

CASE REPORT

A Case of Steatocystoma Multiplex Limited to the Scalp

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We report here on a 63-year-old woman who had several small, yellowish papules on the scalp for the previous 2 years. There was no family history of similar lesions. Yellowish, creamy material was expressed from a papule during punch biopsy. Histologic examination from the lesion revealed the typical features of steatocystoma multiplex. We report here on this rare variant of steatocystoma multiplex that was limited to the scalp. (**Ann Dermatol (Seoul) 21(1) 106~109, 2009**)

-Keywords-

Scalp, Steatocystoma multiplex

INTRODUCTION

Steatocystoma multiplex is a hamartomatous malformation of the pilosebaceous duct junction¹. It is a distinct clinical condition that's characterized by numerous small, skin-colored or yellowish, cutaneous cysts². The lesions typically appear in childhood or early adulthood, but in some patients the lesions have developed late in life^{3,4}. The cysts are located principally on the axillae, groin, trunk, scrotum and proximal extremities and they are rarely localized to the face and scalp³⁻⁶. Steatocystoma multiplex located on the face and scalp has been divided into three types³⁻⁶: 1) a facial papular variant type⁵, 2) sebocystomatosis⁶ and 3) cysts located exclusively on the scalp^{3,4}. We report herein a rare variant of steatocystoma multiplex

Received May 13, 2008, Accepted for publication September 30, 2008
*This work was supported by the SRC/ERC program of MOST/KOSEF (No. R11-2005-017) and Basic Research Promotion Fund, KRF-2007-531-E00045).

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that developed in an elderly woman and this condition was limited to the scalp.

CASE REPORT

A 63-year-old woman visited our clinic because of papules on her scalp, and these papules had been slowly growing over a period of 2 years. On physical examination, several smooth-surfaced, round, yellowish papules were located on the left temporal part of the scalp. The diameter of the lesions varied from 3~5 mm (Fig. 1). No other lesions were observed on the extremities, trunk or face. The patient's past and family histories were non-contributory and the results of routine laboratory findings were within normal limits. A punch biopsy was performed to differentiate an eruptive vellus hair cyst, xanthomatosis and steatocystoma multiplex from other dermatologic conditions. When the surface skin of the yellowish papule was disrupted, a homogenous, creamy, odorless, yellowish material was expressed. The histologic examination showed an empty cyst with a folded wall in the



Fig. 1. Several asymptomatic, yellowish papules on the scalp varying in size from 3 to 5 mm.

mid-dermis. The wall of the cyst was composed of several layers of stratified squamous epithelium without a granular layer (Fig. 2A). Flattened sebaceous gland lobules were lying close to the cyst wall (Fig. 2B): these findings were consistent with steatocystoma. Some of the lesions were surgically excised, but the patient declined further treatment.

DISCUSSION

Steatocystoma multiplex is an uncommon disorder that's characterized by multiple soft, movable, small, cystic nodules. The diameter of lesions typically varies from 0.2 ~2 cm, but the nodules may be larger. The overlying epidermis is usually normal with no central punctum present. The lesions are typically located on the upper trunk, neck, axillae, scrotum and proximal extremities².

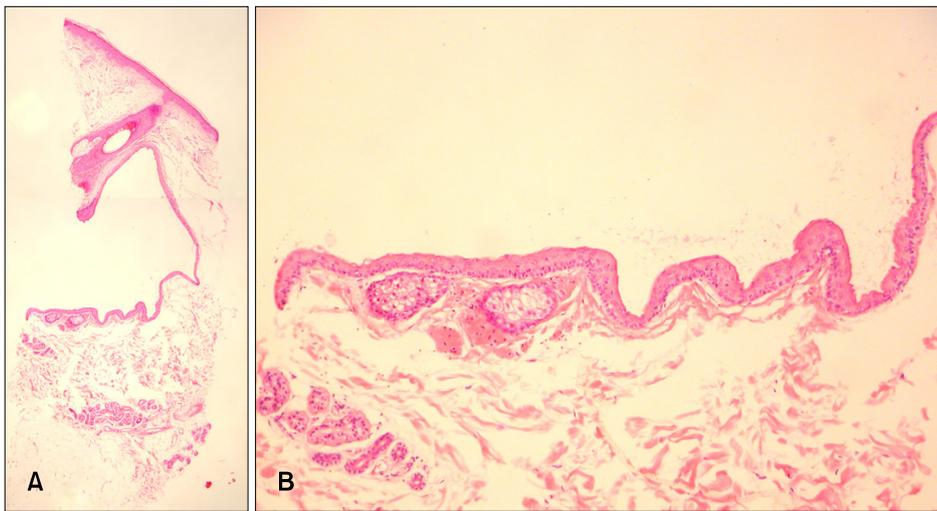


Fig. 2. (A) A folded cyst wall consisted of several layers of stratified epithelium (H&E, ×40). (B) Flattened sebaceous gland lobules close to the cyst wall (H&E, ×200).

