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Gastrointestinal schwannomas: CT findings to differentiate benign and malignant schwannomas

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Abstract
Purpose: To describe the computed tomography (CT) findings for the differentiation of gastrointestinal (GI) benign and malignant schwannomas.
Methods: Five cases of benign gastric schwannomas (GS) and two cases of malignant small intestine (SI) schwannomas identified over a five-year period were retrospectively reviewed by CT. We reviewed the CT findings with regard to tumor size, contour, margin, growth pattern, enhancement pattern, the presence or absence of necrosis, and perilesional lymph nodes.
Results: Benign GS appeared as submucosal tumors with the CT features of round, well-defined, exophytic or mixed growth patterns and homogeneous progressive enhancement. However, malignant SI schwannomas appeared as slightly lobulated, well-defined, exophytic mixed density mass with heterogeneous progressive enhancement. Irregular patchy necrotic density and perilesional lymph nodes were seen in SI schwannomas.
Conclusions: The typical CT findings of benign GI schwannomas manifested as round, well-defined, exophytic or mixed growth patterns and homogeneous progressive enhancement. In contrast, malignant schwannomas appear as lobulated, well-defined, exophytic mixed density mass with heterogeneous progressive enhancement. In addition, irregular patchy necrotic density and perilesional lymph nodes are characteristic features for malignant schwannomas.

Keywords: Gastrointestinal schwannomas, computed tomography, benign, malignant

Introduction
Schwannomas, also known as neurilemomas or neurinomas, are usually benign, slow-growing neurogenic tumors originating from nerve cells that have Schwann cell sheaths. They are common in the central nervous system or peripheral nerves of the body. Gastrointestinal (GI) schwannomas are very rare mesenchymal tumors, and stomach is by far the most common site accounting for 60-70% of all GI schwannomas followed by the colon, and the rectum [1, 2]. The occurrence of GI schwannomas in small intestine (SI) is extremely rare as reported by literature [2]. Gastric schwannomas (GS) account for only 0.2% of all gastric tumors and 4% of all benign gastric neoplasms [3]. GI schwannomas are usually misdiagnosed as other mesenchymal neoplasms preoperatively. Malignant transformation of GI schwannomas is extremely rare, and only few cases have been reported till date [4, 5]. Owing to their rarity, the CT findings of GI schwannomas have not been fully described. The interpretation of CT findings and differentiation from malignant schwannomas, which can metastasize, are clinically important. In this study, we retrospectively reviewed the CT findings of seven cases of GI schwannomas in an effort to differentiate malignancy and other similar tumors.

Materials and methods
1. Patients
We retrospectively reviewed the CT findings of five cases of benign gastric schwannomas and two cases of malignant small intestine schwannomas identified over a five-year period (2018-2023) in our hospital. The institutional review board of hospitals approved the study and did not require additional informed patient consent for reviewing the patient's medical records and images. We also reviewed English-language literature on GI schwannomas based on PubMed records.

2. CT protocol
The CT examinations were performed in all patients with a MDCT scanner in a single institution. Multi-detector row CT images were obtained with a beam collimation of 64 × 0.625 mm, a pitch of 0.984, 5 mm slice thickness, and 5 mm reconstruction intervals. Unenhanced CT and contrast-enhanced CT scans were performed during a single breath hold with patients in a supine position. The scanning ranged from the dome of the diaphragm to the symphysis pubis. All patients received intravenous contrast material (90 mL) as a bolus at the rate of 3 mL/s, and the CT images were obtained during arterial, portal venous, and delayed phases at 30, 60, and 180 seconds after contrast material injection, respectively.

3. CT evaluation
The CT images were retrospectively evaluated by consensus of two experienced radiologists who were unaware of the final diagnosis of the GI mass. The CT features were analyzed with regard to the morphological features of the masses, such as size, contour (round or lobulated), margin (well-defined or ill-defined), growth pattern (exophytic, endoluminal or mixed), enhancement pattern (homogeneous or heterogeneous, progressive or not), presence or absence of necrosis (i.e., intralesional low attenuation area with no

enhancement), and perilesional lymph nodes (considered positive if the shortest diameter was greater than 10 mm). The masses were also evaluated for presence or absence of hemorrhage, calcification, ulceration, adjacent organ involvement, distant tumor metastasis, and ascites.

