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Granular-cell tumor of the anterior abdominal wall

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We report a case of granular-cell tumor (GCT) arising in the subcutaneous tissue of the abdominal wall and describe its radiologic and histologic characteristics. The differential diagnosis of a mass in this site may include multiple benign and malignant stromal lesions. In this case, the presentation, location, and radiological features suggested a desmoid tumor (aggressive fibromatosis). Treatment of the mass involved surgical excision with negative margins, and histological analysis confirmed the presence of a benign GCT. We report a case of this rare, benign tumor to allow the radiologist and pathologist to consider this disease in the differential diagnosis when presented with similar cases.

Case report

A 49-year-old Caucasian female presented with a 16-month history of a nontender, palpable mass in the left upper quadrant of her abdomen. The patient denied any history of trauma. The mass had not changed significantly in size during this time and had remained otherwise asymptomatic. The patient had no family or personal history of colon polyposis, and there was no history of any other significant illnesses.

Palpation of the left upper quadrant about one finger-breadth below the subcostal margin revealed a well-circumscribed, nontender, mobile, firm mass measuring approximately 2 x 3 cm. No tenderness was elicited on palpation of her abdomen, and there was no hepatosplenomegaly or lymphadenopathy. The rest of the examination was within normal limits.

Ultrasound examination revealed an irregular soft-tissue mass in the left subcostal subcutaneous tissue, measuring 2.2 cm in greatest dimension (Fig. 1). A chest-wall MRI

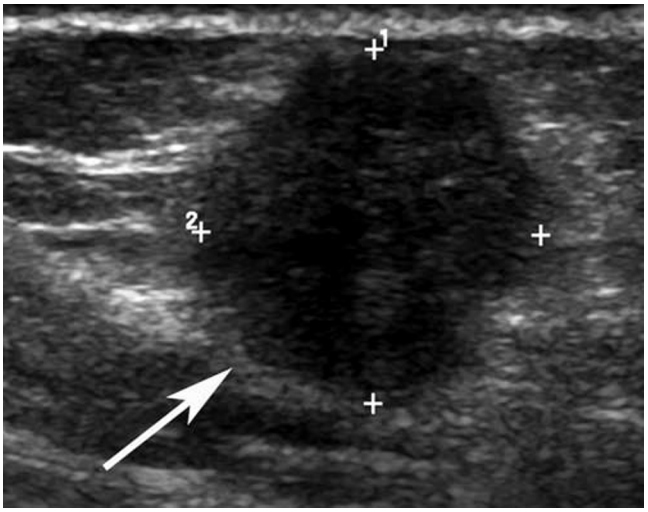

Figure 1. 49-year-old female with granular-cell tumor of the abdominal wall. Ultrasonography of left upper quadrant subcutaneous fat demonstrating a 2.2 cm, slightly ill-marginated hypoechoic mass in the subcutaneous tissue.

revealed a nonspecific enhancing soft-tissue mass within the subcutaneous fat in the same site, measuring up to 2.8 cm in greatest dimension (Fig. 2). The mass exhibited intermediate T1 signal and slightly increased T2 signal (with no fat contained within the lesion). Although the mass was predominantly well-circumscribed, there were spiculated margins anteriorly concerning for possible early invasion. The mass abutted the anterior fascia of the rectus muscle but

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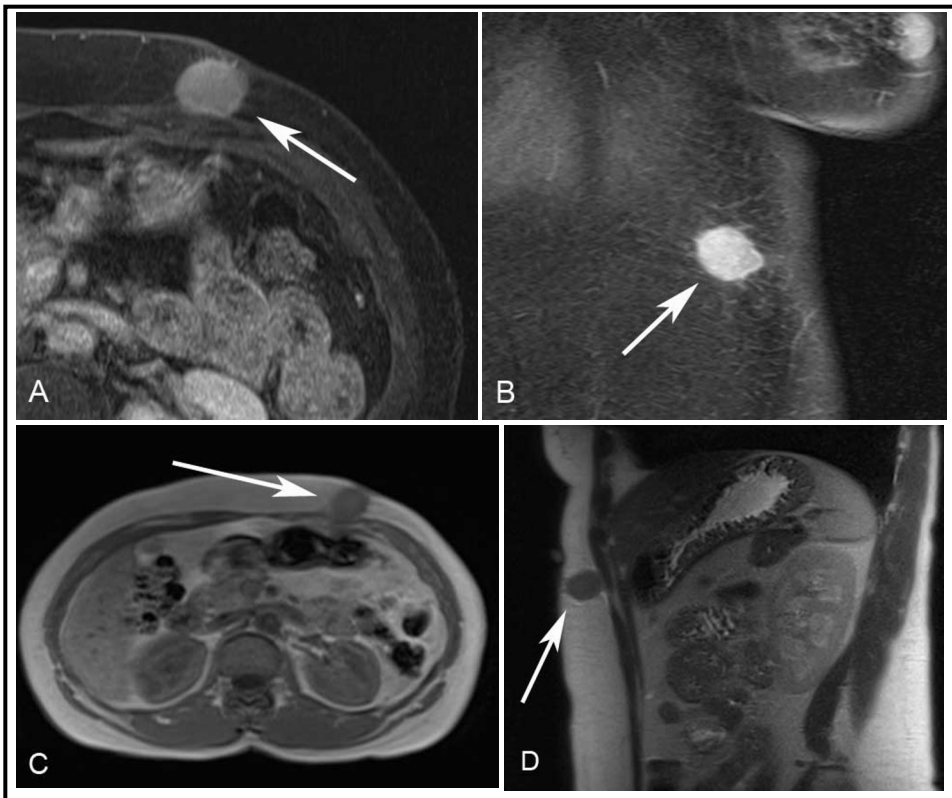

