CASE REPORT

Schwannoma of stomach

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Abstract

Gastrointestinal mesenchymal tumours are a group of tumours originating from the mesenchymal stem cells of the gastrointestinal (GI) tract. Digestive tract Schwannomas are rare mesenchymal tumours occurring most frequently in the stomach. We report the case of a 40-year-old woman with gastric Schwannoma located at the posterior wall of the antrum.

Keywords: Schwannoma, Stomach, Mesenchymal tumour.

Introduction

Gastrointestinal mesenchymal tumours are a group of tumours originating from the mesenchymal stem cells of the gastrointestinal (GI) tract, consisting of GI stromal tumours (GIST), leiomyomas or leimyosarcomas and Schwannoma.[1,2]

Gastric Schwannomas are benign, slow-growing and usually asymptomatic and may present with epigastric pain, dyspepsia, bleeding and gastric outlet obstruction.[3-5] Histologic diagnosis of gastric Schwannoma is confirmed by positivity for S100 protein and negativity for smooth muscle actin and CD117.[3,5] These tumours have an excellent prognosis and surgical removal is sufficient treatment.[5] GI schwannoms are considered to be rare and distinctively different neoplasms from conventional Schwannomas that arise in soft tissue or the central nervous system (CNS). Digestive tract Schwannomas show distinctive histologic features that separate them from conventional Schwannomas. They do not show a nuclear palisading pattern that is usually present in conventional Schwannomas.[1,6-8]

Histologically, gastrointestinal Schwannomas are S-100 protein-positive spindle cell tumours with a microtrabecular pattern, peripheral lymphoid cuffing, and occasional germinal centres.[1.3] These tumours also have an excellent prognosis and surgical removal is sufficient treatment.[5] We present the case of a 40-years-old woman with gastric Schwannoma admitted with epigastric pain, dyspepsia and a mobile epigastric mass.

Case Report

A woman aged 40 was admitted with dyspepsia, epigastric pain, nausea and heartburn for the preceding six months in February 2013. She had history of histamine-2 receptor blocker usages with no complete response.

Physical examination showed a palpable mobile epigastric mass. It was stretched in epigastrium to the right upper quadrant. There was no other pathological findings in other parts of the abdomen. There was no sign of hepatomegaly or splenomegaly.

Ultrasonography revealed a well-defined homogenous mass between stomach and liver. Computed tomography (CT) scan showed a tumoural mass (5×6cm in size) located between liver and greater curvature of stomach. No other abdominal organ abnormality was present (Figure-1).

Routine haematology and biochemistry tests were normal. Surgery was performed and a round-shaped submucosal mass of 6cm in diameter located at the posterior wall of antrum was resected with 2cm safe margin. Postoperative course of the patient was favourable. The specimen received in the pathology department consisted of a well-circumscribed, firm, whitish-gray mass with overlying mucosa measuring 5.8cm in diameter on cut whorl pattern. Microscopic examination of haematoxyline-eosin stained sections showed proliferation of spindle-shaped cells with normochromic elongated to oval shape nuclei and

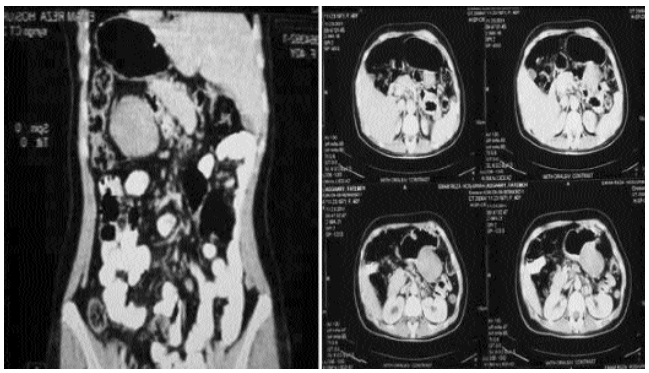

Figure-1: Computed tomography finding.

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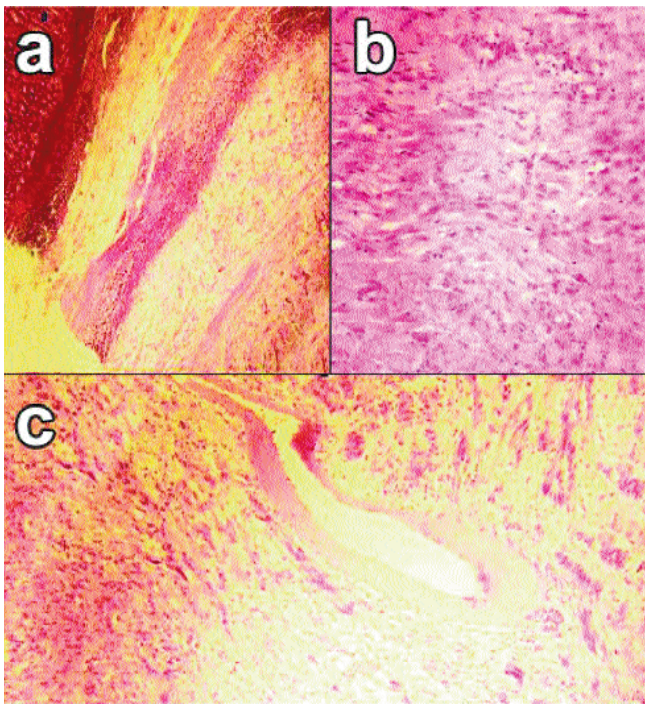

Figure-2: Histological finding, A. Low-magnification feature of gastric Schwannoma with a prominent cuffing by lymphoid aggregates, B. Spindle-shaped cells with normochromic elongated to oval shape nuclei and eosinophilic cytoplasm (H&E staining ×400), C. Hyalinised vessels are present (×400).

eosinophilic cytoplasm arranged in whorl and microtrabecular pattern and also fascicles. Areas of myxoid changes, mononuclear inflammatory cells infiltration and vascular hyalinisation were seen (Figure-2). Immunohistochemistry (IHC) staining for S100 protein was positive and for smooth muscle actin (SMA), CD117 and CD34 were negative. These features were consistent with Schwannoma.