4. Immunohistopathological study

All four patients were treated surgically and specimens were fixed in 10% buffered formalin and embedded in paraffin. Hematoxylin-eosin (H&E) stained slides were reviewed on all cases by experienced pathologists. Immunohistochemical studies were performed for S100 protein, Vimentin, CD117, CD34, Smooth muscle actin (SMA), desmin, human melanoma black 45 (HMB45), DOG-1.

Results

1. Clinical features

All seven patients with GI schwannomas were enrolled in our study; the population consisted of four females and three males with mean age 59 years (age range 49-71 years). Four GS patients presented with upper abdominal pain, one GS patient found as an incidental finding on medical checkup for other unrelated diseases, and two SI schwannomas patient presented with lower abdominal pain with palpable mass. There was no any history of neurofibromatosis in all seven patients. All five GS patients were treated with gastric wedge resection and the SI schwannomas were treated with surgical removal of the tumor along with a perilesional lymph node and part of the involved SI; bladder resection + cystostomy for bladder adhesions.

2. CT findings

The CT findings of GI schwannomas for all seven patients are summarized in Table 1. Three GI schwannomas were located in the greater curvature of gastric antrum (Fig 1, 2), two in the lesser curvature of gastric antrum (Fig 3) and two were located in the small intestine (Fig 4). The tumor size ranged from 1.5 to 12 cm, with a median size of 4.6 cm. All five GS were round (Fig 1, 2, 3), and SI schwannomas were slightly lobulated (Fig 4). All tumors had well-defined margins. The growth patterns of the tumors were diverse with exophytic (n = 6) (Fig 1, 3, 4) and mixed (n = 1) (Fig 2). Non-enhanced CT scan revealed homogeneous soft tissue mass in all five GS patients (Fig 1A, 2A, 3A), and heterogeneous mass in SI schwannomas (Fig 4A). Contrast-enhanced CT scan showed a homogeneous enhancement pattern in all five GS patients (Fig 1B, C, D, 2B, 3B), and heterogeneous enhancement pattern in SI schwannomas (Fig 4B, C, D). The CT value of the plain scans ranged from 21 to 56 Hounsfield units (HU, mean 36 HU), arterial phase with a CT value of 41 to 67 HU (mean 54.25 HU), venous phase with a CT value of 67 to 81 HU (mean 79 HU), and delayed phase with a CT value of 92 to 98 HU (mean 96.25 HU) in all cases demonstrating progressive enhancement pattern. SI schwannomas had irregular patchy necrosis (Fig 4), perilesional lymphadenopathy (Fig 4C) and small amount of pelvic fluid (Fig 4D). It also showed compression of the adjacent bowel and bladder adhesions. There was no necrosis, perilesional lymphadenopathy and ascites in all GS cases. No schwannomas had CT evidence of hemorrhage, calcification, and ulceration. In our study, all cases were misdiagnosed as gastrointestinal stromal tumors (GISTs) preoperatively and received surgery.

3. Immunohistopathological findings

Histopathological studiesin our series revealed abundant spindle-shaped cells (Fig 2C, 3C). On immunohistochemical studies, tumor cells showed strong immunopositivity for S-100 protein (Fig 2D, 3D), and Vimentin and immunonegativity for CD117, CD34, Smooth muscle actin (SMA), desmin, DOG-1 in all cases which were compliable to the diagnosis of GI schwannomas. Additionally, tumor cells of case 6 and case 7showed mitosis and nuclear atypia and immunopositivity for human melanoma black 45 (HMB45), and the perilesional lymph nodes resected during operation also revealed neoplastic cells. Immunohistopathological examinations of the lesion specimen in the first five cases were consistent with benign gastric schwannomas, whereas the last two casesshowed SI pigmented malignant schwannomas.

Table 1: CT findings of the gastrointestinal schwannomas

Case	Sex/Age (year)	Location	Size (cm)	Contour	Margin	Growth pattern	Enhancement pattern	Necrosis	Perilesional lymph nodes
1	M/58	Gastric antrum (GC)	1.5	Round	Well-defined	Exophytic	Homogeneous progressive	No	No
2	F/49	Gastric antrum (GC)	2.2	Round	Well-defined	Exophytic	Homogeneous progressive	No	No
3	F/71	Gastric antrum (GC)	2.1	Round	Well-defined	Mixed	Homogeneous progressive	No	No
4	F/62	Gastric antrum (LC)	2.6	Round	Well-defined	Exophytic	Homogeneous progressive	No	No
5	M/63	Gastric antrum (LC)	2.4	Round	Well-defined	Exophytic	Homogeneous progressive	No	No
6	M/54	Small intestine	12.0	Lobulated	Well-defined	Exophytic	Heterogeneous progressive	Yes	Yes
7	F/56	Small intestine	9.4	Lobulated	Well-defined	Exophytic	Heterogeneous progressive	Yes	Yes

