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CASE REPORT

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Atypical adult non-calcified pilomatricoma

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Abstract We present a rare case of a non-calcified pilomatricoma in a 67-year-old man. This tumor was extremely large in size, and its location, in the lower extremity, was very unusual. The clinical, radiographic, and histopathological features are described in detail. The role of magnetic resonance imaging (MRI) in the diagnosis of this entity is discussed. Definite internal reticulations and septations were observed. A possible explanation for this observation is that the high signal intensity reticulations may represent edematous stroma surrounding basaloid cells.

Key words MRI · Neoplasms, MRI · Soft tissues, MRI · Soft tissue neoplasms · Skin, neoplasms · MRI, pilomatricoma (histology)

Introduction

Pilomatricoma is an unusual benign cutaneous tumor, originating from precursor cells that normally differentiate into hair matrix cells. It has also been described using the term "pilomatrixoma" to signify its origin. The majority of pilomatricomas are seen in the first two decades of life, and they are extremely rare in older patients. Most present as firm, solid nodules in the head and neck region [1]. We present a case of a large pilomatricoma of the lower extremity in an older adult with unusual characteristics.

Case report

A 67-year-old man presented with a large mass on the medial aspect of his left thigh, which had been increasing in size for the previous 18 months. There was no previous medical history of malignancy. However, small lesions may have been present in the region of the mass as long as 7 years prior to presentation. The patient presented to the emergency department with a fungating, foul smelling, pedunculated mass. He stated that the mass bled with trauma, prompting a visit to the hospital. Initial radiographs demonstrated a large soft tissue mass in the region of the medial aspect of left thigh. No calcifications were present.

Magnetic resonance imaging (MRI) was performed to evaluate the mass and to determine possible muscle invasion (Fig. 1). This study revealed an exophytic 11×5-cm mass along the medial left thigh with a 7×9-cm base. The mass had an inhomogeneous appearance, without evidence of a definite capsule. The superomedial edge was irregular due to surface ulceration. Sagittal spin echo (SE) T1-weighted pre-gadolinium images demonstrated a large inhomogeneous mass, isointense relative to muscle, without any specific signal characteristics. Pre-gadolinium axial T2-weighted and T1-weighted post-gadolinium fat-saturation images demonstrated bands of high signal intensity radiating to the periphery, giving the appearance of internal reticulations and septations. No definite enhancement was observed after intravenous administration of gadolinium. There was no invasion into the muscle compartment.

The tumor was excised. It weighed 440 g, and consisted of a circumscribed dermal and subcutaneous polypoid mass. The cut surface appeared variegated with lobulated chalky-white areas admixed with gray and tan areas. Several sur-

