

CASE REPORT

## Synchronous granular cell tumors in the perianus and chest wall

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Granular cell tumor (GCT) is a rare tumor that originates from the Schwann cells in the skin, soft tissues, and internal organs. Usually, GCTs are clinically benign, although malignant and multifocal forms are very rarely known to occur. Cases of GCT of the perianus are rare, and thus far, no study has reported synchronous GCTs of the perianus and the chest wall. We report a case of a 31-year-old woman with synchronous GCTs of the perianus and the chest wall to have a mind of consideration of the possibility of GCT in the differential diagnosis of perianal tumor.

**Key Words:** Granular cell tumor, Perianus, Synchronous

### INTRODUCTION

Granular cell tumor (GCT) is a rare soft-tissue neoplasm that may develop at many different locations in the body, and generally appears as a benign solitary nodule [1].

The gastrointestinal tract is one of the uncommon locations of GCT development [2], and cases of GCTs in the perianal region are extremely rare [3]. Although multiple lesions may appear, synchronous GCT of perianus with chest wall has not reported yet.

A majority of the GCTs show benign behavior, but only 1 to 2% of the GCTs are malignant [4]. The differential diagnosis of malignant GCTs from benign ones is sometimes difficult because the histological appearance of a malignant GCT is similar to that of a benign one, and malignancy can be diagnosed only after the occurrence of metastases [2,4].

We report a case of synchronous GCTs of the perianus and the chest wall to have a mind of consideration of the possibility of GCT in the differential diagnosis of perianal tumor, and possible malignant potential must be considered during therapeutic procedures and follow-up.

### CASE REPORT

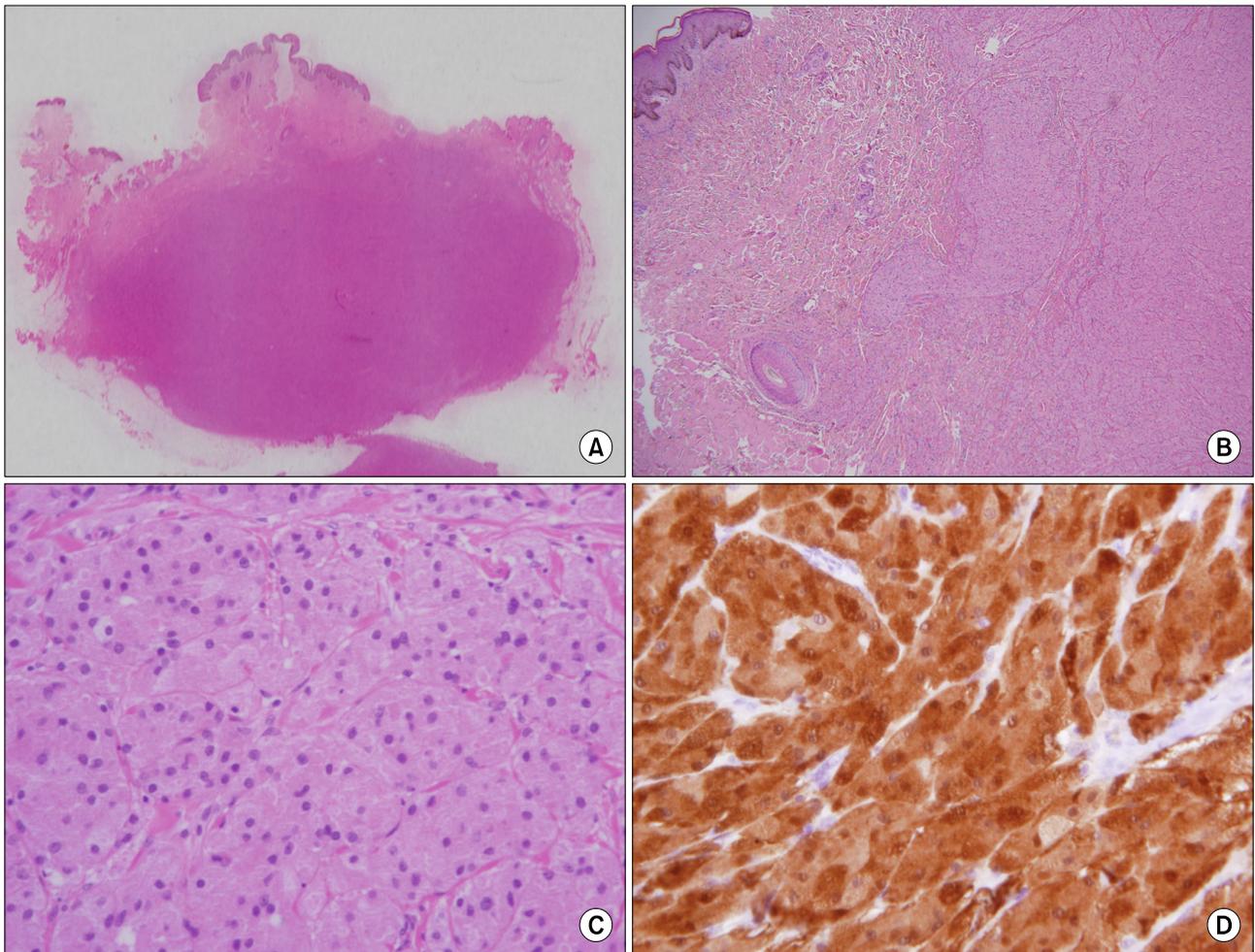
A 31-year-old woman with a 2-year history of a painless subcutaneous nodule on the left upper chest wall presented with a progressively growing perianal mass that had developed 1 year before she consulted this department.