Table 1. Summary of the reported cases of steatocystoma multiplex limited to the scalp

No.	Age (yrs)/ Gender	Number	Size (cm)	Lesions below the neck	Family history	Treatment	Authors
1	71/F	120	<0.2	No	No	Incision	Kumakiri and Yajima ³
2	70/M	4	1.3~2.4	No	No	Excision	Marley et al ⁴
3	50/M	Multiple	—	No	No	—	Ferrandiz and Peyri ⁷
4	40/M	12	—	No	No	—	Ambrojo Antunez et al ⁸
5	58/F	40	0.1~0.4	No	No	Observation	Belinchon et al ⁹
6	70/M	Multiple	0.5~1.1	No	No	Observation	Mortazavi et al ¹⁰
7	35/M	Multiple	0.5~1.5	No	No	Excision	Setoyama et al ¹¹
7	46/M	300	0.2~0.8	No	No	Excision	Lim et al ¹²
8	63/F	7	0.3~0.5	No	No	Observation	Present case

Table 2. Differences among typical steatocystoma multiplex, steatocystoma multiplex limited to the scalp and steatocystoma simplex

	Steatocystoma multiplex		Steatocystoma simplex
	Typical	Limited to the scalp	
Pathogenesis	Nevoid or hamartomatous malformation of the pilosebaceous duct junction		
Inheritance	Autosomal dominant	Sporadic	Sporadic
Age of onset	Adolescence or early adulthood	Adulthood or elderly individuals	Adulthood
Gender predominance	M>F	Equally affected	Equally affected
No. of lesions	Multiple	Multiple	Solitary
Recurrence	None	None	None
Prognosis	May be progressive with inflamed cysts that rupture and heal with scars	Benign	Benign

There may also be cases where there are scattered lesions on the scalp or face; however, it is very rare that the lesions are limited to the scalp (Table 1)^{3,4,7-12}.

The pathogenesis of these lesions has been extensively studied. Steatocystomas were initially thought to be sebaceous or retention cysts¹³. In 1964, Kligman and Kirschbaum¹⁴ included these as a variant of dermoid cysts because of the presence of both hair shafts and sebaceous lobules. Steatocystomas are now recognized as a nevoid or hamartomatous malformation of the pilosebaceous junction¹.

Steatocystoma multiplex is usually hereditary and it is inherited as an autosomal dominant trait. However, it is nonhereditary for cases that the lesions are mainly limited to the scalp. The differences between typical steatocystoma multiplex and that limited to the scalp are presented in Table 2. In 1996, Lee et al⁶ reported on a case of sebocystomatosis that displayed multiple lesions of steatocystoma multiplex confined only to the forehead and frontal scalp together with congenital alopecia, and this was considered as a clinical variant of steatocystoma multiplex. Steatocystoma simplex, which is rarely found on the scalp, was first described as a distinct entity by Brownstein¹⁵ in 1982. It is similar to our case for the pathogenesis, the genetic propensity and the prognosis; however, steatocystoma simplex has a solitary nature (Table 2)¹⁵⁻¹⁸. Lim et al¹² presented a patient with eruptive steatocystoma multiplex on the scalp. It was different from our case in that the lesions had appeared abruptly within a short period of time and the cysts showed an ovoid dilated cyst wall on histological study. Our case is educationally unique for the following points: (1) the presence of several cysts limited to the scalp, (2) no family history of similar lesions, (3) a late onset for the condition and (4) no congenital alopecia.

The causative factors of steatocystoma multiplex remain unclear, yet Setoyama et al¹¹ suggested that trauma, infection or immunological events might be responsible. Some authors have reported that steatocystoma multiplex has been associated with ichthyosis and koilonychia, pachyonychia congenita, acrokeratosis verruciformis of Hopf, hypertrophic lichen planus, hypohidrosis, hypothyroidism, hidradenitis suppurativa and hypotrichosis^{4,18,19}. For steatocystoma multiplex limited to the scalp, there have been no reports about specific causative factors and the relationship with other disorder, as was the case for our patient^{3,4,7-12}.

The clinical differential diagnosis of steatocystoma multiplex includes multiple epidermoid cysts, eruptive vellus hair cyst, neurofibromatosis, lipomas and xanthomatosis. A skin biopsy specimen from a representative lesion is

diagnostic. The characteristic histopathologic finding of the disease is the presence of flattened sebaceous lobules close to the cystic wall. The wall is composed of stratified, squamous epithelium without a granular layer. There are acellular eosinophilic cuticles on the luminal side of the cyst wall and keratin, oil or hairs in the lumen².

Treatment of steatocystoma multiplex is largely symptomatic. Removal of these lesions can be successfully achieved with simple excision or drainage of the cyst materials. Inflamed lesions have been reported to respond to intralesional steroid injections, carbon dioxide laser, oral retinoids or cryotherapy². The lesions are typically asymptomatic, but they are often cosmetically disconcerting. Our patient declined further treatment after the diagnosis was made.

We report here on a rare case of steatocystoma multiplex that sporadically developed in an elderly woman and the condition was limited to the scalp.

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