Discussion

Digestive tract Schwannoma was initially reported in 1988.[6] The clinicopathologic and immunohistochemical review was made of 24 cases of distinctive nerve sheath tumours located in the GI tract. The tumours were microscopically evident in the presence of peripheral lymphoid cuffing and benign nuclear atypia. The tumours arose in the muscularis propria of the stomach in 23 cases and in the ascending colon in 1 case. The patients included 9 men and 15 women whose ages ranged from 36 to 78 years (average, 58). No recurrence developed in any patient who underwent resection. Although positive immunostaining for S100 protein, Leu 7 antigen, and laminin might support the Schwannian nature of these tumours, but the positive immunoreactivity for glial fibrillary acidic protein (GFAP) indicated the possibility of a mesenteric plexus origin. The pattern of S100 protein immunostaining differed from that seen in cases of GI stromal tumours associated with von Recklinghausen's neurofibromatosis or that noted in cases of conventional leiomyomas. We propose that these tumours be designated as benign Schwannoma of the GI tract.

Typical histologic features of gastric Schwannoma are proliferated spindle cells usually arranged in a micotrabecullar-microfollicular pattern and peritumoral lymphoid cuff.[7,8] Our case showed these features too.

Gastric Schwannoma differs histologically from soft tissue Schwannoma by lacking the encapsulation and rarity of nuclear palisading, xanthoma cells, vascular hyalinisation and dilatation,[7] but in our case , vascular hyalinisation and dilatation were seen.

Gastric Schwannomas have cellular structures similar to other mesenchymal tumours such as GI stromal tumour, leiomyoma and leiomyosarcoma and by the aid of IHC staining. Earlier studies[8,9] reported the differences between these spindle cell tumours. Positive desmin and muscle actin stains indicate leiomyoma, or leiomyosarcoma, positive CD34 and CD117 indicate GIST and positive S100 indicate Schwannoma.[1,3,7,8]

Thus, in our case, positivity for S100 and negativity for other markers confirmed the diagnosis of Schwannoma.

Gastric Schwannomas should be accurately distinguished from GISTs which may be malignant or have malignant potential. Schwannomas are biologically benign and patients have an excellent prognosis after surgical resection.[1,3,8] Also, GI Schwannomas are distinctly different from conventional soft tissue and CNS Schwannomas.

Gastric Schwannomas occur more frequently in the fifth to sixth decades of life and commonly in female patients[2,5] as was our case. They are often asymptomatic and can be discovered incidentally. The most common presenting symptom is an episode of upper GI bleeding followed by abdominal pain.[2,4] The typical endoscopic appearance of gastric Schwannoma is a round protruding submucosal mass with overlying ulcerated mucosa. False negative results of endoscopic biopsy would be encountered because normal mucosa overlies the submucosal lesion.[2] CT scan can demonstrate the extent of invasion and help to determine the appearance of a benign lesion. In one recent study, the most consistent CT feature of patients was the homogenous pattern of tumour attenuation on both unenhanced and intravenous (IV) contrast-enhanced scans.[10] Surgical resection, including wedge resection, is sufficient

because prognosis for patients with solitary Schwannoma of the stomach is excellent and malignant transformation of this lesion is rare. A study reported one case with malignant gastric Schwannoma.[3] Another study[11] had 33 patients. The patients were 16 men and 17 women whose ages ranged from 27 to 81 years (median 52.6 years). Tumour size ranged from 10 to 120mm in diameter. Follow-up in 23 cases from 6 months to 13 years showed no recurrences or metastases. Microscopically, all tumours were composed of spindle cells with focal epithelioid cells in four cases. In all cases except one, there were peripheral cuff-like lymphoid aggregates. Immunohistochemically, tumours were strongly positive for S100 protein and vimentin. The tumours were variably positive for nestin (78.8%; 26/33) and GFAP (63.6%; 21/33), three tumours had CD34+ cells, but all were negative for CD117, alpha-smooth muscle actin and desmin. Ultrastructurally, the tumours were composed of elongated spindle-shaped cells with prominent parallel membranous structures. One study[12] reported that with the stomach being the most common site of origin in the GI tract, Schwannomas represent 6.3% of gastric mesenchymal tumours and only 0.2% of all gastric tumours. They are usually located in the middle third of the stomach along the lesser curvature. The majority of the tumours are usually encased by intact mucosa and principally involve the submucosa and muscularis propria, without invading adjacent structures. About half of them show central ulceration.

Due to the difficulty of establishing a definite preoperative diagnosis, but also in order to prevent possible complications such as bleeding or pyloric stenosis, surgical resection should be considered the treatment of choice in patients with gastric Schwannoma. The size and location of the tumour, as well as its relation to the surrounding organs are important factors in determining the type of operation. Local extirpation, wedge resection, partial, subtotal or even total gastrectomy, are all acceptable operations.[13] Laparoscopic techniques can also be used.[14] Recurrence rates are generally very low.

Conclusion

As it is difficult to establish a definite pre-operative diagnosis of a Schwannoma, surgical resection should be the treatment of choice. This prevents complications as bleeding and pyloric stenosis besides providing complete removal of the diseased tissue and achieving negative margins.

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