




Subcutaneous Granular Cell Tumor of the Buttock: An Unusual Case and Brief Literature Review

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Abstract

Granular cell tumors are rare soft tissue tumors with neural origin that present malignant potential. Perianal granular cell tumors with only subcutaneous involvement are uncommonly presented in the literature. Herein, we present the case of a 49-year-old female patient with perianal granular cell tumor of the buttock. The patient underwent successful surgical resection and remains under close postoperative follow-up. There is no standard diagnostic algorithm for these tumors, and imaging provides little benefit. Histopathology and immunohistochemical staining are necessary for the accurate diagnosis of granular cell tumors.

Keywords

- ▶ granular cell tumor
- ▶ oncology
- ▶ colon and rectal surgery

Introduction

Granular cell tumors (GCTs) are benign slow-growing soft tissue tumors of neural crest cell origin, with ~ 1% to 2% of cases reported as malignant.¹ Most GCTs are benign and present excellent outcomes after surgical resection, while malignant GCTs are aggressive and have a poor prognosis, with a high rate of metastasis and recurrence.² These tumors are thought to be derived from neural crest cells, specifically Schwann cells. This hypothesis is based primarily on the similarities to between GCTs and schwannomas in regard to their immunohistochemical profile for biomarkers of neural crest cell and neuroectodermal origin, including GAP43, S100, NSE, and SOX10.³ Granular cell tumors are more prevalent among women when compared with men (in a proportion of 3:1), and they typically occur between the second and sixth decades of life.^{4,5} These tumors most commonly arises in the skin, subcutaneous and soft tissues, the oral cavity, and the gastrointestinal tract.⁵ The most common locations of occurrence are the head and neck (70%), with ~ 10% arising in the gastrointestinal tract.⁴⁻⁶ The esophagus is the most common location within the gastro-

intestinal tract, while the rectum is the least affected.¹ Literature about perianal GCTs localized to the subcutaneous tissue is sparse, with relatively no reported cases arising from the gluteal/buttock region. Herein, we report a rare case of a benign GCT of the left buttock in a healthy 41-year-old female patient.

Case Presentation

A 41-year-old female patient with a medical history of gastrointestinal reflux disease (GERD) presented to the colorectal surgical clinic complaining of a pruritic, uncomfortable left buttock lesion that had been present for 8 years. Other than localized discomfort, the patient denied blood per rectum, weight loss, skin ulceration/bleeding to her buttock, or any personal/family history of colon cancer/inflammatory bowel disease. The physical examination revealed a firm left buttock lesion measuring 2 cm x 2 cm, without skin changes or drainage. Evaluation via digital rectal exam and anoscopy confirmed no fistulous tracts or synchronous lesions in the anus. Due to the progression of her symptoms, the decision was to proceed with surgical treatment.

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A soft tissue mass measuring 4 cm x 2 cm was excised from the left buttock without any complications. The surgical pathology review of the excised mass reported the skin excision to be a GCT spanning ~ 2.5 cm and involving the dermis and subcutaneous tissue, extending to one peripheral margin. Histological review of the tissue demonstrated an ill-defined tumor composed of nests and trabeculae of large polygonal cells with abundant eosinophilic granular cytoplasm. Increased cellularity, prominent cellular spindling, high nuclear-to-cytoplasmic (N/C) ratio, increased mitotic figures, and necrosis were not observed. The tumor was found to be positive for S100 and CD68. The morphology and immunoprofile supported the diagnosis of GCT.

The patient was subsequently discharged home on postoperative day one. She was seen in the clinic at her two-week postoperative evaluation. The patient reported her pruritus and buttock discomfort had resolved, and her incision had healed well, without any signs of dehiscence or infection.

Discussion

The first cases of GCT were described as early as 1854, with reports of diagnosis within the tongue; they were further investigated in 1926 and described as a “myoblastoma” of the tongue.⁵ This soft-tissue lesion is an overall uncommon tumor that is typically identified at a single location; however, there are reports of multiple tumors in different areas of the body.⁶ The oral cavity and tongue remain the most reported sites of GCT occurrence, followed by the skin and subcutaneous tissue. There is sparse information available for GCT of the perianal region.² Less than 100 cases of perianal GCTs have been reported since 1945. While there is a higher prevalence of GCT in women, there has not been a significant difference in gender regarding perianal GCT. Patients diagnosed with GCT in the perianal region were commonly diagnosed in the second or fifth decades of life, with no significant difference in the incidence of age among male and female patients.⁷ Perianal GCT is derived from neural tissue characterized by Schwann cell origin with immunohistochemical staining positive for S100.^{2,8} In the patient herein described, the tumor was located on the left buttock without direct involvement of the skin, which was considered a rare location for GCTs.

