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## Case Report

# Magnetic resonance imaging (MRI) of perianal granular cell tumor: A case report and literature review

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## ABSTRACT

Granular cell tumor (GCT) is a rare soft tissue neoplasm. At present, there is limited information available on the incidence, diagnosis, and treatment of GCTs. Therefore, we perform magnetic resonance imaging (MRI) of a perianal GCT case and review the literature of other reported cases to obtain a more comprehensive understanding of the disease. Here, we describe a rare case of perianal GCT in a 53-year-old female who presented with recurrent abdominal pain. Contrast-enhanced MRI demonstrated a well-defined perianal GCT mass, and the diagnosis was confirmed by immunostaining after mass excision. During a 10-month follow-up, there had been no evidence of recurrence. In addition, we discussed the findings of other perianal GCT cases, with regard to their age, gender, MRI analysis results, pathologic features, and treatment outcomes.

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## Introduction

Granular cell tumor (GCT) was initially found in the tongue by Abrikossoff in 1926 [1]. It has been identified as a rare neurogenic tumor originating from Schwann cells of the peripheral nerves, due to its positivity for S-100 via immunohistochemical staining [2]. GCTs have been observed in many different locations throughout the body, especially in the skin, oral cavity, and subcutaneous tissue [3–5]. Indeed, perianal GCTs are

extremely rare. Here, we describe a rare case of perianal GCT diagnosed by magnetic resonance imaging (MRI) and provide a review of the related literature.

## Case report

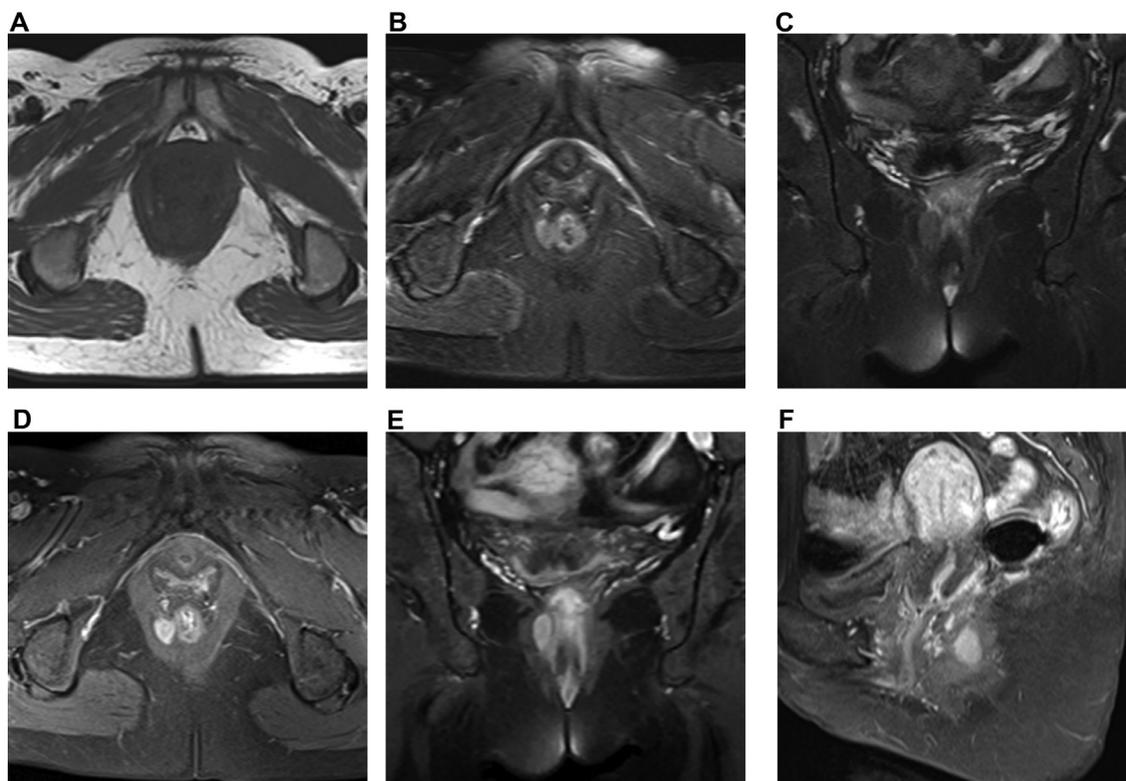
A 53-year-old woman presented with recurrent abdominal pain when being overworked or exposed to cold, due to a

