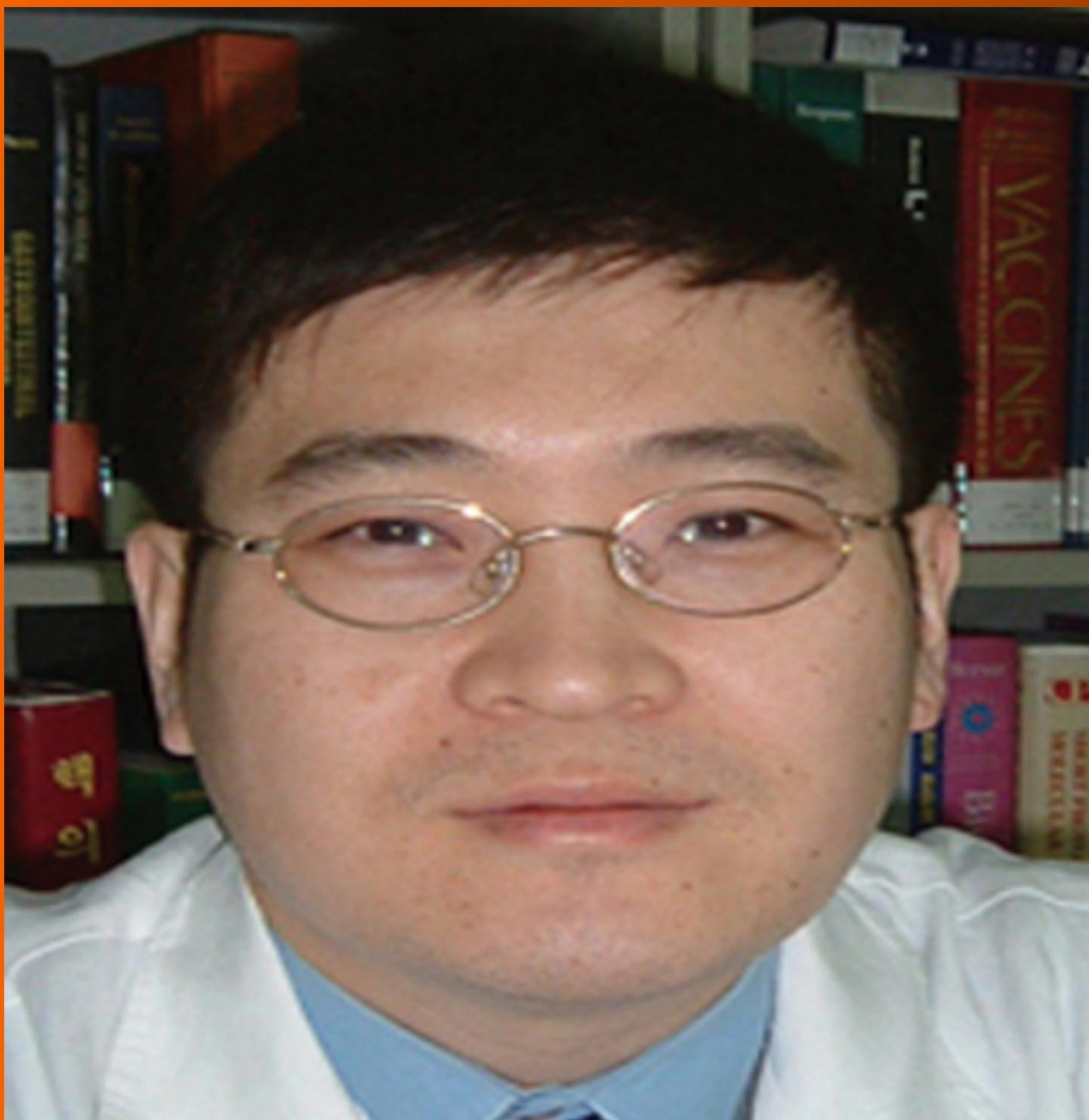


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Primary neuroendocrine tumor in the presacral region: A case report

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Abstract

BACKGROUND

Primary neuroendocrine tumors (NETs) in the presacral region are extremely rare, some of which are caused by other primary tumors or metastatic rectal carcinoids. Nevertheless, cases of NETs have been increasing in recent years. This report describes the first primary neuroendocrine tumor in the presacral region that was found at our hospital within the last five years.

CASE SUMMARY

The patient was identified as a 36-year-old woman with a presacral mass and pelvic floor pain. A digital rectal examination revealed a presacral mass with unclear margins and obvious tenderness. Magnetic resonance imaging (MRI) demonstrated a 57 mm × 29 mm presacral lump. An ultrasound-guided needle biopsy confirmed a well-differentiated neuroendocrine tumor. No other primary

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or metastatic tumors were found.

