Journal of Experimental and Clinical Medicine
http://dergipark.ulakbim.gov.tr/omujecm

Case Report

J. Exp. Clin. Med., 2016; 33(1): 41-43
doi: 10.5835/jecm.omu.33.01.012

Giant pilomatrixoma of the arm: An unusual presentation in a child

Erdal Turk[a*], Irfan Karaca[a], Ragip Ortac[b]

[a] *Department of Pediatric Surgery, Faculty of Medicine, Izmir University, Izmir, Turkey*
[b] *Department of Pathology, Faculty of Medicine, Izmir University, Izmir, Turkey*

ARTICLE INFO

Article History
Received 31 / 10 / 2015
Accepted 13 / 11 / 2015

*** Correspondence to:**
Erdal Turk
Department of Pediatric Surgery,
Faculty of Medicine,
Izmir University,
Izmir, Turkey
e-mail: eturk19@yahoo.de

Keywords:
Arm
Giant pilomatrixoma
Epithelioma of Malherbe
Excision

ABSTRACT

Pilomatrixoma is a benign tumor originating from the matrix of hair follicles. It is commonly found in children and young adolescents as a slowly-growing nodule, mostly in the head and neck regions. It is relatively less common in the upper extremities. Pilomatrixomas are usually asymptomatic, solitary, hard, and mobile dermal and subcutaneous nodules and are mostly misdiagnosed despite being commonly seen in the practise. Most are smaller than 3 cm and usually around 1 cm in size while those larger than 5 cm are called pilomatrixoma and are less common. We present a seven-year-old girl with a giant pilomatrixoma on the upper arm in this study.

1. Introduction

Pilomatrixoma was first defined with the term "benign calcified epithelioma" by Malherbe and Chenantais (1880) as a calcified tumor developing from sebaceous glands. They were named pilomatrixoma by Forbis and Helwig (1961). Forty percent of pilomatrixomas arise in patients younger than 10 years of age and 60% develop within the first two decades of life. There is a 3:2 female to male incidence ratio and multiple tumors occur in 2-3% of cases (Ichikawa et al., 1997). The lesion is common in the head and neck region in 43-58% of the cases and its incidence in the upper extremities is approximately 11-23% (Pirouzmanesh et al., 2003; Hassan et al., 2013; Herrmann et al., 2014). The tumor is usually located subcutaneously. It grows slowly and emerges as a solid and mobile single mass that is generally asymptomatic (Varlıklı et al., 2013). It is actually common but is not rarely missed or misdiagnosed in cases with an atypical clinical presentation (Roche et al., 2010). The vast majority of the lesions are smaller than 3 cm with an average diameter around 1 cm; those larger than 5 cm are named giant pilomatrixoma (Lozzi et al., 2007). We present a girl with a giant pilomatrixoma of the upper arm, a rare location, who was treated with surgical excision.

2. Case report

A seven-year-old girl presented with a painless, hard mass slowly growing on the right upper arm for almost ten months. Patient history revealed no pain, infection

or trauma. A hard, mobile subcutaneous mass about 5x4 cm in size with irregular borders and occasional calcification foci was present at the mid section of the right upper arm (Fig. 1A). The skin on the mass had an ulcerated appearance. The patient was completely healthy except for the mass. Ultrasonography revealed that the mass was a hyperechogenic structure with regular borders and an intense posterior acoustic shadow while magnetic resonance imaging (MRI) showed that it was a non-enhancing calcified structure with sharp borders. The dignosis was pilomatrixoma according to clinical and radiologic findings. The mass was totally excised included the surrounding healthy tissue with a fusiform surgical incision. The surgical specimen was a 5×4×4 cm hard and yellowish-white irregular mass, calcified in its central portion (Fig. 1B). Macroscopic examination revealed a nodular and well-demarcated tumor located in the deeper layers of the dermis adjacent to subcutaneous tissue. The overlying skin was normal. Microscopically, the lesion consisted of solid islands of basaloid and eosinophilic shadow cells and extensive calcification with foreign body giant cells. The diagnosis of giant pilomatrixoma was established. (Fig. 1C and 1D). Local triamcinolone and compressive dressings were used for the patient who developed hypertrophic scar tissue in the post-operative period. The patient is now on the 18th month of follow-up without any problems.