LC = Lesser curvature; GC = Greater curvature

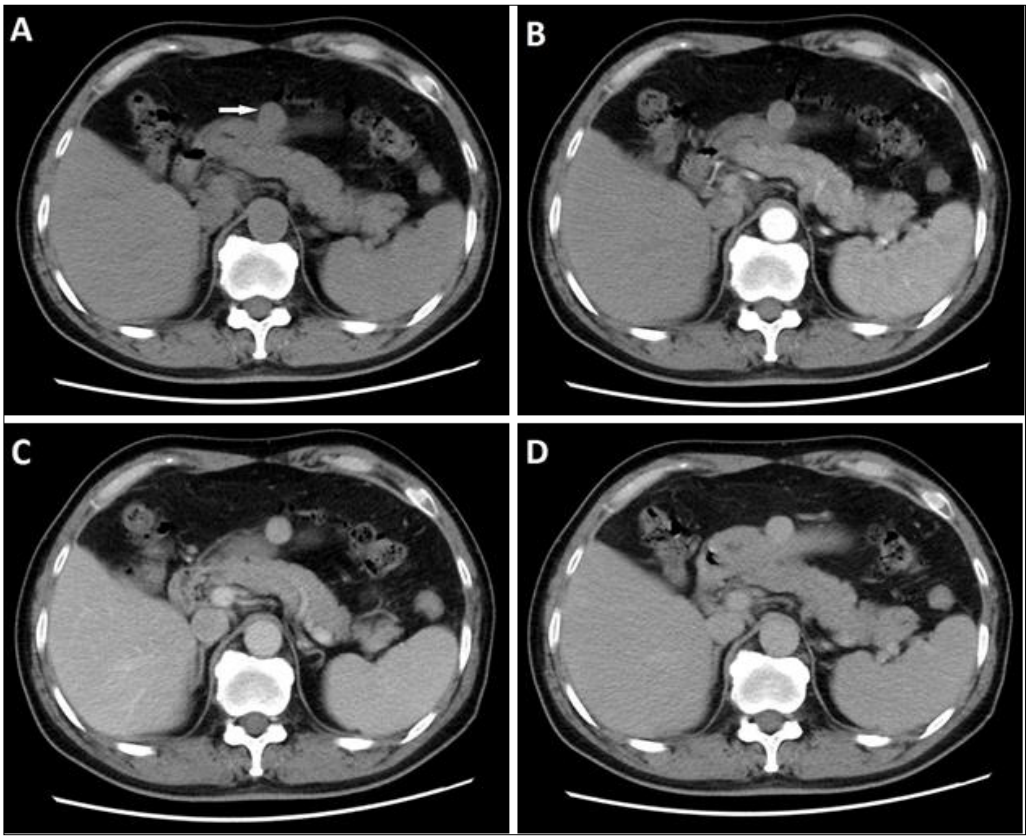

Fig 1: A 58-year-old man with benign gastric schwannoma (case 1). Non-enhanced CT scan of the abdomen shows a round, well-defined, exophytic soft tissue mass arising from the greater curvature of the gastric antrum measuring 1.5 cm in diameter (arrow) (A). Contrast-enhanced CT scan shows a homogeneously enhanced lesion in the arterial phase (B), venous phase with gradual enhancement of the lesion (C), and delayed phase with persistent enhancement of the lesion (D). The CT value of the plain scan (A), arterial (B), venous (C), and delayed (D) phases are 21 HU, 59 HU, 84 HU, and 98 HU respectively.