CONCLUSION

Comprehensive consideration of our case report and literature reported by others suggests that a conclusive diagnosis of NETs should be based on computed tomography/MRI and pathological examinations. The treatment of primary NETs in the presacral region mainly relies on surgical procedures with follow-up.

Key words: Neuroendocrine tumor; Presacral; Biopsy; Case report

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Core tip: This case highlights the need to include neoplastic diseases in the differential diagnosis of any potentially benign perianal abscess. A 36-year-old Asian woman was initially diagnosed with perianal abscess. Ultrasound-guided needle biopsy confirmed a well-differentiated neuroendocrine tumor, and then the patient underwent a sacrococcygeal tumor resection. A whole-body emission computed tomography scan was performed after the patient was discharged from the hospital, which revealed that she had no high expression of somatostatin receptors in the pleural cavity, abdominal cavity, or pelvis. There was no evidence of metastatic disease and no systemic symptoms or signs of carcinoid. These results confirmed that this was a primary presacral neuroendocrine tumor of level G2. The patient was advised to receive regular follow-up with her physician and magnetic resonance imaging.

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INTRODUCTION

Neuroendocrine tumors (NETs) are tumors derived from neuroendocrine cells and peptidergic neurons. Research from the Surveillance Epidemiology and End Results in the United States shows that the incidence of NETs has been increasing. A crude rate of occurrence has been estimated to be 5.85/100000 yearly, globally^[1]. Although NETs occur mostly in the gastrointestinal tract (about 60%) and the broncho-pulmonary tract (about 30%), any part of the body may act as the primary site^[2-4]. The clinical manifestations of NETs mainly depend on the type of hormones produced^[5]. NETs are generally classified according to the Ki67 index and the mitotic index based on the 2010 World Health Organization classification of tumors of the digestive system. NETs are divided into three levels: (G1) mitotic index < 2/10 high-power fields and/or Ki67 index ≤ 2%; (G2) mitotic index 2-20/10 high-power fields and/or Ki67 index 3%-20%; and (G3) mitotic index > 20/10 high-power fields and/or Ki67 index > 20%. G1 and G2 identify NETs, and G3 identifies neuroendocrine carcinomas. Over 5 years, the survival rates of patients diagnosed with G1, G2, and G3 NET were 95.7%, 73.4%, and 27.7%, respectively. The prognosis for treatment is based on the degree of differentiation, with the higher degree leading to a better prognosis after treatment^[6].

This study reports a case of primary well-differentiated NET in the presacral region. This case is an infrequent case of NET without any other primary tumor or metastatic tumor. In addition, we summarize our results regarding the diagnosis and review the literature on presacral NETs for collecting a comprehensive overview of these very rare tumors.

CASE PRESENTATION

Chief complaints

Anal discomfort for 1 mo.

History of present illness

A 36-year-old Asian woman had symptoms including a lump and pelvic pain. On March 11, 2018, she suffered from progressive pain. A pelvic computed tomography scan revealed an elliptical mass in the right side of the adnexa and in the left pelvis; this round mass, about 25 mm × 35 mm, appeared as circular-shaped soft tissue between the rectum and sacrum. A routine blood test revealed a white blood cell count of $11.34 \times 10^9/L$ and neutrophil percentage at 84.3%. The patient then visited our outpatient department due to the intolerable pain in the sacrococcygeal region, and we diagnosed it as a suspected perianal abscess.

History of past illness

Two years before her enrollment, the patient suffered from pelvic pain due to an accidental falling and did not accept any imaging examination or therapy.

Personal and family history

The patient was previously healthy and her family history was unremarkable.

Physical examination upon admission

There was no swelling between the anus and the sacrococcyx, but a mass which was full and had an unclear boundary and obvious tenderness could be touched. The internal anus was not obviously abnormal, and no blood was detected when we did digital rectal examination.

Laboratory examinations

Analysis of peripheral blood showed a neutrophil percentage of 88.01% and 27.3 ng/mL of neuron-specific enolase.

Imaging examinations

Magnetic resonance imaging (MRI) identified a 57 mm × 29 mm presacral mass, and margin and separation were visible (Figure 1). Transanal endoscopic ultrasound revealed a non-homogeneous echo-poor area 6 cm away from the sacral coccyx and in the direction of the sacrum, and a hyperechoic periosteum with a clear membrane but no abundant internal blood flow (Figure 2). On April 24, 2018, the patient underwent a sacrococcygeal tumor resection. The tumor was located in the anterior rectum 5 cm from the anus (Figure 3A). No tumor tissue remained after the resection.

