

# Malignant Granular Cell Tumor of the Abdominal Wall Mimicking Desmoid Tumor: A Case Report with CT Imaging Findings and Literature Review

복벽을 침범한 악성 과립세포종의 증례와 CT 영상소견 보고 및 문헌 고찰

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Granular cell tumors (GCTs) are extremely rare mesenchymal neoplasms of Schwann cell origin. Malignant GCTs (MGCTs) comprise 0.5–2% of all GCTs. In the present report, we describe a case of a 66-year-old man with MGCT of the abdominal wall. The patient visited our hospital due to a recently growing palpable soft tissue mass in the abdominal wall. Computed tomography scan revealed a 4.3 × 4.1 × 2.9 cm sized mass arising from the left abdominal wall, which was contemplated as a desmoid tumor before surgical excision. Histopathological examination confirmed MGCT.

## Index terms

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## INTRODUCTION

Granular cell tumor (GCT) is an extremely rare mesenchymal soft tissue neoplasm of Schwann cell origin. GCT may occur throughout the body, usually in the head and neck, trunk and upper extremities; rarely in the abdominal wall. They are usually benign and solitary; however, approximately 0.5–2% of GCTs are malignant tumors, and 5–10% of GCTs occur in the form of multiple lesions (1-4). While the benign GCTs are smaller and slow-growing tumors, malignant GCTs (MGCTs) are larger and fast-growing tumors, and exhibit a high rate of metastasis and short survival. The radiologic findings of the lesion have been described only in a few reports. In this article, we report a case of 66-year-old man with pathologically proven MGCT arising in the abdominal wall muscles, illustrate the computed to-

mography (CT) findings of the lesion, and discuss imaging differential diagnosis.

## CASE REPORT

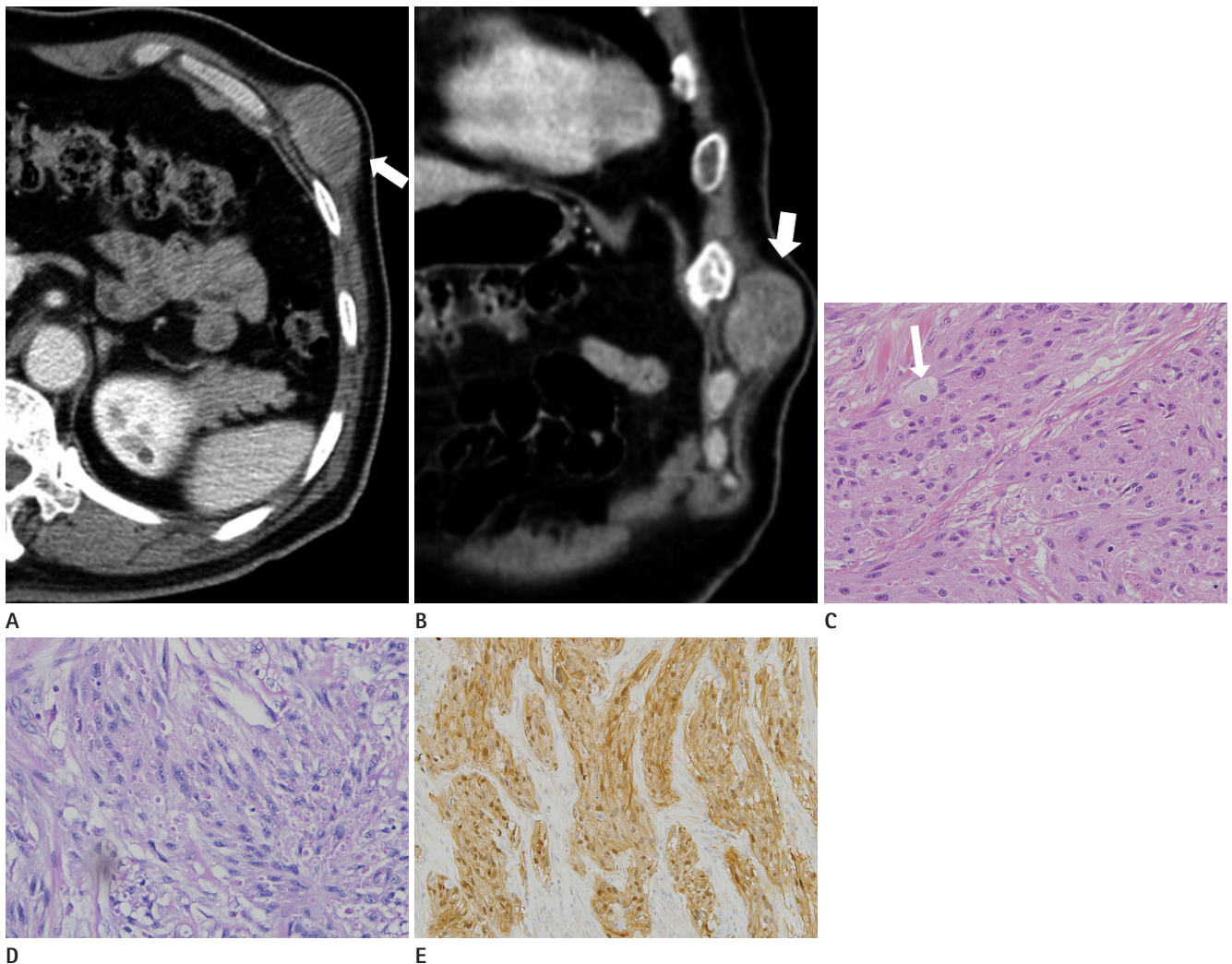
A 66-year-old man visited our hospital with a 30-year history of a palpable and recently growing soft tissue mass in the left anterolateral abdominal wall. Physical examination revealed a hard and well-circumscribed mass with no tenderness. The mass was covered by apparently normal skin. The patient had no history of abdominal trauma or surgery.