Figure 2. 49-year-old female with granular-cell tumor of the abdominal wall. Magnetic resonance images (A, T1, axial, fat-suppressed, post-IV contrast; B, T1, coronal, fat-suppressed, post-IV contrast; C, T1, axial precontrast; D, T2, sagittal) showing the enhancing soft-tissue mass in left upper quadrant subcutaneous fat (dimensions, 2.2 x 2.2 x 2.8 cm).

did not invade into muscle. The radiographic appearances were consistent with possible soft-tissue sarcoma or desmoid tumor. The visualized intra-abdominal structures were unremarkable.

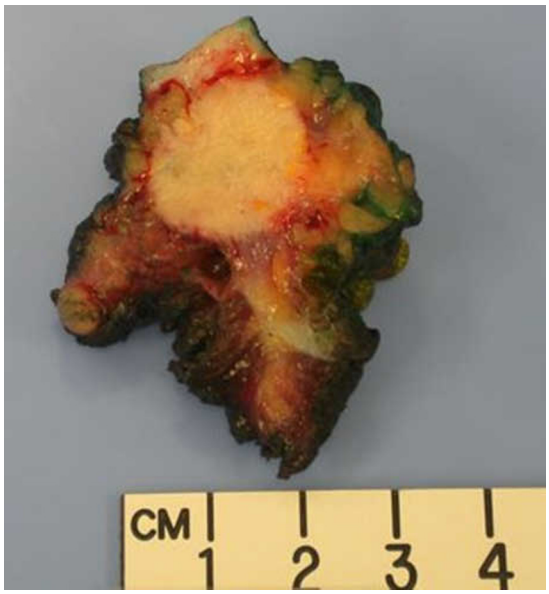

Figure 3. 49-year-old female with granular-cell tumor of the abdominal wall. Gross appearance of the granular-cell tumor with cut surface, showing a relatively well-demarcated, solid pale yellow mass.

The patient underwent wide local excision of the mass, during which an elliptical incision (incorporating skin) was made. Dissection was carried down to the anterior fascia. The lesion was adherent to the anterior rectus fascia, and so an extended area around the lesion in the fascia was excised (including a rim of rectus muscle to constitute the posterior margin). All margins were negative on gross examination, with the closest margin 5 mm from the mass to the cut edge (Fig. 3). On frozen-section examination, the lesion was consistent with granular-cell tumor. The tumor measured 2.1 cm in greatest dimension, and was composed of large polygonal cells with granular-appearing cytoplasm, separated by fibrotic tissue (Fig. 4). The granular cytoplasm stained strongly for S-100 protein (Fig. 5). The mass was completely excised with uninvolved margins and showed no atypical or malignant characteristics.

Discussion

Granular-cell tumors (GCT) are relatively uncommon tumors, first described in the skeletal muscle of the tongue by Abrikossoff in 1926 (1). Initially termed "myoblastic myomata" to describe their possible muscular origin, it was later noted that tumors were often located adjacent to peripheral nerves, implying a neural origin. Although their histogenesis is still uncertain, they are now accepted by most to originate in Schwann cells. They can occur at almost any site, although most occur in the head and neck (with the tongue being the most commonly affected site [2]), and skin and subcutaneous tissues of the chest and upper extremities. There have also been case reports of

