Case Report

Multiple pilomatricomas: A Rare Case Report

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INTRODUCTION

Pilomatricoma is a benign adnexal tumor of childhood first described in 1880 as calcifying epithelioma of the sebaceous gland by Malherbe and Chénantais[1]. This tumor most commonly presents as an asymptomatic, firm, slow-growing subcutaneous nodule on the head and less frequently on the neck and upper limbs[2]. Pilomatricomas usually occur in the first and second decades of life[2]. The exact pathogenesis of this tumor is still unknown. Some pilomatricomas are caused by somatic mutations in the β-catenin gene. β-catenin, as a downstream effector of the Wnt signaling pathway, influences hair follicle cell proliferation and
differentiation[2]. Anetodermic pilomatricoma is a rare variant of pilomatricoma that usually presents as an asymptomatic, rapidly growing, keloid-like lesion in the upper arms or shoulders[3]. It is characterized microscopically by basal keratinocyte hyperplasia, dilated lymphatic vessels, disruption of collagen fibers and absence of elastic fibers in the dermis above pilomatricoma, and a deep dermal pilomatricoma with islands of basaloid cells, eosinophilic shadow cells, and a transition zone of retained nuclei between them. Areas of calcification, ossification or necrosis, and foreign-body granulomatous may also be seen[3]. Multiple pilomatricomas may be sporadic, familial, or syndromic[2]. Multiple pilomatricoma have been reported in association with Gardner syndrome and MYH-associated polyposis, myotonic dystrophy, Rubinstein Taybi syndrome, Sotos syndrome, gliomatosis cerebri, and Turner syndrome[2].

**Case Presentation**

We present a nine-year-old boy with a two-month history of two rapidly growing, smooth-surfaced, firm tumoral lesions in the left arm and left flank (Figure 1). The lesions measuring 1.7×1×0.7 cm and 1.5×1×0.9 cm with resembled keloids clinically, but there was no history of preceding trauma or ulceration. There was no pain or pruritus. The patient was a healthy boy without any previous illnesses. General physical examination was unremarkable. we have done an incisional biopsy from the center of the tumoral lesion in the left arm. After removing the specimen, a creamy white material in the center of the lesion discharged easily by lateral pressure. Microscopic examination of the specimen showed irregular nests of basaloid cells with different stages of trichocyte differentiation and the formation of ghost cells in the dermis. Necrotic areas, calcification, and foreign body type giant
cells were also seen. The diagnosis of pilomatricoma was made based on the histopathologic picture of the lesions (Figure 2).

**Fig 1.** Nodular lesion on the left arm (A) and left flank (B) with smooth and erythematous surface

**Fig 2.** Nests of basaloid cells, ghost cells and calcification in the dermis

A and B. Hematoxylin and eosin stain; original magnification 10 x

C. Hematoxylin and eosin stain; original magnification 40 x
Discussion

A pilomatricoma is an uncommon tumor originating from hair matrix cells. Clinically, these lesions present with atrophic, wrinkled, folded, brownish, keloid-like, or bullous appearances. This tumor most commonly presents on the head and less frequently on the neck and upper limbs and usually occur in the first and second decades of life. pilomatricoma is characterized by epithelial islands of basaloid and shadow cells, Calcification and Ossification occurs occasionally, and foreign-body reaction is usually present [2]. Multiple lesions are rare and usually associated with myotonic dystrophy, Gardner syndrome and MYH-associated polyposis. Rubinstein Taybi syndrome, Sotos syndrome, gliomatosis cerebri, and Turner syndrome[2]. Yagi et al. reported a case of Rubinstein Taybi syndrome with mental retardation, short stature, strabismus, thick eyebrows, long eyelash, beaked nose, and broad thumbs with multiple keloid-like lesions with microscopic features of pilomatricoma[4]. The lesions in our case developed on the arm, 13-year-old with two keloid-like pilomatricomas without any associated abnormalities. Careful follow-up and searching for related problems is advisable in these situations, in addition to complete excision of the pilomatricomas.

In conclusion, pilomatricoma can be diagnosed clinically and excisional biopsy is advised for diagnosis. Because the recurrence rate after excision is low, pilomatrical carcinoma should be considered in the case of local recurrence. Multiple pilomatricomas have been associated with various syndromes so long term follow up is recommended.
PATIENT CONSENT
The parents of the patient provided written informed consent for the publication and the use of his images.

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Conflict of interest
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References