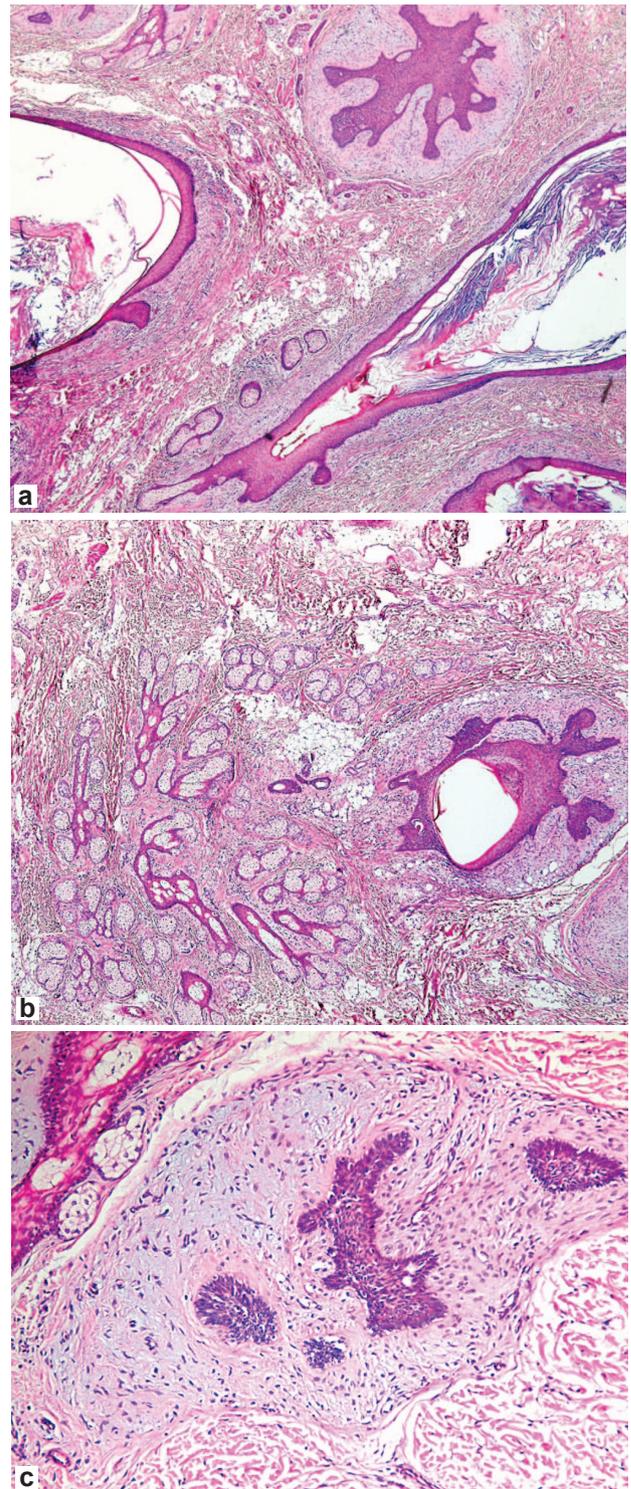
within the dermis and subcutis (Figure 2).

Microscopically, the epidermis was normal. The dermal lesion was composed of markedly dilated follicular cystic structures with sebaceous lobules radiating from its wall, cystic structures lined by keratinized squamous epithelium and some bizarre-shaped follicular structures which were all surrounded by mesenchymal stroma



**Figure 2.** Cut surface of the lesion showed several nodules and cystic spaces like keratinous cysts

consisting of fibrous tissue with increased blood vessels (Figure 3).



**Figure 3.** Typical histopathologic findings in folliculosebaceous cystic hamartoma, including (a) infundibulo-cystic structures that are surrounded by mesenchymal stroma (H&E\*40). (b) sebaceous hyperplasia, abundant fibrous stroma with adipose tissue (H&E\*40). (c) Bizarre-shaped follicular structures surrounded by perifollicular fibroplasia (H&E\*200)

## DISCUSSION

FSCH is a rare cutaneous hamartoma which is composed of dilated, cystic pilosebaceous structures surrounded by various mesenchymal elements. It is mostly found in adults with a striking predilection for the central face and scalp; however, few cases are reported with the involvement of extremities and genital area or the nipple<sup>2,4,6,7</sup>.

To date, only three articles have described childhood FSCH, one on the scalp, one on the leg and one on the nape of the neck. The age of patients was 6 months, 2 years and 9 months, respectively<sup>3-5</sup>. Among these patients, only two had their lesions since birth<sup>3,5</sup>. Therefore, our case was the third case of congenital FSCH that was diagnosed during childhood.

The exact histogenesis of FSCH is still controversial. Schulz and Hartschuh presented convincing evidence that this lesion was a trichofolliculoma at very late stage with the follicular structures in a state of involution<sup>8</sup>. This hypothesis can not explain either congenital FSCH or those without a pre-existing lesion as our case. In another pathogenesis hypothesis, the inductive effect of perifollicular stroma on various portions of a dilated follicle has been proposed. In FSCH, this mesenchymal element induces prominent isthmic and sebaceous differentiation but in trichofolliculoma, focal induction of the lower segment can result in secondary follicle formation. The perifollicular stroma may also induce other cutaneous components such as vascular and neural structures as well as mesenchymal elements including mucin or adipose tissue in different proportions<sup>9</sup>. Using this hypothesis, we can explain the similarities between FSCH and trichofolliculoma.

As a significant number of the reported cases (like ours) were from Asia, Badr proposed that FSCH may have a predilection for Asians<sup>7</sup>.

Although it has speculated that FSCH is slightly more common in females, with previously reported female/male ratios of 3:2 and 5:3, two of three pediatric cases, as well as our case, are male<sup>2</sup>.

The lesion does not have distinctive typifying clinical features and therefore the clinical diagnosis in all reported cases included diagnoses other than FSCH, such as intradermal nevus, sebaceous hyperplasia, basal cell carcinoma, nevus sebaceous,

meningocele, and dermoid cyst<sup>3</sup>.

It usually presents as a solitary papule, nodule or polypoid lesion with intact overlying skin surface that is rubbery to firm in consistency. The majority (90%) do not exceed 25 mm in diameter. However, giant variants have been reported, usually in the genital area and also in a previous congenital case on the posterolateral aspect of the neck<sup>5,10</sup>.

The histopathological features were described by Kimura et al, have not been modified essentially ever since. They include (i) an infundibulocystic structure with sebaceous glands radiating around, (ii) laminated fibroplasia encircling the epithelial component, (iii) mesenchymal changes, mainly increased vascularity and presence of mature fat, (iv) clefts between the epithelial component as well as between this and the adjacent dermis and (v) confinement to the dermis<sup>2</sup>. The present case had histopathological features similar to those of the previously reported cases of FSCH.

The major differential diagnosis of FSCH is sebaceous trichofolliculoma. The most important histopathological clue is the stroma of FSCH that contains thin bundles of collagen, adipocytes, increased number of capillaries and venules and, occasionally, proliferation of nerve fibers. Such a stroma is not found in sebaceous trichofolliculomas<sup>2</sup>.

The lesion is usually excised or shaved. The giant variant described on the scrotum in a 74-year-old man was successfully treated by CO<sub>2</sub> laser and acitretin therapy<sup>10</sup>.

All reported cases of FSCH have proved to be benign biologically; none of them relapsed or developed into carcinoma, and no association with gastrointestinal tract malignancy or Muir–Torre syndrome has been identified<sup>2,3,5</sup>.

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