CT (LightSpeed 16, GE Healthcare, Milwaukee, WI, USA) demonstrated a relatively homogeneous enhancing solid mass in the left anterolateral abdominal wall, arising from the muscle layer (Fig. 1A). A subtle ill-defined low attenuated portion was ap-

parent within the mass and the mass measured  $4.3 \times 4.1 \times 2.9$  cm. The mass did not show extension to the parietal peritoneum or subcutaneous fat layer. The mass was well-defined, but focally ill-defined at the inferior aspect (Fig. 1B). There was no calcification or cystic portion within the mass.

Initially, the CT findings suggested the presence of a benign soft tissue mass, which was considered as a desmoid tumor. Subsequently, surgical mass excision was performed. The mass showed adhesions to the adjacent subcutaneous fat tissue. On macroscopic inspection, a partially ill-demarcated round mass,

measuring  $4.5 \times 3.4 \times 3.0$  cm was found. On histological examination, tumor cells with abundant cytoplasm and intracytoplasmic granules (Fig. 1C) were observed and they were found to be positive for periodic acid-Schiff staining reaction (Fig. 1D) and S-100 (Fig. 1E). Up to 3 mitotic figures were counted in 10 high-power fields. The histologic report confirmed the diagnosis of malignant GCT. The patient was discharged after the surgery without any complications. There was no evidence of local recurrence or metastasis on follow up imaging studies after 30 months.



**Fig. 1.** A 66-year-old man with a malignant granular cell tumor of the abdominal wall muscle.

**A.** The axial contrast-enhanced abdominal CT scan reveals a well-defined, relatively homogeneous enhancing solid mass in the left anterolateral abdominal wall, arising from the left internal oblique and transverse abdominal muscles. A focal ill-defined low attenuated portion can be seen in the mass (arrow).

**B.** The coronal image demonstrates a focally ill-defined mass at the inferior edge (arrow).

**C.** Hematoxylin-eosin staining ( $\times 400$ ) shows tumor cells with abundant cytoplasm and intracytoplasmic granules (arrow).

