CASE REPORT

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Pilomatricoma of the Upper Extremities: A Case Report

Üst Ekstremite Yerleşimli Pilomatrikoma: Olgu Sunumu

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ABSTRACT

Pilomatricoma is a benign skin tumor, which is generally spotted in neck region along with the head. Isolated lesions are the usual detected. Malignant transformations are considered as seldom occurrences. A 17-year-old boy with a pilomatricoma on his left arm was presented as our case in this study. On ultrasonography, the hypoechoic solid lesion with calcification spots defined 15x8 mm sized, well-defined margins localized in the subcutaneous fatty tissue. Definitive diagnosis is made by histopathology. From histopathological point of view, shadow or ghost cells are characteristic. Pilomatricoma should be taken into consideration in the differential diagnosis of superficial or subcutaneous, painless masses of head, neck and extremities. Surgical excision is curative and the recurrent rate is low.

Keywords: Benign tumor, Pediatric, Pilomatricoma, Pilomatrixoma, Skin lesions

ÖZ

Pilomatrikoma, genellikle baş ve boyun bölgesinde yerleşim gösteren iyi huylu bir deri tümörüdür. Genellikle izole bir lezyon olarak görülür ve nadiren malign seyreder. Bu yazıda sol kolunda pilomatrikoma saptanan 17 yaşındaki erkek olgu incelenmiştir. USG'de cilt altı yağlı dokuda lokalize düzgün sınırlı 15x8 mm boyutlu mm'lik kalsifik odaklar içeren hipoekoik solid lezyon izlenmiştir. Pilomatrikomanın kesin tanısı histopatolojik olarak konulur. Histopatolojik incelemede gölge ve hayalet hücrelerin görülmesi karakteristiktir. Baş, boyun ve ekstremitelerde cilt altı yüzeyel yerleşimli ağrısız kitle ile karşılaşıldığında pilomatrikoma ayırıcı tanıda değerlendirilmelidir. Cerrahi eksizyon tam kür sağlamaktadır ve nüks oranı düşüktür.

Anahtar Sözcükler: İyi huylu tümör, Pediatrik, Pilomatrikoma, Pilomatriksoma, Deri lezyonları

INTRODUCTION

Pilomatricomas (PMX) are benign, cutaneous tumors of the hair matrix cells (1). A pilomatricoma is typically present as a superficial, mobile, small, hard mass. The most commonly, it presents as a single lump found on head or neck with varying sizes from 0.5 to 3 cm in diameter (2,3). Pilomatricomas may be found in all age groups with bimodal peaks in those under 20 years and a second less significant peak between 50-65 years of age (4). It demonstrates a higher prevalence in female (3). It is the second most common superficial tumor excised in children (5). This lesion is considered relatively common. According to the study by Marrogi et al., PMX accounts for one of every 500 specimens submitted by dermatologists (6). Complications of PMX are uncommon. However occasionally they grow to giant size and malignity has been very rarely reported.

A firm, slow growing, and non-tender are the most frequent clinical features encountered. Superficial tumors develop a bluish-gray hue, and occasionally protuberant, red nodules are present. Showing a central whitish or grayish-blue structureless area on dermoscopy may suggest PMX. If the nature of the skin lesions is unclear, ultrasound scan might be recommended. Fine-needle aspiration cytology can be a useful method to diagnose PMX including pediatric patients (7).

Management of PMX is marginal excision and histopathological examination is required for definitive diagnosis (1). The tumor is encompassed of a basaloid proliferation resembling the hair matrix cells, which matures into structureless esosinophilic cells lacking nuclei called shadow cells. The shadow cell area signifies differentiation towards the hair cortex. Frequently there are areas of calcification within the shadow cell regions. For PMX and most benign skin tumors, surgical removal is the most preferred treatment. They do not disappear by themselves, and if incompletely removed, they might recur.

Our aim was to keep PMX in mind during the differential diagnosis of subcutaneous mass lesions that do not match the lymph node localization in the childhood.

CASE REPORT

A 17-year-old boy was presented with a one-year-old enlarging skin lesion on his left arm. He denied any history of trauma, fever, weight loss, tingling or fatigue. According to the patient he had contact with animals in the village. Last year he felt his extremities cold. The patient did not report pain or any other symptoms. Physical examination was unremarkable except for a 20x10 mm solid, painless mass over the left arm. It was superficial and easily mobile. Laboratory parameters were normal (Table 1).