3. Discussion

Pilomatrixoma is the second most common skin tumor of the head and neck region but it is not considered often in the differential diagnosis due to the paucity of reports in the literature (Roche et al., 2010; Varlıklı et al., 2013). It is commonly in the form of a single nodule and 1-3 cm in size, but giant pilomatrixoma cases up to 15 cm in size have been reported (Lozzi et al., 2007). Although radiologic studies are not used routinely in the common locations as the lesion is superficially located, imaging techniques such as ultrasonography, computed tomography or MRI are required in rare locations and in cases with large tumors (Varlıklı et al., 2013). Tumor in our case was a rare location and as well as larger than usual. Therefore we used radiologic imaging techniques such as ultrasonography and MRI in order not to miss other potential pathologies as well as for surgical planning.

Spontaneous regression of pilomatrixoma has never been observed and its malignant transformation is quite rare (Roche et al., 2010). Its treatment is total excision together with the ulcerated skin on the mass. Recurrence may develop in case of inadequate excision. It is recommended that the excision include 1-2 cm of the healthy tissue to prevent recurrence (Varlıklı et al., 2013). The excision of a pilomatrixoma is usually easy, but malignancy should be suspected if the mass is adherent to the surrounding tissues or if its borders are

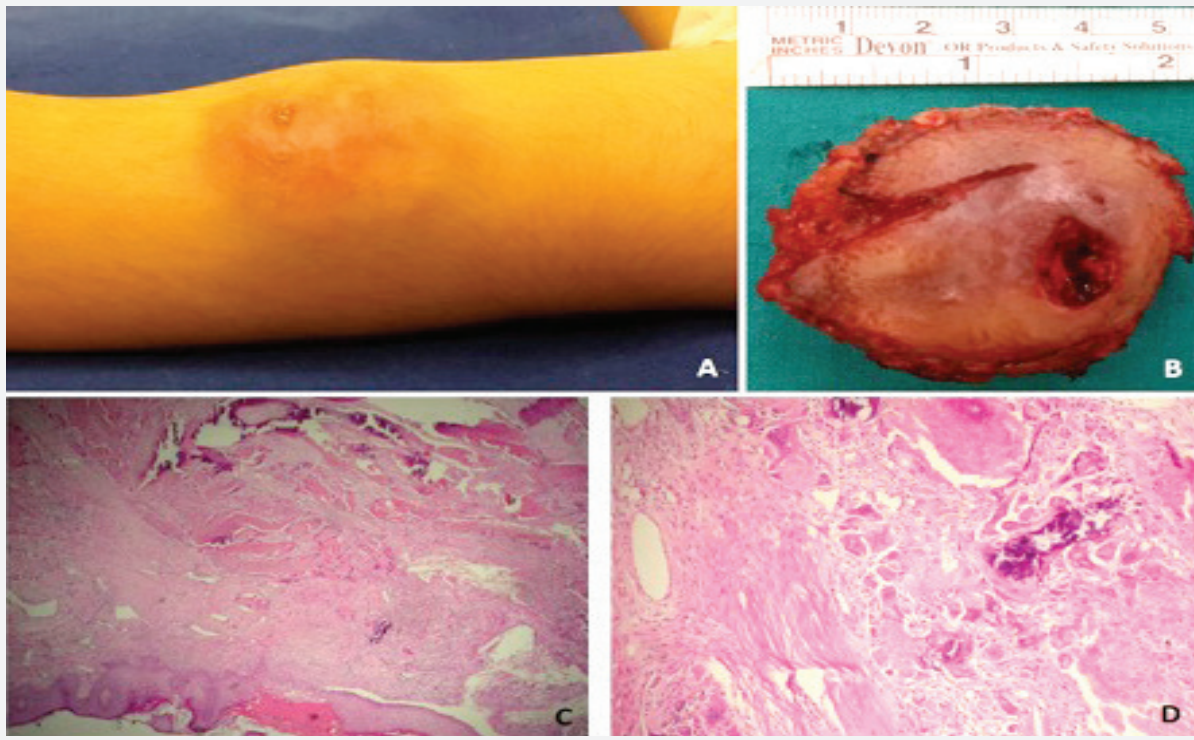

Fig. 1A. Pre-operative appearance of the tumor; **1B.** Appearance after excision; **1C.** Tumor tissue with predominant well-limited ghost cells in the subcutaneous tissue (HE; 40x); **1D.** Calcification and foreign body giant cells accompanying tumor tissue (HE; 200x)

