Giant pilomatricoma of the arm: An unusual presentation (A case report)

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Abstract Pilomatricoma is a benign tumor of the hair matrix. It is most often seen in children and young adults as a slowly growing nodule in the skin of the head and neck. The frequency of presentation of this lesion in the upper extremities is relatively rare. Pilomatricoma is a frequently misdiagnosed entity in clinical practice. They are usually asymptomatic, solitary, firm or hard, freely mobile, dermal or subcutaneous nodules. Most tumors are smaller than 3 cm in diameter. We report a 32-year-old woman with a giant pilomatricoma of the arm. Surgical excision was curative without recurrence. In this paper, we discuss the clinical, histopathologic characteristics of pilomatricoma and review the literature regarding pilomatrixomas in the upper extremity.

1. Introduction
Pilomatrixomas are of ectodermal origin and arise from the outer root sheath cell of the hair follicle (Rook et al., 1998). It is most often seen in children and the most common anatomical location is the head and neck region (Pirouzmanesh et al., 2003). Involvement of the upper extremities is relatively uncommon (Pirouzmanesh et al., 2003; Daoudi et al., 2006; Ioannidis et al., 2010). Pilomatrixomas usually are asymptomatic, deeply seated, firm, nontender subcutaneous masses adherent to the skin but not fixed to the underlying tissue (Rook et al., 1998). Most tumors are smaller than 3 cm in diameter. Pilomatricoma prognosis is generally good. Cure without recurrence is the rule after total surgical resection. We present a case of an unusual presentation of giant pilomatrixoma in the arm of a 32 year old woman.

2. Case report
A 32-year-old woman with an unremarkable medical history, presented with a nodular mass on the left arm. The patient had noted the mass in her left arm, one year before initial presentation. The lesion had begun growing over the last few months. There was no history of pain, infection or trauma. Clinical examination revealed a sharply outlined subcutaneous mass, which was calcified in its central portion. MRI reveals a rim-enhancing lesion with small areas of signal dropout which...
The tumor was removed with a fusiform surgical excision. The surgical specimen revealed a hard irregular mass, calcified in its central portion, measuring 6·6·4 cm, and was yellowish-white in color (Fig. 3). Macroscopic examination revealed a nodular tumor located in the deeper layers of the dermis adjacent to the subcutaneous tissue. The overlying skin was healthy. Microscopically it consisted of solid islands of shadow cells and extensive ossification. Even the usual nests of small basaloid cells were absent. The diagnosis of giant pilomatricoma was established. Follow-up at 3 years revealed no evidence of recurrence.

3. Literature review

Pilomatrixoma, pilomatricoma or calcifying epithelioma of Malherbe is a benign neoplasm derived from hair follicle matrix cells (Rook et al., 1998). These lesions are typically found in the head and neck region (Pirouzmanesh et al., 2003). Only a few isolated locations at exceptional members as the case of our patient have been reported in the literature (Pirouzmanesh et al., 2003; Daoudi et al., 2006; Ioannidis et al., 2010). Despite the frequency of presentation of this lesion in the upper extremities, discussion of this lesion is essentially limited to the literature of otolaryngology, pathology, and dermatology (Silva et al., 2003; Khammash et al., 2001). A search of the literature revealed few well-documented cases of pilomatrixomas in the arm (Daoudi et al., 2006; Ioannidis et al., 2010). Since the first description of pilomatrixoma, there has been a gradual increase in understanding of the morphologic features and clinical presentation of this tumor (Kaddu et al., 1994). However, difficulties still persist in making clinical and cytologic diagnosis (Kaddu et al., 1994). Calcification of the tumor was observed in 80% of cases, sometimes achieving a true osteoma subcutaneous (Daoudi et al., 2006; Ioannidis et al., 2010). The usual size is less than 3 cm, however, few cases of giant pilomaricoma than 5 cm in diameter were observed as in our case (Percin et al., 1996; Khammash et al., 2001). Pilomatrixoma presents most commonly in children and young adults, and they are noted more commonly in females (Pirouzmanesh et al., 2003). It occurs mainly during the second decade of life and rarely beyond (Pirouzmanesh et al., 2003). Our patient is 32 years old which is an unusual age of onset. Diagnostic imaging is generally not obtained in the evaluation of pilomatrixomas as they are usually superficial, small, and well-circumscribed. Conventional radiography in the pilomatrixomas may demonstrate foci of calcification (Daoudi et al., 2006; Ioannidis et al., 2010; Haller et al., 1977). Ultrasound demonstrates a well-defined mass with inner echogenic foci and a peripheral hypoechoic rim or a completely echogenic mass with strong posterior or acoustic shadowing in the subcutaneous layer (Hughes et al., 1999; Hwang et al., 2005). Computed tomography (CT) demonstrates a sharply demarcated, subcutaneous lesion of soft tissue density, with or without calcification (Agarwal et al., 2001). MRI may reveal a rim-enhancing lesion with small areas of signal dropout.
which may be consistent with calcifications as is the case in our patient (Agarwal et al., 2001). In all cases histology confirmed the diagnosis (Heenan et al., 1996). It is important to note that the pilomatrixoma was incorrectly diagnosed by fine needle aspiration cytology (Wang et al., 2002). The microscopic appearance of pilomatrixomas is characterized by irregularly shaped well-circumscribed masses of epithelial cells. Basophilic cells and shadow cells may be observed in the cellular stroma (Rook et al., 1998; Pirouzmanesh et al., 2003; Heenan et al., 1996). The differential diagnosis of these lesions should include sebaceous, dermoid and epidermoid cysts, metaplastic bone formation, foreign body reaction, parotid gland tumor, hematomata, osteochondroma, trichoeithelioma and basal cell epithelioma (Pirouzmanesh et al., 2003; Kaddu et al., 1996).

Recommended management is surgical excision (Pirouzmanesh et al., 2003). Recurrence is uncommon after adequate excision (Pirouzmanesh et al., 2003; Khammash et al., 2001). All patients need dermatological evaluation with close long term follow-up. The clinical course is generally benign although, malignant transformations have been reported (Pirouzmanesh et al., 2003; Mikhaeel and Spittle, 2001).

Pilomatrixoma is a rare benign neoplasm derived from hair follicle matrix cells. Involvement of the arm is relatively uncommon. And, most tumors are smaller than 3 cm in diameter. In this patient’s case, the definitive diagnosis was made only after histologic examination following excision of the mass. Management of pilomatrixomas typically involves marginal excision to prevent recurrence.

References