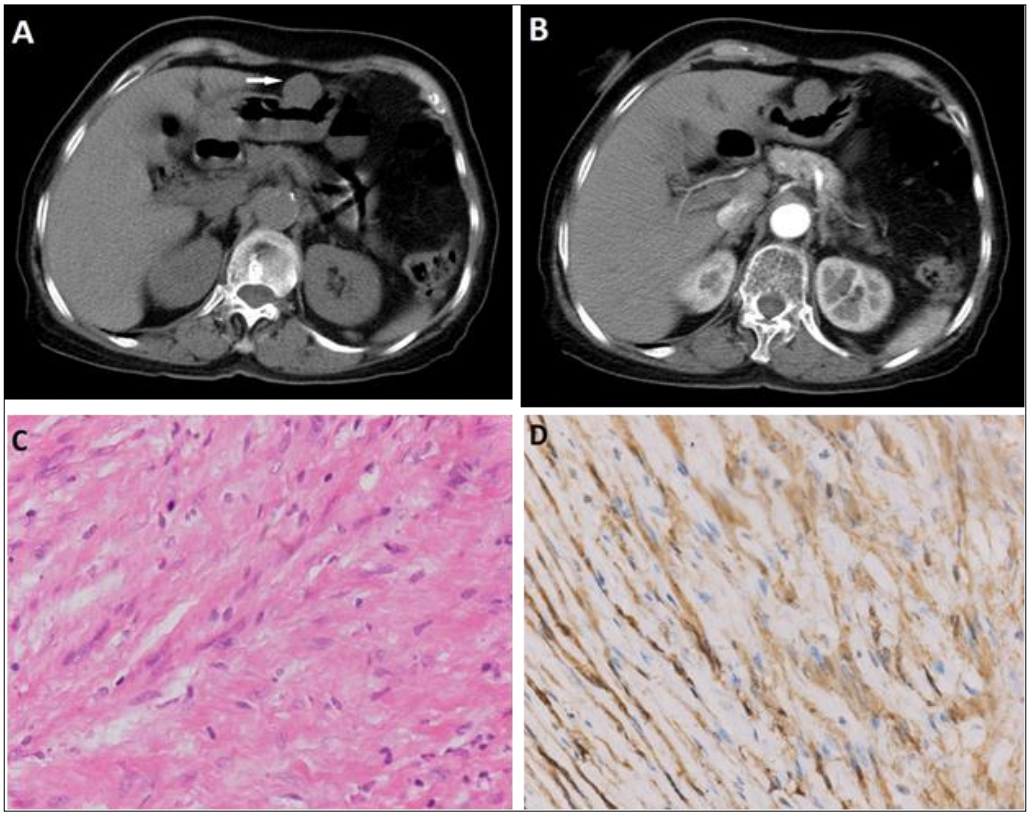

Fig 2: A 71-year-old woman with benign gastric schwannoma (case 2). Non-enhanced CT scan of the abdomen shows a round, well-defined, exophytic soft tissue mass arising from the greater curvature of the gastric antrum measuring 2.1 cm in diameter (arrow) (A). Contrast-enhanced CT scan shows a homogeneously enhanced lesion in the arterial phase (B). Histopathological study revealed abundant spindle-shaped cells (H&E stain, ×40) (C). Immunohistochemical staining (×40) shows that tumor cells are strongly positive for S-100 protein (D).