not well defined (Pirouzmanesh et al., 2003; Varlıklı et al., 2013). The mass of our patient was on the left upper arm. Its borders were regular, it could be differentiated easily from the neighboring tissues and it was covered with ulcerated tissue. The excision of the mass was quite easy, but its large dimensions caused a large skin defect afterwards. Primary closure of the skin was therefore performed under tension. It should be kept in mind that the post-excision defect may be large in these patients and a graft or flap may be required.

The differential diagnosis of pilomatrixoma varies according to the localization. Epidermoid and dermoid cysts, calcified lymph nodes and calcified hematoma, hemangioma and parotid gland tumors should be considered in the head and neck region. Parotid tumors, sebaceous cyst, calcified hematoma, branchial remains and giant cell tumors should be considered in the preauricular region, and epidermal inclusion cyst, lymphadenopathy, and foreign body reaction lipoma should be considered in other regions (Roche et al., 2010; Hassan et al., 2013; Varlıklı et al., 2013). We primarily considered sebaceous, dermoid and epidermoid cysts, metaplastic bone formation, foreign body reaction, hematoma, osteochondroma, trichoepithelioma, basal cell epithelioma and other benign and malignant soft tissue tumors in our patient due to the location on the left upper arm.

In conclusion, pilomatrixoma is a quite common tumor in the head and neck, especially in childhood and adolescence, but can also be seen in the extremities. Clinical presentation is typical in most cases and the diagnosis is made easily. However, the diagnosis is notalways easy in lesion with an unusual location and size, and radiologic investigations should be conducted in such cases. The definite diagnosis is made with histopathologic methods. Management of pilomatrixomas typically involves marginal excision to prevent recurrence.

REFERENCES

Forbis, R., Helwing, E.B., 1961. Pilomatrixoma (calcifying epithelioma). Arch. Dermatol. 83, 606-608.

Hassan, S.F., Stephens, E., Fallon, S.C., Schady, D., Hicks, M.J., Lopez, M.E., Lazar, D.A., Rodriguez, M.A., Brandt, M.L., 2013. Characterizing pilomatricomas in children: A single institution experience. J. Pediatr. Surg.. 48, 1551-1556.

Herrmann, J.L., Allan, A., Trapp, K.M., Morgan, M.B., 2014. Pilomatrix carcinoma: 13 new cases and review of the literature with emphasis on predictors of metastasis. J. Am. Acad. Dermatol. 71, 38-43.

Ichikawa, T., Nakajima, Y., Fujimoto, H., Koyama, A., Honma, M., Yatsuzuka, M., Ohtomo, K., Uchiyama, G., Ushigome, S., Ohba, S., 1997. Giant calcifying epithelioma of Malherbe (pilomatrixoma): Imaging features. Skeletal Radiol. 26, 602-605.

Lozzi, G.P., Soyer, H.P., Fruehauf, J., Massone, C., Kerl, H., Peris, K., 2007. Giant pilomatricoma. Am. J. Dermatopathol. 29, 286-289.

Malherbe, A., Chenantais, J., 1880. Note Sur L'epitheliome calcifie des glades sebacees. Programme Med. 8, 826-828.

Pirouzmanesh, A., Reinisch, J.F., Gonzalez-Gomez, I., Smith, E.M., Meara, J.G., 2003. Pilomatrixoma: A review of 346 cases. Plast. Reconstr. Surg. 112, 1784-1789.

Roche, N.A., Monstrey, S.J., Matton, G.E., 2010. Pilomatricoma in children: Common but often misdiagnosed. Acta. Chirurgica. Belgica. 110, 250-254.

Varlıklı, O., Yıldız, T., Çetin, G., Yıldırım, M., Erdem, MT., 2013. Pilomatrixoma in Children (Calcifying Epithelioma). Türkderm. 47, 84-87.