

## An 8-Year-Old Girl with Multiple Subcutaneous Nodules: Pilomatrixoma

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### Abstract

Pilomatrixoma commonly occurs in children as a single tumor. Multiple tumors are rare. This case report describes the presentation of an 8-year-old girl with multiple pilomatrixomas. We report this for the benefit of pediatricians.

**Keywords:** Multiple pilomatrixom; Childhood

### Introduction

A pilomatrixoma (also called *Malherbe's calcifying epithelioma*) is a benign tumor originating from hair follicle matrix cells [1]. The tumor most commonly occurs in the head and neck region [2]. The age of onset is bimodal. In over 60% of patients, the first peak occurrence is before the third decade of life, while the second peak takes place between the sixth and seventh decades [3]. It commonly occurs in children as a single tumor [4]. Although pilomatrixoma has been properly defined over the years, it is frequently misdiagnosed. Because most case reports are documented in the dermatology and otolaryngology literature, general pediatricians might not be familiar with the condition [5]. This case report describes the presentation of multiple pilomatrixomas in a child.

### Case Report

An 8-year-old girl presented with 4 subcutaneous, rock-hard nodules. The average time from the onset of the appearance of other nodules was about 12 months. The sites of occurrence were the right eyebrow, neck, right scapular region, and upper left region of the abdomen. Only the nodule in the abdominal region was 1 cm in diameter, the others were 5 mm in diameter. The nodules were nontender and painless except the one in the right scapular region. The overlying skin was normal in appearance, with no evidence of ulceration or discoloration. No concurrent disorders were observed. The preliminary clinical diagnosis was multiple pilomatrixoma. The nodules were tender and painful, and the larger one was surgically excised. Histologically, the nodules consisted of acellular material in which ghost cells (Figure 1) were prominent, together with foreign body giant cells and calcification. At the periphery, there were focal areas of basaloid cells (Figure 2). The pathological diagnosis was pilomatrixoma. No recurrence has been observed during 8 months' follow-up.

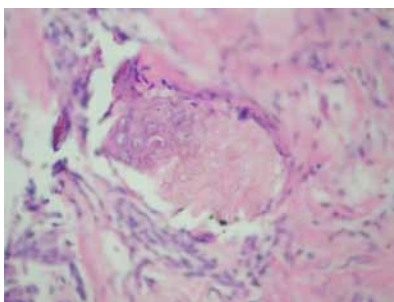


Figure 1: Ghost Cells.

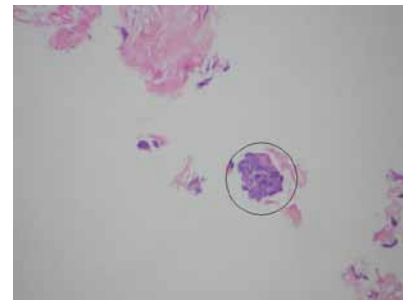


Figure 2: Basaloid Cells.

### Discussion

Pilomatrixoma, a benign neoplasm of the hair follicle, was initially thought to arise from sebaceous glands and was called *calcifying epithelioma of Malherbe* by Malherbe and Chenantais [6]. Pilomatrixoma is the most common superficial tumor in children and occurs with greater frequency in girls [7]. Pilomatrixomas are usually solitary nodules, but multiple occurrences have been observed in 2% to 10% of reported cases [2]. However, Turhan Haktanir et al. [8] reported multiple pilomatrixomas 50% in their series of Turkish patients. Our patient, similar with these Turkish series, had multiple tumors. A further study reviewing a wide series of Turkish patients are necessary. Multiple tumors may be found in association with Gardner's syndrome, myotonic muscular dystrophy, and Turner's syndrome [2]. In our patient, the tumors were not of any syndrome, anomaly, or other disorder.

On over 50% of patients, pilomatrixoma most commonly occurs in the head and neck with the upper extremity a distant second, followed by the trunk and lower extremities [2,6]. In our patient, the occurrence of tumors was similar to that reported in the literature. Typical tumors measure 5 mm to several centimeters [7]. Pilomatrixomas grow slowly and typically attain their full size over 6 to 12 months. They are usually asymptomatic [2]. In our patient, the tumors were 5 mm and 1 cm in

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size for nearly 12 months, and 1 of 4 tumors was symptomatic with tenderness and pain.

The diagnosis of pilomatricoma can generally be made with a clinical examination but requires careful examination and a high index of suspicion based on palpation of a superficial, rock-hard nodule, and confirmed by histopathologic examination [1]. However, other entities to be considered in the differential diagnosis include epidermal inclusion cyst, foreign body reaction, calcinosis cutis, and lipoma. As described by Sanchez and associates, the classic cytologic appearance of pilomatricoma consists of varying amounts of basaloid and ghost cells [9]. Basaloid cells are basophilic and are about three times the size of a lymphocyte. They contain large uniform nuclei with prominent nucleoli, and their cytoplasm is scant with poorly defined borders. In contrast, ghost cells are eosinophilic and anucleate, which gives the cell a hollow or 'ghostlike' appearance. Other key components that assist in the diagnosis are foreign body giant cells and nucleated squamous cells. Calcium deposits are less definitive according to Wang et al. [10] as they were present in only 55% of the cases in their study. In our patient, histopathologic findings were all typical for pilomatricoma.

As spontaneous regression of pilomatricoma has never been observed and malignant degeneration is rare, surgical excision with clear margins is the treatment of choice [1]. Small nodules are left untreated unless they become large or symptomatic [11]. In our patient, the 2 nodules, which were 5 mm and asymptomatic, were left untreated. The incidence of recurrence after surgery has been reported as being between 0% and 6% [1,2]. At 8 months' follow-up, there has been no recurrence in our patient.

Although pilomatricoma occurs mostly in children, general pediatricians are not as well informed about this tumor as are dermatologists and otolaryngologists. We report this case for the benefit of pediatricians.

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