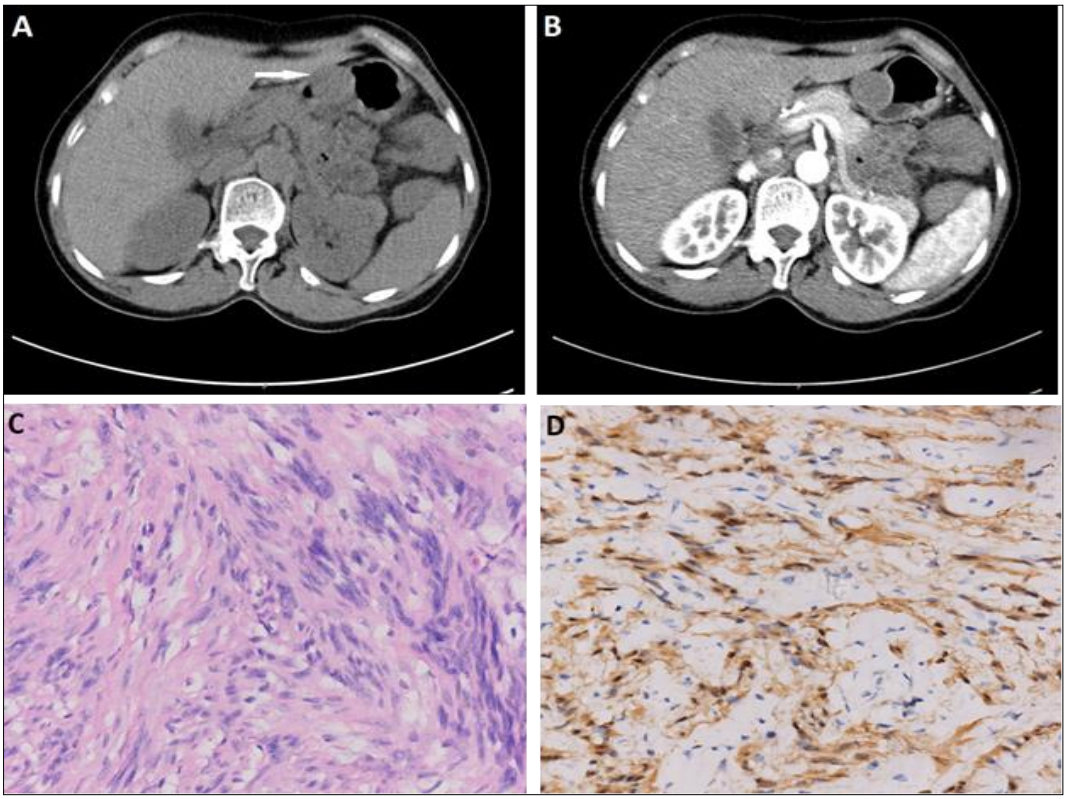

Fig 3: A 62-year-old woman with benign gastric schwannoma (case 3). Non-enhanced CT scan of the abdomen shows a round, well-defined, exophytic soft tissue mass arising from the lesser curvature of the gastric antrum measuring 2.6 cm in diameter (arrow) (A). Contrast-enhanced CT scan shows a homogeneously enhanced lesion in the arterial phase (B). Histopathological study revealed abundant spindle-shaped cells (H&E stain, ×40) (C). Immunohistochemical staining (×40) shows that tumor cells are strongly positive for S-100 protein (D).