Perianal GCTs of the subcutaneous tissue are largely asymptomatic; however, these tumors can present with localized discomfort and bleeding. Preoperative workup and diagnosis may be challenging, with a broad differential for skin and soft tissue masses including slow growing lipoma, superficial hemangioma, fibroma, leiomyoma, sebaceous cyst, or superficial myxoma.⁸ The gold standard for diagnosis remains immunohistochemical staining and histopathology.^{2,8,9} Immunohistochemical staining for GCT has routinely expressed neuron-specific enolase (NSE), S100, as well as CD57, a marker of natural killer cell maturation.⁸ Interestingly, individuals with congenital GCT had negative histochemical testing for S100.^{2,9} These tumors have been

found to show positivity for vimentin, a major intermediate filament within non-muscle cells, as well as CD68, a macrophage marker responsible for cytoplasmic lysosomal aggregation.⁸ The hypothesis that GCT signifies a non-specific degenerative progression of mesenchymal origin via self-phagocytosis has been raised.² As with the case herein reported, the histopathology demonstrated positive immunostaining for S100 and CD68, while negative for AE1/AE3, markers for high molecular weight cytokeratins which exclude an epidermal source. **►Fig. 1 (A-D)** demonstrates multiple cross-sectional images of different immunohistochemical stains. The cytoplasm of our specimen exhibited large polygonal cells with abundant eosinophilic granular cytoplasm, consistent with findings in GCT. Vimentin and NSE were not tested in this particular case.

Most GCTs present as benign, with less than 2% diagnosed as malignant.¹⁰ Benign and malignant tumors may share similar histopathology, making definitive diagnosis difficult. Malignant GCTs have previously been divided into two groups. The benign type (type 1) is clinically malignant with benign histopathology. The malignant type (type 2) presents histologically malignant features.^{2,10,11} In the present study, 6 histologic criteria were assessed: necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity (> 2 mitoses/10 high-power fields at 200x magnification), N/C ratio, and pleomorphism.¹⁰ However, there is no consensus regarding a histological algorithm for diagnosis. Malignant GCT is associated with faster tumor growth, a diameter > 4 cm, cell necrosis, spindle-shaped tumor cells, cellular atypia, active mitosis (more than 2/10 high-power fields at 20x magnification), high N/C ratio, local recurrence, and high p53 and K_i-67 expression.² In the patient herein described, we did not observe increased cellularity, prominent cellular spindling, high N/C ratio, increased mitotic figures, or necrosis. While there are no radiologic results specific to GCTs, magnetic resonance imaging (MRI) is the best imaging modality to differentiate between benign and malignant lesions. Within the muscle, benign GCTs will appear brighter than muscle on T1-weighted imaging.¹² On T2-weighted sequences, the center of the lesion is isointense to muscle with peripheral enhancement. **►Fig. 1 (E-F)** shows T1- and T2-weighted images of GCTs in the perianal region.¹²

The treatment for GCTs, whether benign or malignant, is wide surgical excision with negative margins and close postoperative follow-up.¹³ Sentinel lymph node biopsy is only recommended for clinically suspicious lesions or when histology is concerning for malignancy.¹³ The risk of recurrence with negative margins is rare with benign GCTs, with a rate ranging from 2% to 8%, but it increases to > 50% with an R1 resection.^{13,14} Malignant GCTs present a significantly higher risk of recurrence, of nearly 41%, even with an R0 resection. The risk of metastasis increases to ~ 60% 3 years after the diagnosis. The role of chemoradiation in GCTs is limited. Currently, there is no standard chemotherapy regimen, and adjuvant radiation has been controversial and ill-defined, with no substantial survival benefit.^{14,15}