Table 1: Laboratory parameters

Parameters	Range	Values
White blood cells ($10^3/\mu$L)	4.27-9.84	4.630
Hemoglobin (g/dL)	10.2-13.4	17.3
Platelet ($10^3/\mu$L)	144-597	276
Neutrophils (%)	22.4-74.5	56.2
Eosinophils (%)	0-4.7	2..1
Lymphocytes (%)	15.5-68.6	33..9
Glucose (mg/dL)	74-106	89
Urea (mg/dL)	16.6-48.5	17.9
Creatinine (mg/dL)	0.4-0.6	0.98
Alanine transaminase (U/L)	0-41	17
Aspartate aminotransferase (U/L)	0-40	29
Alkaline phosphatase (U/L)	142-335	100
Lactate dehydrogenase (U/L)	120-300	223
Creatine Kinase (U/L)	20-200	99
Calcium (mg/dL)	8.8-10.8	10.9
Fosfor (mg/dL)	2.7-4.9	3.3
Magnesium (mg/dL)	1.58-2.55	2.1
Cholesterol (mg/dL)	0-200	148
Triglycerides (mg/dL)	0-200	74
HDL Cholesterol (mg/dL)	35-75	45
LDL Cholesterol (mg/dL)	0-130	88
Serum iron (μg/DL)	33-193	171
Sodium (mmol/L)	136-145	137
Potassium (mmol/L)	3.5-5.1	4.8
Chloride (mmol/L)	98-107	100
Blood urea nitrogen (mg/dL)	6-20	8.4
C-Reactive Protein (mg/dL)	0-0.5	0

On ultrasonography, the hypoechoic solid lesion with calcification spots defined 15x8 mm sized, well-defined margins localized in the subcutaneous fatty tissue. There was no vascularization in the lesion. Radiologically, these results suggested benign lesions.

Under general anesthesia, the lesions were excised (Figure 1). After cross-sectioned, it was seen that it had yellow-orange colored calcified areas in places. Histopathologically, the lesion has typical basaloid cells (Figure 2), ghost cells (Figure 3) and calcification. The patient was discharged on same day, recovered well and is in perfect health.

DISCUSSION

Pilomatricomas is a neoplasm of the hair follicle matrix cells. Towards the end of 19th century, Malherbe and Chenantais first described it as a “calcifying epithelioma” thought to be

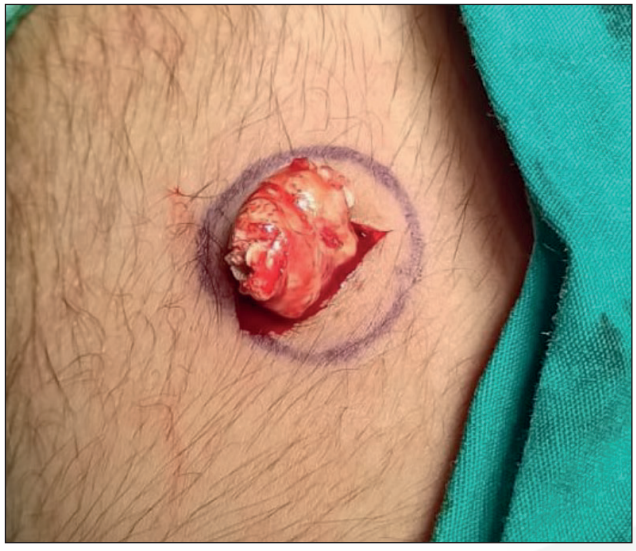

Figure 1: Encapsulated solid mass removed thorough left arm.

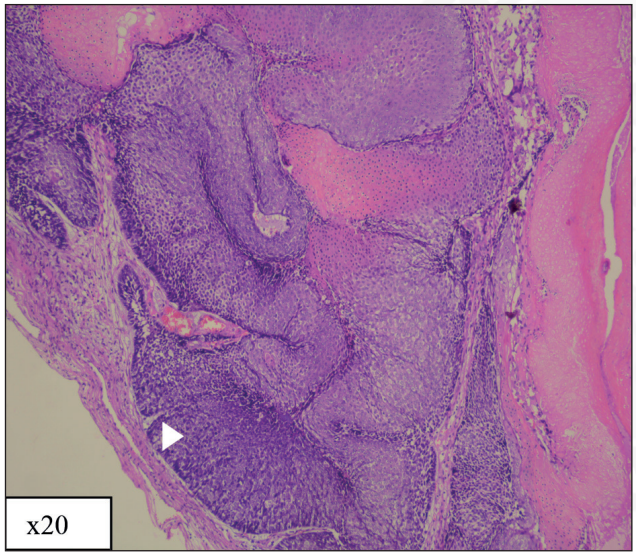

Figure 2: Basaloid cells area is seen where indicated by the black arrow. Slide stained with hematoxylin and eosin (x20).