**D.** Periodic acid-Schiff stain ( $\times 400$ ) shows tumor cells with positive purple-magenta staining.

**E.** Staining for S-100 ( $\times 150$ ) shows tumor cells with positive red-brick color staining.

## DISCUSSION

GCTs are rare and almost invariably benign masses, which usually develop as painless masses with normal overlying skin. GCTs occur in a wide variety of visceral sites, and skin and subcutaneous tissue. In majority of the cases, the head, neck and oral cavity (30%), especially the tongue, are involved. McGuire et al. (5) reported that MGCTs are twice as likely to occur in females as in males in the age range of 30–50 years. The neoplasm is typically slow-growing, well-circumscribed, firm and rounded, with a diameter ranging from 5–20 mm, although larger tumors may be seen. The skin surrounding the tumor is usually normal, although it may be thickened, hyperpigmented, and on

rare occasions, it may be ulcerated (4, 6).

Most of the GCTs exist as a benign solitary nodule with very scarce cases of MGCTs in the form of a high-grade sarcoma of Schwann cell origin and they are also reported as Abrikossoff's tumor (7). Although GCT was initially reported as myoblastoma of the tongue based on the presence of signs of infiltration between the striated muscles, indicating a muscular origin, recent studies have indicated that it has a peripheral neuroectodermal origin as the tumor cells are strongly positive for S-100 protein, similar to the Schwann cells in the peripheral nerve (4, 8).

Histologically, benign GCTs are composed of large polygonal cells containing numerous eosinophilic granules. The nuclei are relatively small and mildly pleomorphic with prominent nucle-

**Table 1. Summary of the 11 Cases of Reported Granular Cell Tumors of the Abdominal Wall**

Author (Year)	Age/Sex	Symptoms	Size (cm)	Location	Imaging Findings	Preoperative Diagnosis	Pathology (B/M)
1 Gorelkin (1978)	58/F	Incidental finding on physical exam	8 × 4	Rectus muscle	-	-	B
2 Vamsy (1992)	30/F	-	9	-	-	-	M
3 Fanburg (1998)	49/F	-	11	Abdominal wall	-	-	M
4 Fanburg (1998)	32/M	-	5.5	Rectus sheath	-	-	M
5 Joshi (2003)	37/M	Painless lump	2.7 × 2	Rectus muscle	-	Organized hematoma	B
6 Chelly (2005)	67/F	Abdominal pain	10	Anterior abdominal wall (subcutaneous layer to peritoneal cavity)	Heterogeneous iso to low attenuated solid mass (CT)	Recurrent hydatid cyst	M
7 An (2007)	44/F	Non-tender, hard mass	3.6 × 2.5	Rectus muscle	Well-defined oval shaped hypoechoic mass (US), ovoid mass (CT)	Desmoid	B
8 Claudhry (2008)	70/F	Firm, fixed mass	10 × 7	Internal oblique and transversus abdominis	Spiculated homogeneous iso attenuated solid mass (CT)	Sarcoma	B
9 Kang (2010)	44/F	Non-tender, painless lump	1.7 × 1.3	Rectus muscle	Iso to high attenuated solid mass (CT) Ill-defined spiculated margin, hyperechoic rim, posterior acoustic shadowing (US)	-	B
10 Panunzi (2012)	29/F	Swelling	1.2 × 1.0	Rectus muscle	Well-defined homogeneous hypoechoic mass without vascularity (US)	-	B
11 Toelen (2013)	68/M	Hard, painless, palpable nodule	3	Subcutaneous layer	Homogeneously iso attenuated lesion without calcification and necrosis (CT)	Desmoid or fibromatosis	

B = benign, M = malignant, US = ultrasonography

oli (3). Approximately 0.5–2% of GCTs are malignant, and they have the following characteristics which indicate more likelihood of malignancy; mass size larger than 5 cm, nuclear pleomorphism, metastatic lymph nodes, aggressive clinical behavior, rapid growth, and ulceration (1-4).

In order to diagnose MGCT, Fanburg et al. devised a classification system by analyzing 73 atypical MGCTs using the following six parameters: increased nuclear-to-cytoplasmic ratio, nuclear pleomorphism, necrosis, spindling of tumor cells, vesicular nuclei with prominent nucleoli, and a mitotic count of more than two in 10 high-power fields (200 × field). According to the Fanburg criteria, GCT was classified as atypical if it satisfied 1–2 criteria, and it was classified as malignant if it satisfied 3–6 criteria. Although rare, complete surgical resection of all GCTs is recommended because of its malignant potential and possibility of local recurrence (9).