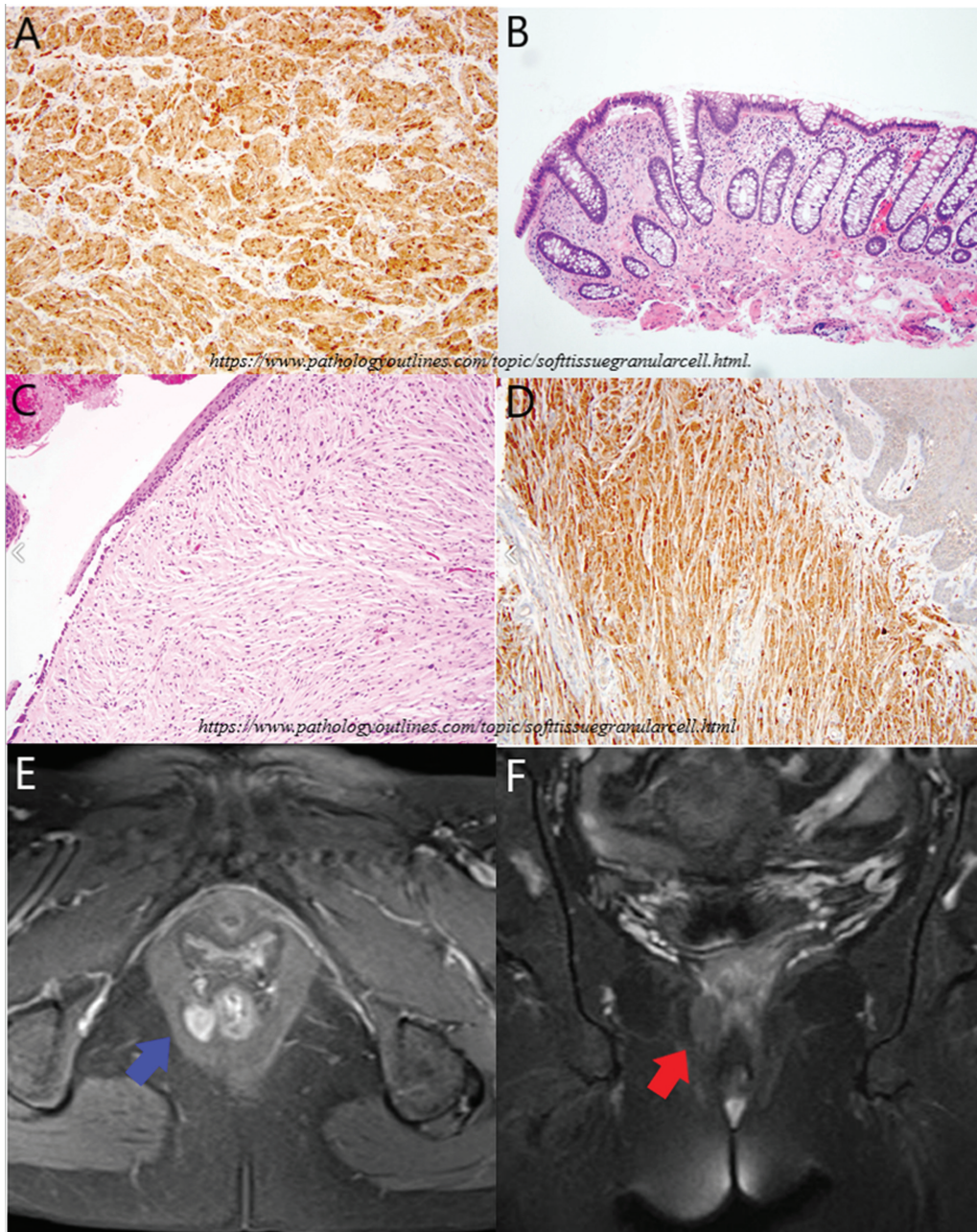


Fig. 1 Different immunohistochemical stains on magnetic resonance imaging (MRI) scans of a perianal granular cell tumor (GCT). (A) Immunohistochemical staining in a GCT that is S100+. (B) Hematoxylin and eosin (H&E) staining. (C) H&E staining demonstrating fibrous and cellular spindling appearance. (D) CD68+ staining. (E) Axial view of T2-weighted MRI of the pelvis demonstrating perianal GCT (blue arrow). (F) Coronal view of T1-weighted MRI showing perianal GCT (red arrow).

A GCT located in the subcutaneous tissue of the buttock is an infrequent diagnosis, with an overall low risk of malignant conversion. However, a complete workup should always be conducted with histopathological evaluation for accurate diagnosis and treatment. Due to the high recurrence rate of malignant GCTs and with incomplete surgical excision, close follow-up is strongly recommended.

Conflict of Interests

The authors have no conflict of interests to declare.

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