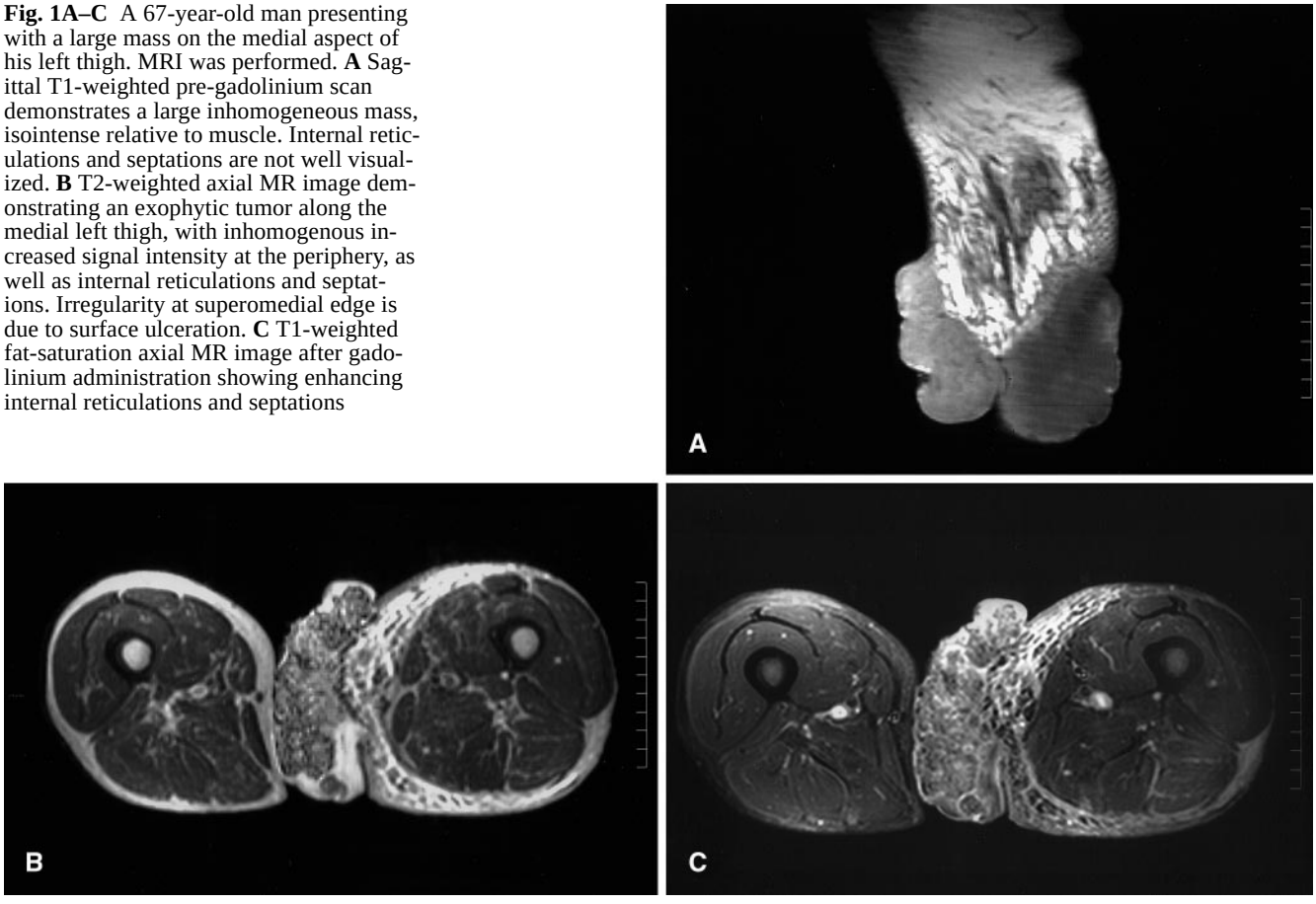

Fig. 1A–C A 67-year-old man presenting with a large mass on the medial aspect of his left thigh. MRI was performed. **A** Sagittal T1-weighted pre-gadolinium scan demonstrates a large inhomogeneous mass, isointense relative to muscle. Internal reticulations and septations are not well visualized. **B** T2-weighted axial MR image demonstrating an exophytic tumor along the medial left thigh, with inhomogenous increased signal intensity at the periphery, as well as internal reticulations and septations. Irregularity at superomedial edge is due to surface ulceration. **C** T1-weighted fat-saturation axial MR image after gadolinium administration showing enhancing internal reticulations and septations

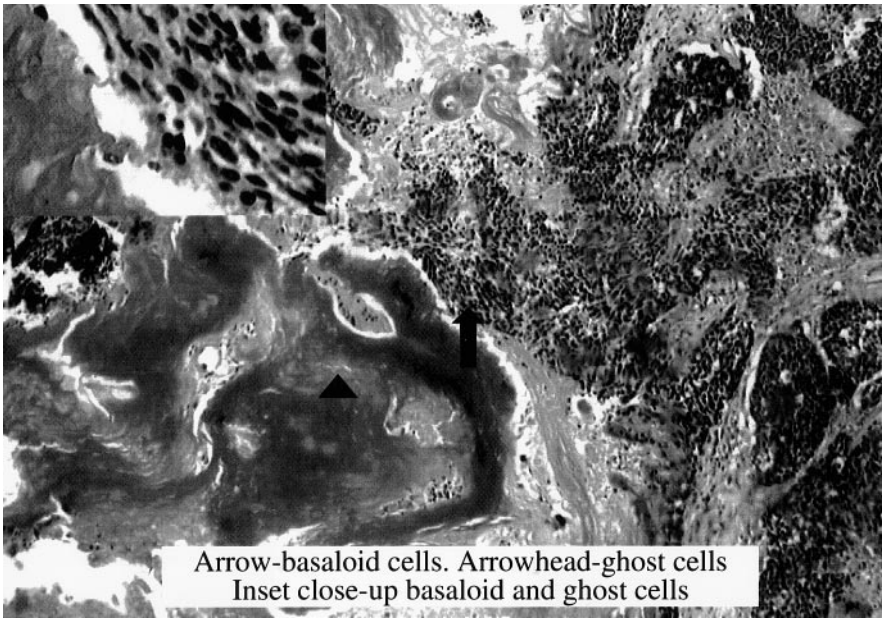

Fig. 2 Photomicrograph of the resected specimen shows a large keratin accumulation, peripheral basaloid cells, and several centrally located shadow ("ghost") cells, which are typical for pilomatricoma (H&E ×100; courtesy of Dr. Kittane Ranganath)