In our study, the lesion was confirmed to be MGCT and we observed a mass with soft tissue attenuation located in the abdominal wall muscle with relatively homogeneous attenuation including a subtle internal low attenuated portion, which is considered to reflect the pathologically necrotic portion. This lesion was first thought to be a desmoid tumor. Typically, 28–69% of desmoid tumors have been reported to present as an intra-abdominal or an abdominal wall mass in various studies, and since a desmoid tumor manifests as a soft tissue mass with variable attenuation and enhancement on CT, it is difficult to differentiate it from GCT due to overlap of many imaging features (10). The radiologic characteristics of MGCTs are unclear since there are few reports on the imaging findings, but generally, since the disease presents as a mass with soft tissue attenuation, differential diagnosis with soft tissue tumorous lesions is necessary. If the lesion appears heterogeneous or has a part showing low attenuation, the possibility of MGCT should be considered.

GCTs rarely develop in the abdominal wall. There are 10 cases of GCTs of the abdominal wall in the English literature and one case in the Korean literature (11). These cases are summarized in Table 1, including patient characteristics such as age, sex, symptoms, locations, preoperative diagnosis, imaging findings, and pathologic group. According to the table, four out of the eleven cases were confirmed to be of MGCT; therefore, in GCTs with abdominal wall involvement, a higher prevalence of malignancy is more likely than in the previously known cases.

All the four previously reported MGCTs of the abdominal wall were more than 5 cm in terms of mass size, but the lesion in our case was smaller than 5 cm. Also, two of the six benign abdominal GCTs measured 8 cm and 10 cm, respectively. This proves that it is difficult to differentiate between benign and malignant lesions based on the size alone.

There is only one case report describing an abdominal MGCT. The reported CT imaging finding of MGCTs was a heterogeneously low attenuated mass within the subcutaneous layer of the abdominal wall extending into the peritoneal cavity. However, since our case presented as a well-defined, relatively homogeneous enhancing solid mass despite being a MGCT, the imaging findings of MGCTs can be considered to be non-specific. Therefore, it is difficult to differentiate between benign and other malignant lesions such as sarcoma or desmoid tumor based on the imaging findings alone or the size criteria.

In conclusion, although GCTs of the abdominal wall are extremely rare, they should be considered in the differential diagnosis of an abdominal wall mass arising from the abdominal muscles. Furthermore, even if the lesion is less than 5 cm and presents as a homogeneously enhancing solid mass, it is difficult to differentiate benign lesions from malignant lesions based on the size or imaging findings alone. Therefore, it is necessary to confirm the diagnosis through biopsy or to perform complete surgical resection of a mass with abdominal wall involvement.

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## 복벽을 침범한 악성 과립세포종의 증례와 CT 영상소견 보고 및 문헌 고찰

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과립세포종(granular cell tumor)은 Schwann 세포에서 기원하는 매우 드문 종류의 중간엽 세포성 종양이다. 악성 과립세포종은 전체 과립세포종의 약 0.5~2%를 차지하는 것으로 알려져 있는데, 본 증례 보고에서는 66세 남성에서 복벽을 침범한 악성 과립세포종의 사례를 다루고 있다. 환자는 최근 들어 좌측 복벽에서 촉진 가능하면서 지속적으로 크기 증가를 보이는 종괴를 주소로 내원하였으며, CT에서 좌측 복벽의 약  $4.3 \times 4.1 \times 2.9$  cm 크기의 종괴가 확인되어 섬유모세포 종양(desmoid tumor) 의심하에 수술적 제거를 시행하였으나 최종 병리소견에서 악성 과립세포종으로 확인되었다. 이에 저자들은 복벽을 침범한 악성 과립세포종의 영상 및 병리소견과 함께 관련 문헌 고찰을 보고하고자 한다.

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