The final pathology report showed that the tumor was a well-differentiated neuroendocrine tumor with a Ki67 index of 30% (Figure 3G). No nerve or lymphatic invasion was observed. Immunohistochemistry tested positive for neural cell adhesion molecule (CD56) (Figure 3E), CK2 (Figure 3F), and Syn (Figure 3B).

FINAL DIAGNOSIS

There was no evidence of metastatic disease and no systemic symptoms or signs of carcinoid syndrome (Figure 4). These results confirmed that this was a primary presacral neuroendocrine tumor of level G2. The patient was advised to receive regular follow-up with her physician and MRI.

TREATMENT

On March 11, 2018, a small amount of pus was withdrawn during surgery, and the transanal endoscopic ultrasound showed that this was a solid mass. According to the patient's symptoms and signs, an abscess incision was performed with antibiotic therapy and an ultrasound-guided needle biopsy confirmed a well-differentiated neuroendocrine tumor. On April 24, 2018, the patient underwent a sacrococcygeal tumor resection.

OUTCOME AND FOLLOW-UP

One month later, a whole-body emission computed tomography (ECT) scan was performed after the patient was discharged from the hospital. The whole-body ECT scan did not show high expression of somatostatin receptors in the pleural cavity, abdominal cavity, or pelvis.

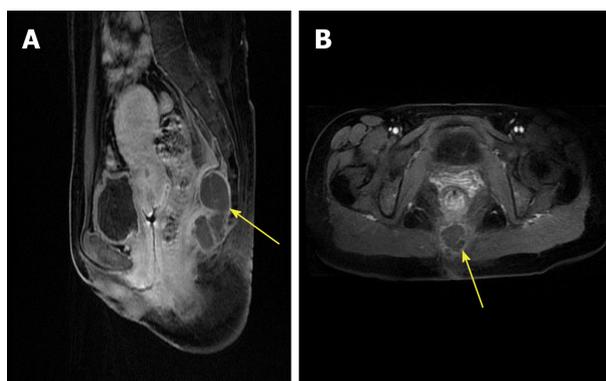


Figure 1 Magnetic resonance imaging scans. A: Sagittal view demonstrating a 57 mm × 29 mm presacral mass, with margin and separation being visible; B: T2STIR-weighted axial MRI image showing high intensity of the tumor.

DISCUSSION

The number of reported NET cases has increased in past decades because of improvements in diagnostic modalities and surveillance^[7,8], but primary NETs in the presacral region are extremely rare. The sacrococcygeal tumors usually remain clinically silent for a long period of time. A physical mass and pelvic pain are often discovered in the context of nerve root compression. For adults, primary tumors arising in this area include chordomas, myxopapillary ependymomas, paragangliomas, schwannomas, liposarcomas, and chondrosarcomas, *etc.*^[9,10] Some NETs can also result from the metastasis of rectal carcinoids^[11].

As shown in this case, a colonoscopy was not performed and a gastrointestinal tract NET was not considered since NETs in the presacral region are very rare, the patient's previous symptoms aligned with those of an abscess, and the doctor had little experience in this kind of tumor. Along the lines, masses in the presacral region are difficult to pre-diagnose and are reported as NETs by biopsy much later. The current methods for diagnosing NETs include: (1) Somatostatin-receptor nuclear imaging; (2) CT/MRI; and (3) biopsy. NETs are then classified according to the Ki67 index and the mitotic index. At the same time, NET phenotypes can be divided into functional and non-functional tumors based on the absence or presence of hormones and the presence of hormone-related symptoms^[5]. While hormone-related markers were not detected in the current case, it was unclear whether the patient suffered from hormone-related symptoms. Consequently, it was also difficult to distinguish whether the tumor was functional or non-function in this case.

Studies have shown that somatostatin-receptor nuclear imaging should be performed as a baseline preoperative test to detect whether a strong SSRT2A immune response is observed in tumor tissues^[12]. In this case, the whole-body ECT did not show high expression of somatostatin receptors in the pleural cavity, abdominal cavity, or pelvis.