She had no significant medical history. Physical examination showed a light gray-colored, polypoid, well-defined, firm, movable perianal mass (size, 1.5 cm) adjacent

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**Fig. 1.** Histopathological findings of the perianal mass. The lesion was characterized by polygonal cells with abundant eosinophilic granular cytoplasm, and an immunohistochemical study revealed that the cells were positive for the S-100 (A, H&E,  $\times 1$ ; B, H&E,  $\times 40$ ; C, H&E,  $\times 400$ ; D, S-100 protein).

to the anal verge and extending to the perianal skin. A benign fibrous tissue tumor was first considered and a quiescent perianal abscess should be considered in the differential diagnosis. The mass in the chest wall (size, 1.5 cm) was a firm and well-circumscribed subcutaneous nodule. For pathological diagnosis, a surgical excision was performed for both chest and perianal masses under the local anesthesia.

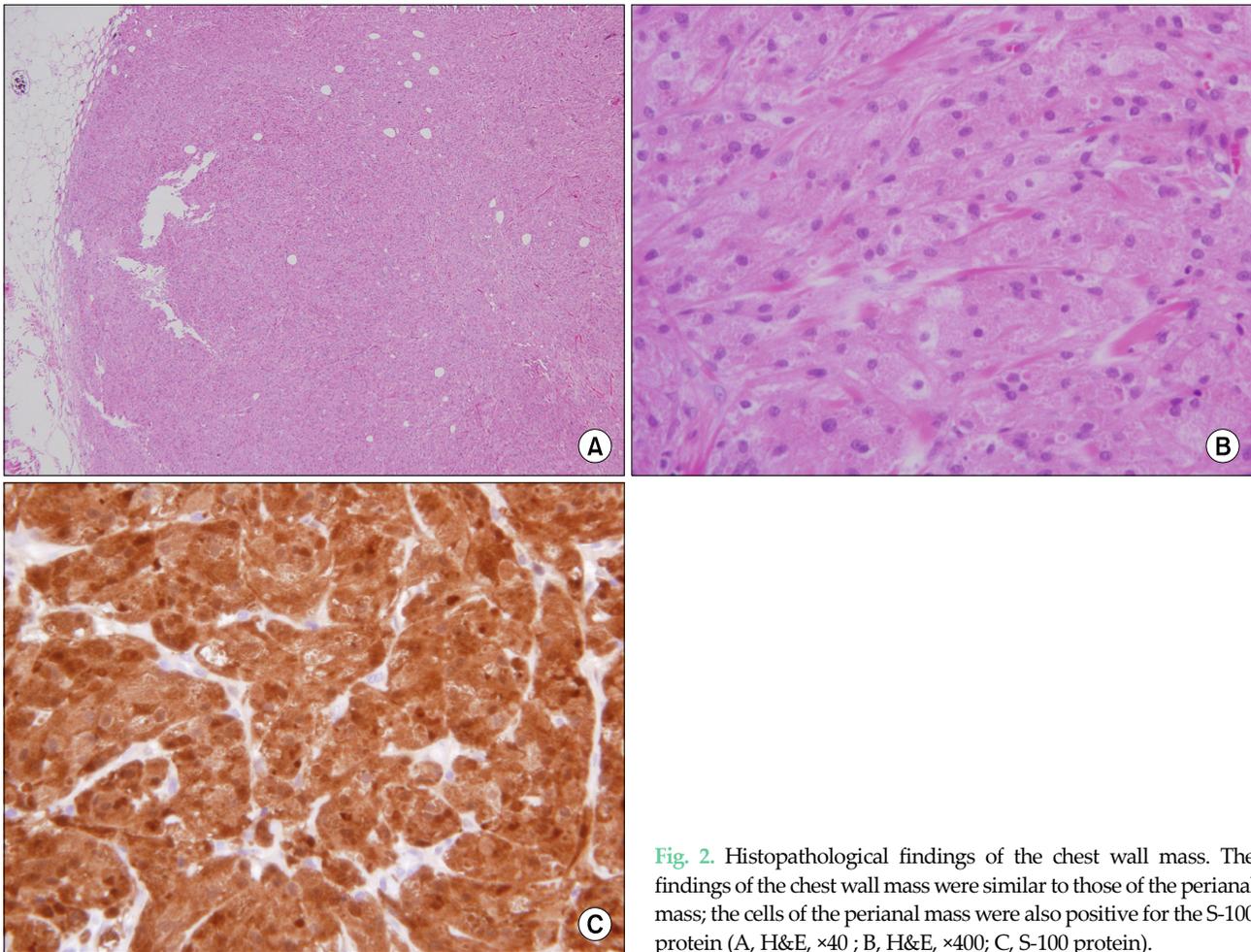
Histopathological examination of the perianal mass showed that the mass was characterized by polygonal cells with abundant eosinophilic granular cytoplasm, and an immunohistochemical study revealed that the cells were positive for the S-100 protein (Fig. 1). No nuclear mitosis and necrosis associated with malignant cells were ob-

served, and these histopathological findings were consistent with a typical benign GCT. Histopathological and immunohistochemical findings of the chest wall mass were similar to those of the perianal mass (Fig. 2).

The postoperative course was uneventful. The patient was followed up for 1 year, and no evidence of recurrence was observed.

## DISCUSSION

GCTs were first described by Abrikossoff [5] in 1926. They are rare and usually benign soft-tissue neoplasms that appear in the form of solitary small nodules. GCTs



**Fig. 2.** Histopathological findings of the chest wall mass. The findings of the chest wall mass were similar to those of the perianal mass; the cells of the perianal mass were also positive for the S-100 protein (A, H&E,  $\times 40$ ; B, H&E,  $\times 400$ ; C, S-100 protein).

may occur at any sites of the body, although they most frequently affect the skin or the subcutaneous tissues of the chest, upper extremities, tongue, breasts, and female genital region. Cases of GCT development in the gastrointestinal tract are rare. Approximately 8% of all GCTs have been reported to occur in the digestive tract, and the most common site of these GCTs was reported to be the esophagus, followed by the large intestine [1]. Cases of GCTs in the perianal region are extremely rare [3], and thus far, no study has reported synchronous GCTs in the perianus and the chest wall.