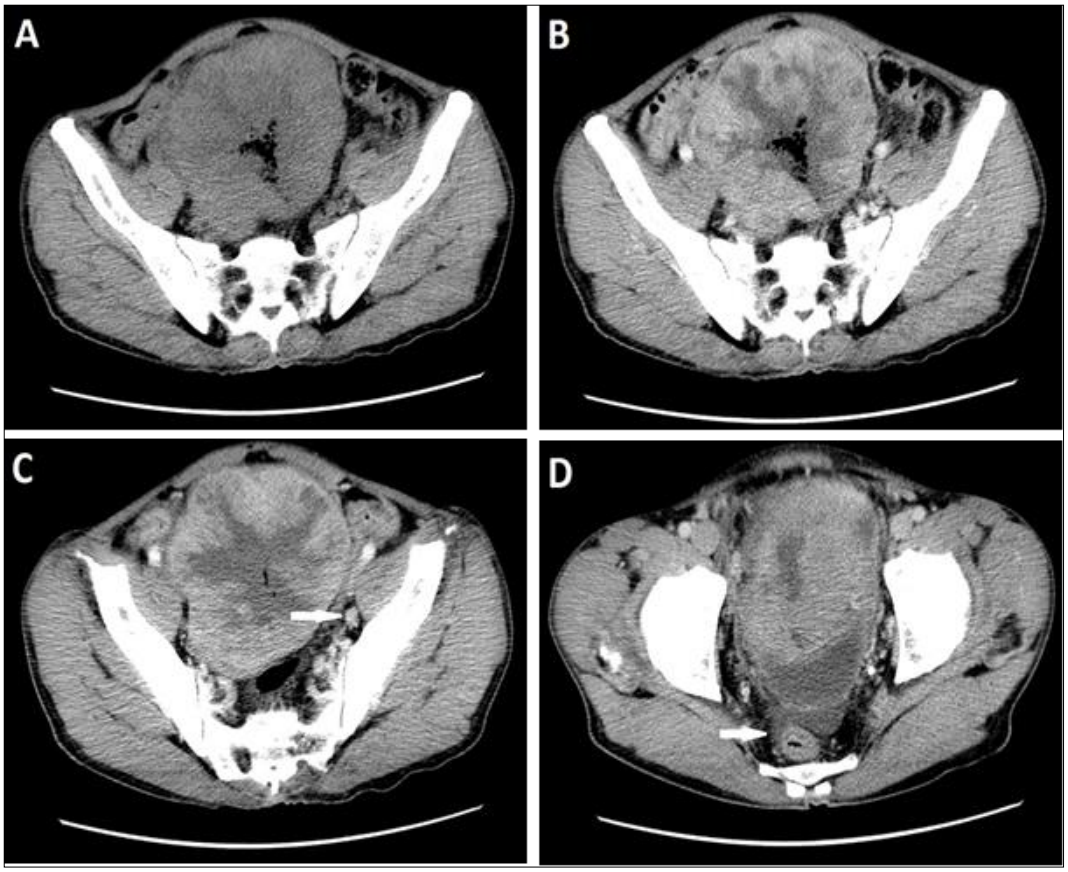

Fig 4: A 54-year-old man with malignant small intestine schwannoma (case 4). Non-enhanced CT scan of the abdomen shows a slightly lobulated, well-defined, exophytic mixed density mass with irregular patchy necrosis arising from the small intestine measuring 12 cm in diameter (A). Contrast-enhanced CT scan shows a heterogeneously enhanced lesion in the arterial phase (B), venous phase with gradual enhancement of the lesion showing perilesional lymph node (arrow) (C), delayed phase with persistent enhancement of the lesion showing small amount of pelvic fluid (arrow) (D). It also shows compression of the adjacent bowel and bladder adhesions.

Discussion

Schwannomas are neurogenic tumors of ectodermic origin that is usually solitary and benign. They usually involve peripheral nerves. Gastrointestinal (GI) schwannomas are very rare mesenchymal tumors; the stomach is the most commonly involved site followed by the colon, then rectum and the small intestinal schwannomas are exceedingly rare [1, 2]. Malignant GI schwannomas are even rarer, reported with a high potential for recurrence and metastasis [6]. GI schwannomas arise from Schwann cells of the nerve sheath of Auerbach's plexus or, less commonly, from Meissner's plexus [7]. According to previous reports, GI schwannomas can occur at any age but are most frequently observed in the fifth to sixth decade of life with a female predominance [8]. Our study also shows the mean age of 59 years with female predominance. Patients can cause non-specific symptoms, although many are detected incidentally. Sometimes, a palpable mass may also be noted when the tumor size is large and exophytic, as in our last two cases. Patients with benign GI schwannomas have good prognosis after surgical resection, however malignant schwannomas, which can metastasize, are clinically important.

CT is the imaging modality of choice for GI tumors [9]. Due to the rarity of GI schwannomas, CT findings have not been fully described. In our study, we have described the CT findings in an effort to differentiate benign and malignant GI schwannomas. The most common site for GI schwannomas in our series was the greater curvature of gastric antrum (3/7), followed by the lesser curvature of gastric antrum (2/7), and small intestine (2/7). Documented CT findings of typical benign gastric schwannomas frequently include round, well-defined, exophytic or mixed growth patterns, homogeneous attenuation on unenhanced CT and homogeneous progressive enhancement on contrast-enhanced scans. On unenhanced CT scan of the abdomen, most schwannomasin our series showed round (5/7), well-defined (7/7), exophytic (6/7), and homogeneous attenuation (5/7) without hemorrhage, calcification, and ulceration. Only malignant tumors in SI showed irregular patchy necrotic density, perilesional lymph nodes and small amount of pelvic fluid. The heterogeneity in SI tumors may just have occurred due to the size of SI tumors. In our study, contrast-enhanced CT scan showed homogeneous progressive enhancement pattern (5/7) in benign schwannomas and heterogeneous progressive enhancement pattern (2/7) in malignant schwannomas. These CT findings are compliable to the previous studies and illustrate that malignant schwannomas are different from benign schwannomas in that they show lobulated contour, heterogeneous progressive enhancement pattern, irregular patchy necrotic density, and perilesional lymph nodes. However, these CT findings consistent with malignant schwannomas have also been described in some benign gastric schwannomas in previous reports [10]. Recently, Ji *et al.* [11] reported CT findings of eight benign cases of gastric schwannomas as ovoid (7/8), well-defined (8/8), exophytic (4/8) or mixed (3/8) growth patterns, homogeneous progressive enhancement (7/8) and heterogeneous progressive enhancement pattern in a largest tumor (11.4 cm) with necrosis and perigastric lymph nodes. These perigastric lymph nodes were reactive inflammatory lymph nodes without evidence of neoplastic cells or malignancy. However, perilesional lymph nodes resected in the last two cases of our series revealed neoplastic cells.