For treatment, surgery with follow-up is the main form of treatment for NETs in the presacral region. In addition, somatostatin analogs are appropriate initial therapies for most patients with unresectable metastatic NETs for the control of carcinoid syndrome and inhibition of tumor growth. SSAs are typically selected as the first-line systemic therapy^[13-15]. In this case, the symptoms of recovery were pain relief and reduced mass size after pus withdrawal. With the mass remaining in the presacral region and a biopsy likely necessary to determine the pathological tendency, there was a risk for exacerbation and the patient needed a sacrococcygeal tumor resection. Nevertheless, the entire body ECT did not show high expression of somatostatin receptors in the pleural cavity, abdominal cavity, or pelvis. Therefore, there was no evidence of metastatic disease and no systemic symptoms or signs of carcinoid syndrome in the end.

Among the 11 cases of primary tumor in the presacral region that we found to be previously reported, the tumors were located mostly within the sacrum^[16-26], with one case located at the anorectal junction^[18]. Seven of the eleven patients were male. This prompted us to consider the possibility of a relationship between hormones and tumor location (Table 1)^[16-26]. Of note, five were associated with a retrorectal cystic hamartoma which was also known as "Tailgut cyst"^[16,18,19,24,25]. From those, three originated from a carcinoid tumor, one from a sacrococcygeal carcinoid^[22], one from an anterior tibial neuroendocrine tumor^[17], and one from a poorly differentiated large cell carcinoma^[26].



Figure 2 Transanal endoscopic ultrasound revealing a non-homogeneous echo-poor area 6 cm away from the sacral coccyx and in the direction of the sacrum and hyperechoic periosteum with clear membrane. No abundant internal blood flow was observed.

The main symptoms of the tumor were pain and the presence of a mass, which are also reported in [Table 1](#). Biopsies generally showed positive tumor markers, but not every case had detected SSRT2A. Surgery was the main treatment. If metastasis was observed, adjuvant therapy was needed mainly for SSAs.

The average follow-up time of reported cases was 41.8 mo^[16-26]. Most of the patients were alive at the time of the clinical follow-up control. Patients with well-differentiated presacral NETs that only showed a local disease often showed a good prognosis. There was even a patient who had been followed for 28 years after radiotherapy^[17]. However, patients with distant metastases need more investigation.

CONCLUSION

In summary, primary NETs in the presacral region are very rare, and the clinical presentation often involves pain and the presence of a mass. The imaging performance of these tumors is indistinguishable from that of the other tissue masses in this area, making diagnosis difficult. Currently, the most common diagnostic method for NETs is still based on biopsy. If biopsy confirms a well-differentiated NET, a colonoscopy is needed to exclude metastatic disease. In terms of treatment, surgical excision is needed to remove the primary tumor, and follow-ups are necessary to prevent recurrent attacks and metastases.

Table 1 Eleven cases of sacrococcygeal region tumor published in PubMed

Ref.	Year	Sex/age	Size (cm)	Associated anomalies	Symptoms	Biopsy	Treatment	Follow-up
Jehangir <i>et al</i> ^[18]	2016	M/71	NA	Tailgut cyst	No	CD56 ⁺ AE1/AE3 ⁺ CgA ⁺	Surgery	5 Y
Lokesh Bathla <i>et al</i> ^[19]	2013	F/46	6×6×6	Tailgut cyst	Constipation	Syn ⁺ NSE ⁺	Surgery	6 M
Fanny <i>et al</i> ^[17]	2009	M/72	NA	Neuroendocrine tumor	Pain	Neurosecretory granules ⁺	Radiation-therapy	28 Y
Stefano <i>et al</i> ^[16]	2010	F/73	3.9 × 3.2	Tailgut cyst	Pain	Chromogranin ⁺ Syn ⁺	Surgery	10 M
Luong <i>et al</i> ^[22]	2005	M/37	7	Sacrococcygeal carcinoid	Pain	NA	SSAs	NA
Mathieu <i>et al</i> ^[23]	2005	F/49	NA	Carcinoid tumor	Mass	NA	NA	NA
Krasinet <i>et al</i> ^[21]	2001	F/40	5 × 5	Carcinoid tumor	Mass pain	Chromogranin ⁺ Keratin ⁺⁺	Chemotherapy + radiotherapy	4 Y
Theunissen <i>et al</i> ^[26]	2001	F/51	7 × 5 × 5	Poorly differentiated large cell carcinoma	Pain	NA	Chemotherapy	NA
Prasad <i>et al</i> ^[25]	2000	F/36-69	20- 12	Hamartoma	Pain Bleeding	Syn ⁺ SSRT2A ⁺	Surgery	NA
Oyama <i>et al</i> ^[24]	2000	M/52	22	Tailgut cyst	Mass	Syn ⁺	NA	NA
Fiandaca <i>et al</i> ^[20]	1988	F/35	NA	Carcinoid tumor	NA	NA	NA	NA

Sph: Synaptophysin; cg A: Chromogranin A; NSE: Neuron-specific enolase; NA: Not available.