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**Fig. 1 – Contrast-enhanced MRI of the perianal region. (A) Axial T1-weighted nonfat-saturated MRI image showed a low signal tumor (white arrow) in the perianal area. (B and C) Axial and coronal T2-weighted fat-saturated images demonstrated slightly high signals. (D-F) Axial, coronal, and sagittal gadolinium-enhanced T1-weighted images indicated an obvious enhancement.**

palpable mass in the perianal area. The tumor mass had been in the region over the past 15 years. The patient exhibited no weight loss, abdominal pain, abdominal distension, diarrhea, and hematochezia. No treatment was applied, and the abdominal pain was relieved by resting. Digital rectal examination revealed a hard, about  $1.0 \times 1.5 \text{ cm}^2$  mass from the anal verge at 8-9 o'clock direction. Physical examination indicated 2 mixed hemorrhoids at 2 and 7 o'clock directions, respectively. The results of electronic colonoscopy were normal, and the laboratory data were unremarkable. No additional disease was found in the patient's medical history.

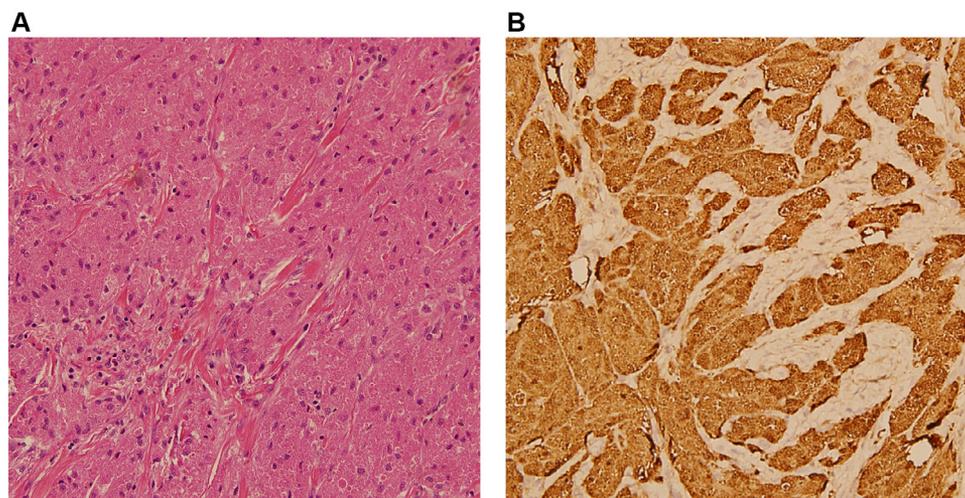
MRI (Siemens Verio, Shanghai, China) scanning of the perianal region was performed using the belt body coil. The images of axial T1WI (Fig. 1A), fat-saturated TSE T2WI (Fig. 1B), and coronal fat-saturated TSE T2WI (Fig. 1C) were then acquired. The patient underwent contrast-enhanced MRI following an intravenous administration of 30 mL gadolinium (2.0 mL gadolinium per second). Finally, axial, coronal, and sagittal TSE T1WI imaging (Fig. 1D-F) were carried out after contrast injection.

MRI images showed an ovoid lesion located approximately 2 cm above the anal verge at 8-9 o'clock direction. The size of the mass was approximately  $1.2 \text{ cm} \times 1.1 \text{ cm} \times 1.6 \text{ cm}$ , with a well-defined margin and outward growth. The tissue mass was connected to the anal canal by a narrow base without

definite invasion margins. Moreover, the mass exhibited isointensity on T1-weighted images, and slightly slight hyperintensity compared to the surrounding tissues on T2-weighted images. After intravenous administration of gadolinium diethylenetriaminepentaacetic acid, the lesion showed obvious enhancement, and the edge enhancement was more pronounced than in the central region. There were no obvious exudation and enlarged lymph nodes around the lesion, and no fistulous tract was observed across the anal sphincter.