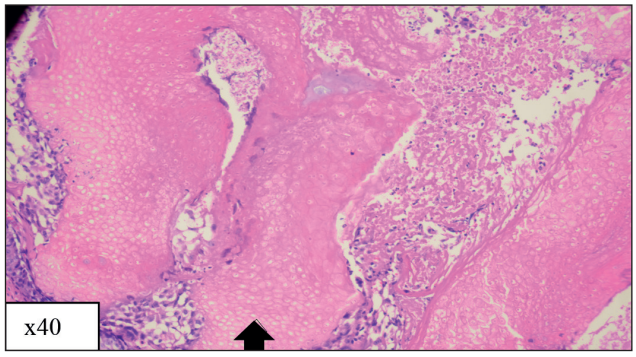

Figure 3: "Ghost cells" is seen where indicated by the black arrow. Slide stained with hematoxylin and eosin (x40).

of sebaceous gland origin (8). Forbis and Helwig examined the cortex of hair follicle and describes it as cell of origin. After this elucidation, PMX term has been proposed by them in '61 (9). The etiopathogenesis of PMX is unknown. Literature shows it is associated with trauma, β-catenin mutations, polyoma virus and genetic disorders such as xeroderma pigmentosum, myotonic dystrophy Gardner syndrome, Turner syndrome, basal cell nevus syndrome (2-4). Moehlenbeck's statistical study, which analyzed 140,000 skin tumor samples at their department of dermatology, showed an incidence of 0.12% of PMX (10). In society, 97% of PMX seen in white people (11). Gender distribution demonstrates a female predominance (4).

The head and neck regions are the most typical areas to locate these lesions. It is more likely to find PMX within the head region compared to the neck region. Within the head region specifically, the tumor locations in decreasing order are the cheek, periorbital and periauricular areas (4). They have also been described in various upper and low extremity locations. No cases have been reported on soles, palms or genital region (12). Clinically, PMX can be present as a solid mass, mobile attached to the skin. It can be present with overlying normal skin or with reddish and bluish tint (4). In this case, characteristics of palpation outcome of a superficial firm nodule are painlessness, or tenderness.

The differential diagnosis for PMX is varied. The differential diagnosis includes dermoid cyst, epidermal inclusion cyst, lipoma, hemangiomas, ossified hematoma, osteoma cutis, degenerating fibroxanthoma and foreign body reaction. Pilomatricomas may be mistaken for these lesions. According to literature, it has been emphasized that "tent sign" and "teeter-totter" findings may be useful in distinguishing PMX from other lesions. Stretching of the skin over the tumor shows the "tent sign" with multiple facets and angles, a pathognomonic sign for PMX (13). Pressing on one edge of the lesion causes the opposite edge to bulge from the skin like a "teeter-totter" (14). Imaging methods are used preoperatively in cases where a definitive diagnosis cannot be made. In this case, ultrasound as first option for us since it is a noninvasive tool, relatively quick, cost effective and sedative-free. It can demonstrate the degree of calcification, position and the continuity of the lesion deeper structures. There are also articles in which CT an MRI are used in the diagnosis phase (15,16).

On fine needle aspiration, the diagnosis of PMX may be extremely difficult. The presence of the complete spectrum of PMX cytological characteristics, such as basaloid cells, calcium deposits, naked nuclei, shadow ("ghost"), and giant cells, inflammatory background, allows a significant well-done preoperative diagnosis (17). In our case, demonstrated a characteristic histologic appearance including shadow (or ''ghost'') cells, basaloid cells and areas of calcification.

In pediatric patients, PMX is now considered as a more familiar diagnosis for cysts. In the past, it was not widely known on cytology.

Surgical excision remains the gold standard of treatment with low recurrence rates during the follow-up period (4). In cases with tumor adherence to the dermis, the overlying skin might be excised. Tumor size does not correlate with prognosis, although recurrence may occur if incompletely excised. If a lesion recurs after excision or rapidly enlarges, it should be excised due to malignant potential or possible misdiagnosis (18).