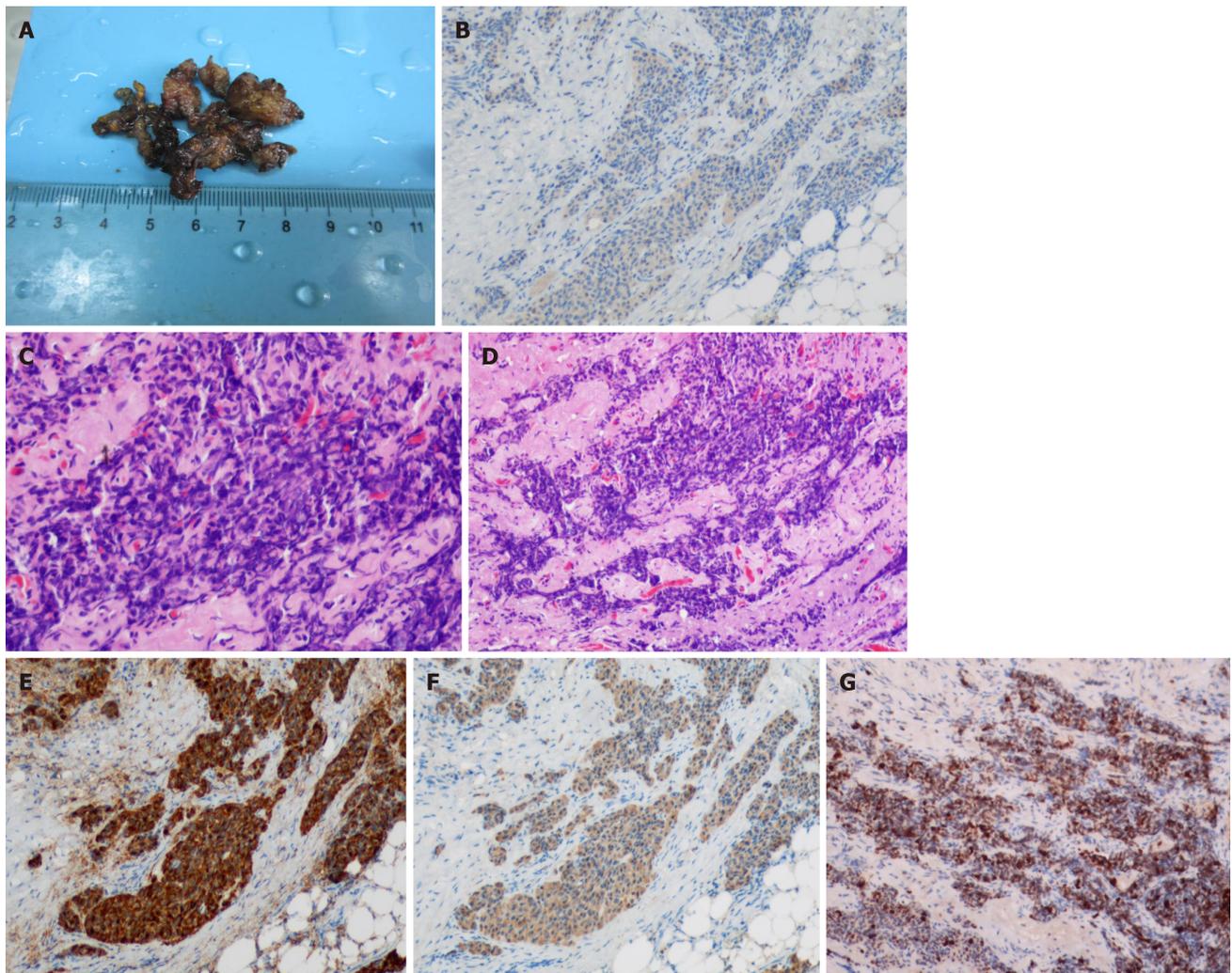


Figure 3 Final pathological images of the tumor. A: The mass removed by resection; B: The tumor component tested positive for the marker Syn ($\times 100$); C: The tumor stained using H&E ($\times 100$); D: The tumor stained using H&E ($\times 200$); E: The tumor component was diffusely positive for the marker CD56 ($\times 100$); F: The tumor component was positive for CK ($\times 100$); G: The tumor's Ki67 index was 30% ($\times 100$).



Figure 4 Two months after the resection, only the small fistula was not healed.

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