face ulcers were present. Initial histologic examination demonstrated basaloid cells with numerous "ghost" cells in a fibrocollagenous matrix with an inflammatory background. Numerous foci of keratin were evident, with a multinucleated giant cell reaction to this material. No invasion of the adjacent subcutaneous tissues was present, and there were no signs of malignancy (Fig. 2). These findings are consistent with the diagnosis of pilomatricoma.

Discussion

Pilomatricoma is somewhat unusual in adults. Behenke et al. reported four elderly patients, with histologically proven pilomatricomas [2]. The lesion is, however, common in childhood, and between 60 and 70% of the cases occur in the first two decades of life [3]. In this group, there is a female to male ratio of 2 to 1. Most occur in the head, neck, and chest regions. The remainder occur on the upper extremities, with extremely rare occurrences in the lower extremities, making this a very unusual case presentation. The typical pilomatricoma is a firm, solitary, calcified, dermal nodule. The majority are between 1.0 and 1.5 cm in size [4], as opposed to our case, which was much larger. In rare instances, multiple pilomatricomas are associated with myotonic dystrophy. When present, familial incidence suggests that multiple pilomatricomas occur due to a pietropic effect of the myotonic dystrophy gene [5]. Pilomatrix carcinoma is a somewhat subjective diagnosis. The usual criteria are high mitotic activity, with crowded, atypical, pleomorphic basaloid cells demonstrating locally aggressive behavior [6]. Fewer than 20 cases of pilomatrical carcinoma have been reported in the literature, mostly occurring in the older population. Distant metastases are rare [4].

Histologically, the appearance of this tumor was typical for benign pilomatricoma. Epithelial cells with basophilic cytoplasm are seen at the periphery of these tumors, arranged in an arc-like fashion [6]. In 30%, basaloid cells are transformed into shadow cells, also known as "ghost" cells, which are more centrally located. These cells have lost their nuclei and are filled with keratin and stain eosinophilic. The unstained area in the center of the ghost cell appears as a shadow due to the lost nucleus [6]. Foreign body giant cells are not unusual.

The tumor that we describe is unusual in several respects. First, it was quite large, and to our knowledge, is one of the largest ever reported [7]. Second, the location of the tumor is atypical. Third, the age of our patient was significantly older than the typical younger age of presentation, although Taaffe et al. have described a second, smaller age peak in the sixth decade of life [8]. Finally, no calcifications were demonstrated on either the radiographs or histologically. This presentation is unusual, as 80% of these tumors are known to be calcified [1].

A recent article by Hoffman et al. described the MRI characteristics of a pilomatricoma in the neck of a child [3]. T2-weighted fat-suppressed images showed bands of hyperintense signal radiating away from a lower signal intensity center towards the periphery. With gadolinium administration, there was enhancement of the periphery, but no enhancement was noted in the center of the lesion. The authors raised the possibility that the basaloid bands present on histologic specimens may correspond to the hyperintense bands seen on MRI. De Beuckeleer et al. described MRI findings in pilomatricoma with homogeneous intermediate signal intensity on T1-weighted images and inhomogeneous intermediate signal intensity on gradient echo and T2-weighted images [1]. Finally, Ichikawa et al. noted the same findings on T1-weighted and T2*-weighted images [7]. Amorphous calcification accounted for the inhomogeneous appearance. Our case suggests that pilomatricoma indeed does have internal reticulations and septations, which are equally well seen on T2-weighted and post-gadolinium T1-weighted fat-saturation images. This patient's tumor contained no calcifications, which was very unusual. Since the gross specimen is not available to us for precise radiologic-pathologic correlation, we are unable to correlate various signal intensities with underlying tissue histology. Hoffman suggested that internal reticulations noted on T2-weighted and post-gadolinium T1-weighted images represent basaloid cells. Another possible explanation is that the high signal intensity reticulations represent surrounding edematous stroma rather than basaloid cells, as the basaloid cells are sheets of avascular epithelial cells.

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