CT of pilomatrixoma in the cheek.

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[http://www.ajnr.org/content/19/7/1219](http://www.ajnr.org/content/19/7/1219)

This information is current as of November 13, 2023.
Summary: Pilomatrixoma is an uncommon benign tumor arising from hair follicles. They occur most commonly in the head and neck region, and are usually found in girls during the first two decades of life. These tumors may contain calcification, which, when present, is helpful in suggesting the diagnosis. We present a classic case of pilomatrixoma in the cheek of a young woman. The tumor was documented on CT studies, which showed a subcutaneous, noninvasive mass with calcifications.

Pilomatrixomas are uncommon benign neoplasms of hair follicle origin that were first described by Melherbe and Chantantois in 1880 (1). In 1961, it was Forbes and Helwig who first suggested the name pilomatrixoma (1, 2). These tumors are uncommon neoplasms that favor the hair-bearing areas of the head and neck, with approximately half the reported tumors occurring in the head and neck region (3). The majority of these tumors arise during the first two decades of life; however, a second peak of occurrence is seen in older patients (4). Overall, there is a female predominance. Calcium deposition can occur within the tumors and, when present, has been helpful in suggesting the diagnosis at fine-needle aspiration cytology (5). To our knowledge, only two prior reports of this tumor have been documented with CT. In one case, a noncalcified tumor was seen in the parotid region. Because the tumor was inseparable from the parotid gland on CT scans, the imaging diagnosis was pleomorphic adenoma (1). In the second case, multiple giant pilomatrix carcinomas, malignant counterparts of pilomatrixomas, were seen on CT scans in the posterior part of the neck and calvaria (6).

The purpose of this report is to document the CT findings in a case of pilomatrixoma with calcifications that occurred within a well-defined subcutaneous cheek mass in a young woman. Because this appearance reflects the majority of clinical and pathologic reports of these tumors, the presence of these CT findings should favor the addition of pilomatrixoma to the imaging differential diagnosis (1–12).

Case Report

A 21-year-old woman had an 8-month history of a painless mass of the left cheek. On physical examination, a firm, 2.5-cm mass was seen 5-cm dorsal to the angle of the mouth. The lesion was freely mobile with respect to the subcutaneous fat, but was tethered to the dermis. The clinical diagnosis of a pilomatrixoma was suggested because none of the other lesions in the clinical differential diagnosis present with tethering to the dermis. A CT study was performed to ascertain the presence of any other subclinical disease and to provide possible supporting evidence for the suspected diagnosis. The CT scans showed a noninfiltrating mass in the subcutaneous fat of the left cheek (Fig 1). The mass could not be separated from the overlying skin, and the lesion did not extend into the underlying facial musculature. Located within the caudal half of the mass were multiple calcifications. The initial imaging diagnosis was consistent with a pilomatrixoma. The differential diagnosis included a sebaceous cyst, a foreign body reaction, or an unusual tumor.

At surgery, with the patient under general anesthesia, the tumor was removed together with an ellipse of overlying skin. The lesion did not involve the facial musculature and it was easily dissected free from the subcutaneous fat. The postoperative course was uneventful and the pathologic report was pilomatrixoma.

Discussion

Pilomatrixoma, or calcifying epithelioma of Melherbe, is a rare benign tumor of hair-follicle origin that usually arises in the head and neck region, most commonly in the first two decades of life (7). At histopathologic analysis, one sees matrixlike basaloid hair cells and shadow or ghost cells, which have a central unstained area that represents the shadow of a lost nucleus. Intracellular and stromal calcification is reported in about 70% of cases, and tumors with large areas or nests of shadow cells proportionally have increased calcium deposition (1). It has been suggested that osteopontin (a protein marker associated with bone production) may be produced by macrophages and play a role in the deposition of calcium phosphate in the shadow cell nests (8).

The clinical presentation is typically that of an asymptomatic, superficial, solitary, firm mass that is often accompanied by a reddish-blue discoloration of the overlying skin. These tumors usually increase...
slowly in size and are located in the head and neck (78% of cases) or chest and upper limbs (1, 4). Pathologically, the tumors are situated in the dermis or subcutaneous tissue, and complete surgical excision is curative (8, 9). Multiple tumors account for 2% to 3.5% of reported cases; in rare instances they are associated with myotonic muscular dystrophy, Gardner syndrome, and skull dysostosis. A rare familial occurrence has also been reported (1).

Fine-needle aspiration biopsy has produced either clustered or isolated basaloid cells with variably sized nuclei and prominent nucleoli. Multinucleated giant cells and either ghost cells or acellular masses are suggestive of ghost cell nests. When present, calcium deposition adjacent to basaloid cells also contributes to the correct diagnosis (5, 10).

One previous report included MR imaging findings in pilomatrixomas, which included homogeneous intermediate T1-weighted signal intensity, low-to-intermediate T2-weighted signal intensity, and no enhancement. The lesion was also hyperechoic at sonography (11).

The differential diagnosis includes sebaceous cyst, ossifying hematomata, giant cell tumor, chondroma, dermoid cyst, foreign body reaction, degenerating fibroxanthoma, metastatic bone formation, and osteoma cutis (1). The rare, malignant counterpart of pilomatrixoma is pilomatrix carcinoma, and the differential diagnosis of this malignant lesion includes basal cell carcinoma with matrix differentiation and metastasis (6).

Conclusion

The imaging finding of a noninfiltrating mass containing calcifications located within the subcutaneous tissues of the head and neck should raise the possibility of a pilomatrixoma, especially if such a mass occurs in a young woman who is in the first two to three decades of life.

References