The preoperative differential diagnosis of GI schwannomas includes gastrointestinal stromal tumors (GISTs), lymphomas, leiomyomas and GI adenocarcinomas. The main differential diagnoses of GI schwannomas are GISTs, which are the most common mesenchymal neoplasms of the GI tract. In our study, all cases were misdiagnosed as GISTs. Although GISTs have a heterogeneous appearance on CT due to presence of hemorrhage, necrosis and cystic change, 8-13% of these tumors may appear as homogeneous tumors mimicking the GI schwannomas [12, 13]. Levy *et al.* [13] have reported that GISTs have a peripheral enhancement pattern in 92% of their cases due to presence of hemorrhage, necrosis and cystic change within the tumors. In our study, homogeneous progressive enhancement pattern in benign GS and heterogeneous progressive enhancement pattern in malignant SI schwannomas might aid in differentiating them from GISTs, which commonly show marked heterogeneous enhancement on the arterial phase and decreases in the venous and delayed phases. Choi *et al.* [14] have reported that perilesional lymph nodes are much more common in GS than GISTs and pathologically confirmed that these lymph nodes include numerous inflammatory cells.

Lymphomas and leiomyomas of stomach may have a CT appearance of homogeneous enhancement pattern similar to those of gastric schwannomas. However, homogeneous lymphadenopathy and usually a relative range of involvement of stomach without a well-defined capsule are frequently observed in gastric lymphomas, which are helpful distinguishing features of lymphoma [15]. Gastric leiomyomas mainly occur in the cardia, frequently show endoluminal growth, and have lower enhancement than GS [16]. GI adenocarcinomas are frequently infiltrative and interrupt the mucosal margins with surface ulceration accompanied by heterogeneous-attenuation perilesional lymphadenopathy as a result of lymphatic metastasis [17].

Magnetic resonance imaging (MRI), positron emission tomography (PET) and Ultrasonography (USG) have all been used to evaluate GI neoplasms, but the features are non-specific for schwannomas. Gastric schwannomas show hyperintensity on T2-weighted and diffusion-weighted MRI and accumulation of [^{18}F]-fluorodeoxyglucose (FDG) on PET [18]. Endoscopic ultrasonography (EUS) may reveal marginal hypoechoichalos in GS, corresponding to lymphoid cuffs. Endoscopically, GISTs may also show the same feature of marginal halos [19].

Definitive diagnosis of GI schwannomascan only be made on the basis of histological examination and immunohistochemical studies of the resected specimen. The characteristic histopathologicalfeature includes abundant spindle-shaped cells with a characteristic peripheral lymphoid cuff [20]. The immunohistochemical features are strong immunopositivity for S-100 proteinand Vimentin and immunonegativity for CD117 (KIT protein), CD34, Smooth muscle actin (SMA), desmin, DOG-1 which are found in true GISTs and smooth muscle tumors [21]. In our study, all GI schwannomas showed similar immunohistopathological findings, and additionally, tumor cells of malignant schwannomas showed mitosis and nuclear atypia and immunopositivity for human melanoma black 45 (HMB45), and the perilesional lymph nodes resected during operation also revealed neoplastic cells. The treatment of choice is complete surgical excision of the tumor when no metastasis has occurred.

There were some limitations to our study that should be mentioned. Firstly, our study was performed retrospectively. Secondly, a small number of patients were included because of the rarity of GI schawannomas. A further limitation is that our study did not differentiate the CT findings of GI schawannomas to other GI diseases. In the future, these limitations will be needed to better understand features of GI schawannomas.

Conclusions

The CT findings of benign GI schwannomas typically manifested as round, well-defined, exophytic or mixed growth patterns and homogeneous progressive enhancement. In contrast, malignant schwannomas appear as lobulated, well-defined, exophytic mixed density mass with heterogeneous progressive enhancement. In addition, irregular patchy necrotic density and perilesional lymph nodes are characteristic features for malignant schwannomas. Malignant GI schwannomas are extremely rare with a high potential for metastasis. Radiologically, it is difficult to distinguish GI schwannomas from other mesenshymal tumors due to overlapping imaging features. Further research is necessary to better describe the radiological features of GI schwannomas.

Compliance with Ethical Standards

Conflict of Interest: The authors declare that they have no conflict of interest.

Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee.

Informed consent: The institutional review board of our hospital approved the study and did not require additional informed patient consent for reviewing the patient's medical records and images.

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