The patient underwent local excision of the perianal GST under lumbar anesthesia. The tumor was excised en bloc with hemorrhoids via a transanal approach. The mass was approximately 1.5 cm in diameter, smooth, and movable between the anal canal and the levator ani muscle. The surgically excised specimen appeared to be gray-white and hard without capsule. Histopathological examination revealed nests of mononuclear cells with abundant granular eosinophilic cytoplasm and uniform nuclei (Fig. 2A). Immunohistochemical staining with the specific primary antibodies gave the following results: S-100 (+) (Fig. 2B), CD-68 (+), SOX-10 (+), KP-1 (+), CK (-), Desmin (-), Calretinin (-), Inhibin $\alpha$  (-), and less than 5% for Ki-67 index.

The patient's follow-up consisted of physical examinations. There had been no evidence of recurrence during the 10-month follow-up period.



**Fig. 2** – H&E-stained image and cell immunohistochemistry image of the perianal GCT. (A) H&E-stained image (x 200) revealed a noncapsulated mass composed of nests of mononuclear cells with abundant eosinophilic granular cytoplasm and bland nuclei. (B) Cell immunohistochemistry image (x 200) showed strong and diffuse positive staining for S-100.

## Discussion

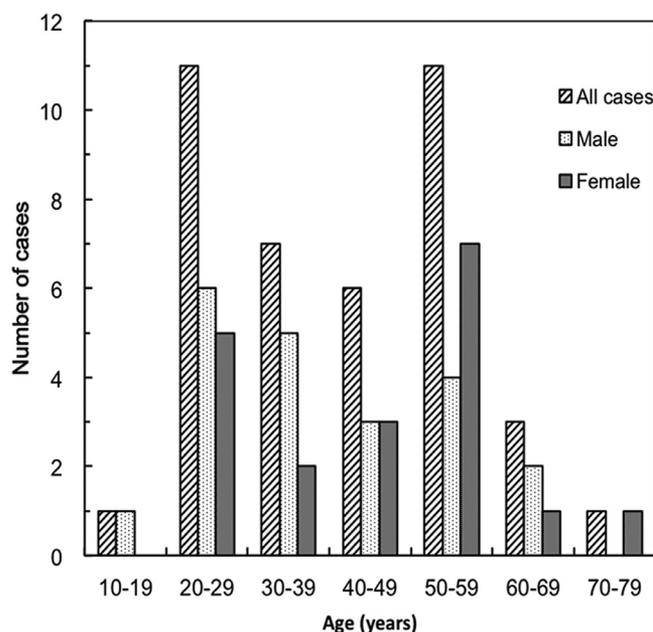
GCTs are soft tissue neoplasms of mesenchymal origin and derive their name from the presence of coarse cytoplasmic granularity that is typically found among their constituent cells. They are believed to arise from Schwann cells as they are positive for S-100, myelin, and myelin-associated glycoprotein. GCTs are uncommon, benign masses that often present as solitary small nodules.

### Demographic characteristics

Approximately 8% of GCTs have been reported to occur in the digestive tract [6]. The most common occurrence site in the digestive tract is the esophagus, followed by the large intestine. Typically, GCTs are rarely found in the perianal. Only forty cases of perianal GCTs have been reported in the English literature since 1945 [7], including the current case. Notably, these cases were consisted of 21 males and 19 females, indicating that there is no significant difference in GCT occurrence between genders. It was clearly seen that the tumor could occur at any age ranged from 18 to 75 years. Age data revealed that perianal GCT patients were most likely to be diagnosed in the second or fifth decade of life (Fig. 3). The mean age  $\pm$  SD of males was  $39.00 \pm 13.87$  years (range: 18-66 years), while that of females was  $44.47 \pm 14.94$  years (range: 22-75 years), with an overall value of  $41.60 \pm 14.47$  years. No statistically significant difference was found in the incidence age between male and female patients ( $P = .237$ ).

### Clinical characteristics

Patients with GCT are lacking of typical clinical presentation. Only 14 out of the 40 perianal GCT cases exhibited clinical symptoms. Of them, 10 cases were manifested as painless and palpable masses. Three cases presented with painful masses



**Fig. 3** – Age at presentation of perianal GCTs.

and 1 of them had hemorrhoids. Rectal bleeding was only found in 1 case. The data on tumor size were only available for 10 cases. Maximum tumor diameter of the patients varied from 0.70 to 5.00 cm (mean  $\pm$  SD,  $2.66 \pm 1.54$  cm).