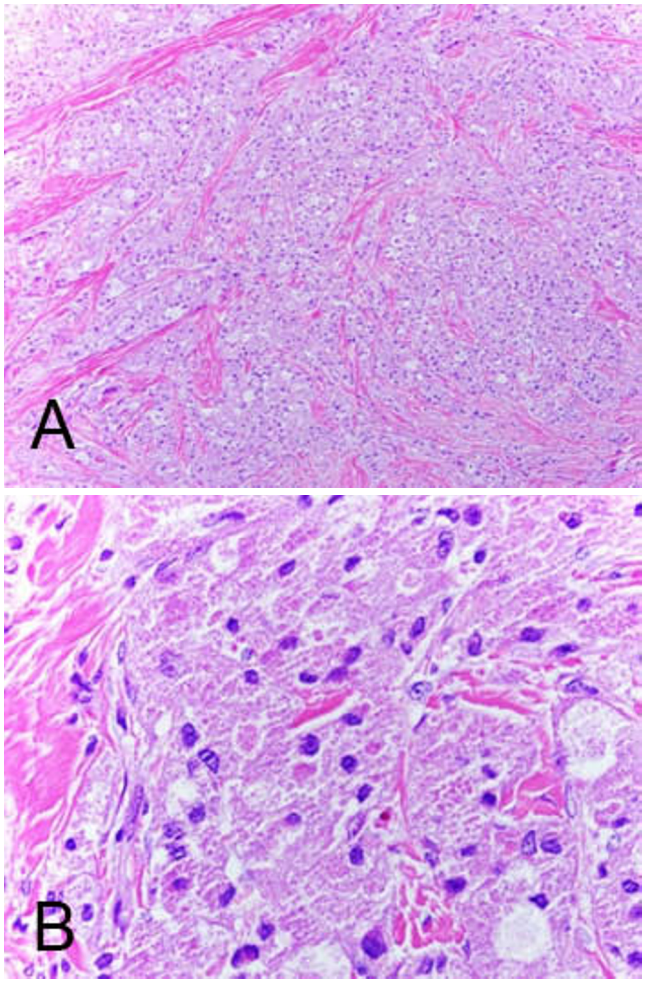

Figure 4. 49-year-old female with granular-cell tumor of the abdominal wall. Hematoxylin and eosin stain showing granular-cell tumor composed of large polygonal cells with granular cytoplasm, separated by slender fibrous tissue (A, x 100 magnification; B, x 600 magnification).

GCTs arising in the breast (3), stomach (4, 5), esophagus (6), pancreas (7), large bowel (1, 8-11), thyroid (12, 13), bronchus (14), abdominal wall (15), and kidney (16).

The presentation of a painless solid mass adjacent to the rectus sheath in a young woman is a common presentation of a desmoid tumor (aggressive fibromatosis); its coalescence with the adjacent aponeurosis radiologically also pointed toward this entity. The use of an intra-operative frozen section in this case was helpful, avoiding a potentially more extensive wide excision for a presumed sarcoma or desmoid tumor.

The usual presentation of a GCT is a painless, slowly growing mass. Patients are usually in their second to fourth decades, with a female predilection. The histological features should be distinguished from rhabdomyoma, hibernoma, oncocytoma, schwannoma, extra-gastrointestinal stromal tumor, and non-neoplastic reactive changes associated with injury and trauma. Excisional biopsy is the mainstay for a pathological diagnosis. The tumors are defined histologically by large polygonal cells with an abundant eosinophilic granular cytoplasm and relatively small nucleus. Tumor cells are typically arranged in cords or nests

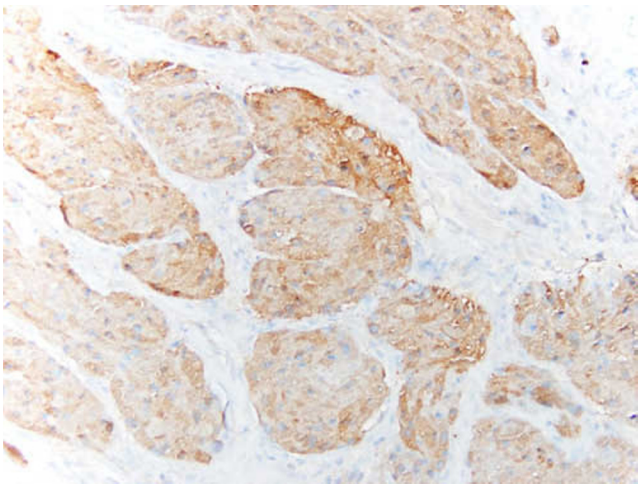

Figure 5. 49-year-old female with granular-cell tumor of the abdominal wall. The granular cytoplasm expresses S-100 protein (S-100 immunohistochemical stain, x200).

separated by fibrous connective tissue, and characteristically express CD68, neuron-specific enolase, and S-100 protein. There have been rare reports of cases in which the tumor is locally aggressive or malignant with distant metastases (17-19).

Treatment is complete resection of the mass. There is no role for medical management. Prognosis is good, with a low recurrence rate. As there are very few true malignant cases reported, diagnostic criteria and a standard strategy of treatment for such rare entities is lacking (17).

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