The histogenesis of GCTs remains controversial. Abrikossoff [5] proposed that GCTs originated from degenerating striated muscles, but later modified his view in favor of the origin of GCT from embryonic muscle cells. He termed the lesion myoblastic myoma.

In 1946, Fust and Custer [6] first hypothesized that the

“myoblastoma” had a neural origin. Since then, many studies presented light microscopy, histochemical, and electron microscopy data that suggest that the lesions may have a neurogenic origin.

More recently, Aparicio and Lumsden [7] and Sobol et al. [8] suggested that GCTs originate from undifferentiated mesenchymal cells. Because of the uncertainty in the origin of these lesions, terminology of GCT rather than myoblastoma would be appropriate for this tumor.

Recently, many studies on GCTs have mainly focused on the recognition and correct preoperative diagnosis of this tumor in clinical practice, and on determining the histogenesis of this tumor by performing immunohistochemical, ultrastructural studies and cytogenetic analysis, but not on the management and clinical behavior of the tumor [9].

Although generally GCTs are clinically and histologi-

cally benign, some cases of malignant GCTs have been reported [10]. Less than 60 cases of malignant GCTs have been reported in the literature, but no case of a malignant GCT of the perianus has not been reported yet.

The general features associated with the malignant GCT are rapid growth to a size greater than 4 cm, cell necrosis, spindling of tumor cells, cytologic atypia, high mitotic activity, high nucleus-to-cytoplasm ratio, and local recurrence. The presence of any mitotic activity is the best predictor of aggressive tumor behavior [1,4].

Immunohistochemical findings of a positivity rate of more than 50% for p53 and more than 10% for the Ki-67 index have been reported to be significantly correlated with malignancy [4]. The tumor mass in this patient was considered to be a benign GCT because it was less than 4 cm in size, showed no cell necrosis, had low mitotic activity, and did not express p53 and Ki-67. Furthermore, no evidence of recurrence was observed at the 1-year follow-up.

Usually, GCTs are detected incidentally, but they may be symptomatic, especially when the anorectal region is involved; symptoms include perianal discomfort and bleeding.

Because GCTs are invariably benign, adequate local excision is effective for both diagnosis and treatment of anorectal GCTs; careful follow-up should be performed after excision because of the risk of recurrence [1,3,4].

Despite the low risk of malignancy and only occasional instances of recurrence, careful follow-up, including repeat endoscopy, should be considered after local excision of these tumors, particularly due to the rare involvement of the perianal region.

Management of malignant is not clearly defined by the rarity of their occurrence in the literature and the lack of adequate follow-up, but wide excisional margin is preferred and addition of radiation is will be considered.

Aksoy et al. [10] reported additional chemotherapy and/or radiation did not alter disease-free survival or overall survival in 9 patients with local recurrence or metastasis. However, Khansur et al. [1] reported a case malignant GCT of the subcutaneous chest wall with excision and adjuvant radiation (unknown dose) without evidence of disease at 5-years follow-up.

Despite the undefined treatment options, it is reason-

able to offer a surgical excision with the addition of radiation to patients diagnosed with malignant GCT, and close follow-up should be advocated.

When a GCT is located close to the squamous epithelium, as in the case of this patient, it can induce pseudoepitheliomatous hyperplasia of the overlying epithelium; this can be occasionally mistaken for squamous-cell carcinoma, if an inadequate biopsy is obtained [4]. Therefore, pathologists and clinician should pay attention to the diagnosis and management of perianal tumors.

In conclusion, we reported first case of synchronous GCTs of the perianus and the chest wall to draw attention to a differential diagnosis of perianal tumor for GCT, and malignant potential must be considered during therapeutic procedures and follow-up.

## CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

## ACKNOWLEDGEMENTS

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