### MRI features

MRI is the imaging technique of choice for preoperative evaluation of soft tissue masses because of its ability to show high soft tissue contrast and direct image at multiple planes. The use of preoperative MRI may assist in surgical

**Table 1 – Literature review of imaging appearance of perianal GCTs.**

Reported by (years)	Age (years)/gender	MRI imaging	Follow up (months)	Prognosis
Wan et al. 2014 [8]	48/female	A brightly enhancing mass	3	No recurrence
Rashidi et al. 2017 [9]	36/female	An ovoid, solid mass in the right perianal region, uniform low T1 and intermediate to high T2-weighted signal intensity, homogeneous and avid enhancement, no definite invasion to the anal canal, no surrounding enlarged lymph nodes	Not reported	No recurrence
Terrah et al. 2018 [10]	52/female	A heterogeneously enhancing soft tissue perianal mass with restricted diffusion, no invasion into the ischioanal fat involve both the internal and external anal sphincters, mildly enlarged pelvic lymph nodes	8	Recurrence
Current case	53/female	An oval-like lesion, well-defined margin, low signal intensity on T1-weighted images and slightly high on T2-weighted images, obvious enhancement, no obvious exudation around the lesion, no surrounding enlarged lymph nodes	10	No recurrence

decision-making process for GCT patients with uncertain malignant potential. The MRI results of all the case are listed in Table 1. Four cases, including the present case, had been diagnosed by MRI scanning [8–10]. MRI images revealed an ovoid and solid mass with well-defined borders. Three tumors exhibited low signal intensity on T1-weighted images and slightly high intensity on T2-weighted images. The status of tumor diffusion was not reported in 1 case. All cases showed enhancement after injection of gadolinium-based contrast agents. The enhancement was apparent either homogeneous or heterogeneous. Taken altogether, several features should arouse suspicious of GCT, including well-defined masses, location between the anal canal and the levator ani muscle, isointensity on T1WI and hyperintensity on T2WI, as well as a heterogeneous or homogeneous enhancement on T1WI.

#### Pathologic features

GCTs display a characteristic appearance under histopathologic examination. The masses are typically uniform and contain enlarged eosinophilic cells with granular cytoplasm. Immunohistochemical staining is the key method of GCT diagnosis, as the cells can be intensively stained with an anti-S-100 antibody. The results of immunohistochemical staining had been reported in 10 perianal GCT cases, and all of them were positive for S-100 protein immunoreactivity. Among the 10 cases, 4 GCT cases were positive for CD-68 and 3 cases were positive for NSE.

#### Treatment and follow-up

At present, surgical resection is the most effective treatment for GCTs. It can prevent local recurrence and metastasis. Almost all the GCT patients underwent a single procedure of gross total resection. In most GCT cases, the prognosis is favorable after complete surgical resection, without any recurrence and metastases. Benign lesions have an excellent prognosis following surgical excision, whereas malignant lesions tend to

recur after surgery. The prognosis of malign perianal GCTs is even worse. Follow-up data were available for 7 perianal GCT cases. Five of them had no recurrence. One patient had local recurrence during the 8 months of follow-up. In the remaining 1 patient, recurrent perianal tumor and bilateral metastatic inguinal lymph nodes were observed 17 months after surgery. In this report, the patient manifested with pulmonary and hepatic metastases at 32 months postsurgery, and recovered within the same year. Therefore, regular follow-up visits are necessary, especially for GCT cases with typical histology or large in size.

#### Conclusion

In conclusion, we have reported a case of perianal GCT and reviewed the relevant literature in order to get a better insight into the occurrence, diagnosis, and treatment of this rare tumor. Although our study only reveals a crude picture of the disease, the data of GCT cases in terms of age, gender, MRI analysis results, pathologic features, and treatment outcomes reported here can provide a reference for improving its diagnosis and treatment in the future.

#### Conflicts of interest

The authors report there are no conflicts of interest.

#### Acknowledgments

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## Supplementary material

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2019.05.010](https://doi.org/10.1016/j.radcr.2019.05.010).

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