Hair matrix cell originated benign tumors are pilomatricomas that mostly appear in children. It should be taken into consideration in the differential diagnosis of superficial or subcutaneous masses in children. Imaging studies are generally not necessary unless the symptoms or lesion's location warrants such diagnostic assessments.

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Author Contributions

Concept, design, validation, writing-original draft preparation, methodology, writing review and editing: **Yusuf Doruk Bilgi,** Data analysis: **Gülden Taşova Yılmaz.** The final version article has been read and approved by all authors.

Conflicts of Interest

There is no conflict of interest.

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Ethical Approval and Consent

Not available.

Review Process

Extremely peer-reviewed and accepted.

REFERENCES

1. Birman MV, McHugh JB, Hayden RJ, Jebson PJ. Pilomatrixoma of the forearm: A case report. Iowa Orthop J 2009;29:121-123.
2. Kwon D, Grekov K, Krishnan M, Dyleski R. Characteristics of pilomatrixoma in children: A review of 137 patients. Int J Pediatr Otorhinolaryngol 2014;78(8):1337-1341.
3. Pirouzmanesh A, Reinisch JF, Gonzalez-Gomez I, Smith EM, Meara JG. Pilomatrixoma: A review of 346 cases. Plast Reconstr Surg 2003;112(7):1784-1789.
4. Schwarz Y, Pitaro J, Waissbluth S, Daniel SJ. Review of pediatric head and neck pilomatrixoma. Int J Pediatr Otorhinolaryngol. 2016;85:148-153.
5. Aydoğar BE, Kaçar H, Nepesov Mİ, Arık D, Dinleyici M, Kılıç Ö. Pediatric pilomatrixoma; A case report. Osmangazi Journal of Medicine 2020;42(1):110-113.
6. Marrogi AJ, Wick MR, Dehner LP. Pilomatrical neoplasms in children and young adults. Am J Dermatopathol 1992;14(2):87-94.
7. Lemos LB, Brauchle RW. Pilomatrixoma: A diagnostic pitfall in fine-needle aspiration biopsies. A review from a small county hospital. Ann Diagn Pathol 2004;8(3):130-136.
8. Malherbe A, Chenantais J. Note sur l'epitheliome calcifie des glandes sebacees. Prog Med 1880;8:826-837.
9. Forbis R, Helwig EB. Pilomatrixoma (calcifying epithelioma). Arch Dematol 1961;83:606-617.
10. Moehlenbeck FW. Pilomatrixoma (calcifying epithelioma). A statistical study. Arch Dermatol 1973;108(4):532-534.
11. Bulut Y, Karadeniz C, Kurugöl Z, Koturoğlu G, Akşit S. An uncommon skin tumor: Pilomatricoma: Case report.Turkiye Klinikleri J Pediatr 2008;17(1):67-70.
12. Boyd AS, Martin RW 3rd. Pathologic quiz case 1. Pilomatricoma (calcified epithelioma of Malherbe) with secondary ossification. Arch Otolaryngol Head Neck Surg 1992;118(2):212-215.
13. Graham JL, Merwin CF. The tent sign of pilomatricoma. Cutis 1978;22(5):577-580.
14. Pant I, Joshi SC, Kaur G, Kumar G. Pilomatricoma as a diagnostic pitfall in clinical practice: Report of two cases and review of literature. Indian J Dermatol 2010;55(4):390-392.
15. Won S, Cho KJ, Ryu S, Seo JH, Lee JS, Park JJ. Evaluation of ultrasonography and CT for imaging of head and neck pilomatricoma in children and adolescents. Korean J Otorhinolaryngol-Head Neck Surg 2021;64(12):922-927.
16. Kato H, Kanematsu M, Watanabe H, Nagano A, Shu E, Seishima M, Miyazaki T. MR imaging findings of pilomatricomas: A radiological-pathological correlation. Acta Radiol 2016;57(6):726-732.
17. Ieni A, Todaro P, Bonanno A M, Catalano F, Catalano A, Tuccari G. Limits of fine-needle aspiration cytology in diagnosing pilomatrixoma: A series of 25 cases with clinico-pathologic correlations. Indian J Dermatol 2012;57:152-155.
18. Acer E, Tad M, Erdoğan HK. Alt ekstremite yerleşimli pilomatrikoma. Dermatoz 2016;7.