Gastric Schwannoma: a rare Schwann cell tumour of the GI tract

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Schwannomas are benign, slow-growing, encapsulated nerve-sheath tumors arising from Schwann cells, which are responsible for the myelin sheath in the peripheral nervous system.1 Schwannomas are most commonly found in the cranial vault, involving the myelin-forming cells of the 8th cranial nerve, in a condition called vestibular neuroma. In rare cases, they can also occur in the GI tract, usually in the stomach.3 GI schwannomas are often non-encapsulated, but well demarcated.3

Gastric schwannomas account for only 0.2% of all gastric tumors and 4% of all benign gastric neoplasms.5 The peak incidence is in the 4th and 5th decade of life, usually involving females.5,6 They can also occur in children, and can be malignant in rare cases.7 The majority of patients have remained asymptomatic for extended periods of time, with most cases discovered incidentally on imaging or laparotomy.3,8 Symptomatic patients most often present with ulcerations and upper GI bleeding, and a palpable mass may be present if the tumor grows exophytically.5 Malignant transformations are exceedingly rare, but have been reported.5

CASE

The patient is a 34-year-old female who presented to the ER with RLQ pain and lower back pain persisting for the past 10 days. Her vital signs were stable with a normal white cell count.

CT-scan of her abdomen showed a normal appearing appendix. Fluid was seen in the pelvis, possibly secondary to a ruptured ovarian cyst. There was an incidental finding of a mass that appeared to be arising from the stomach. The mass appeared intramural and measured 5.1 cm x 4.4 cm. An ultrasound was done to look at lesions in the liver which were felt to be hemangiomas.

The patient’s pain was somewhat controlled with morphine, although it was still present. No cause of pain was determined. She was discharged from the emergency department with Percocet and scheduled to return for an endoscopic ultrasound (EUS) approximately 4 weeks later.

Upon return for the EGD, the patient’s pain was much improved and she discontinued all her pain medications. The EGD showed a submucosal mass 5 cm below the gastroesophageal junction. The patient was referred to GI to for endoscopic ultrasound (EUS).

When seen by GI one month later, the patient revealed a prior history of anxiety disorder and was being treated with Paxil. She also had constitutional symptoms of anorexia and reported a 45-pound weight loss. An upper endoscopy was performed and the gastric mass was visualized along the lesser curvature of the stomach. The mass felt firm when probed.

On endoscopic ultrasound, the mass appeared hypoechoic but had evidence of hyperechogenicity foci. It measured 5.2 cm x 3.3 cm in its largest dimensions and was well demarcated, originating from the muscularis propria of the stomach. There was evidence of a perigastric lymph node as well as two celiac lymph nodes. A fine needle aspiration (FNA) for cytology as well as a core biopsy for histopathology was performed on the primary mass and one of the nodes. The mass was not endoscopically resectable.

Microscopic evaluation of lesion cells revealed a palisaded growth configuration, with elongated spindled nuclei and pointed ends. No mitosis or necrosis was seen. Immunohistochemistry was positive for S-100; negative for CD117, CD34, desmin, muscle specific actin and caldesmon. These findings were consistent with a diagnosis of schwannoma.

An ultrasound was done approximately 6 months later to reassess the mass, which appeared unchanged. She is currently being followed with annual CT-scans; no surgery is planned.

DISCUSSION

This is an interesting case of a gastric mass found incidentally for the presentation of right lower quadrant pain.

Briefly, the presenting pain was relatively severe and never explained. Appendicitis is the most likely cause of acute RLQ pain, but our patients’ appendix appeared normal on CT-scan. There was also suspicion of a ruptured ovarian cyst based on fluid seen in the pelvis on ultrasound. However, the ovaries were normal and this...
diagnosis was somewhat inconsistent with the patient’s 10-day history of pain: fluid from an uncomplicated ruptured ovarian cyst is normally reabsorbed in 24 hours, with the pain resolving in a few days.\(^{11}\) The pain did resolve, and further investigations focused on the incidental gastric mass.

The differential diagnosis of a submucosal gastric mass is broad and includes gastrointestinal stromal tumor (GIST), leiomyoma, leiomyosarcoma, and schwannoma, among others.\(^{12}\) Upper endoscopy is useful in examining epithelial tumors; however, it is inadequate for examining tumors below the mucosa.\(^{13}\) In contrast, EUS is effective in visualizing submucosal tissue adjacent to the gastric wall and can be used to precisely guide needle biopsies.\(^{13}\)

It is important to differentiate between gastric schwannomas and other mesenchymal tumors, particularly the main differential of GIST, which has a higher chance of being malignant. The diagnosis of a GIST would mandate surgical resection if possible. Imatinib is used both as an adjuvant therapy when the GIST is completely excised and palliatively for unresectable or metastatic disease.\(^{14,15}\) It was found that virtually all cases of GIST would exhibit malignant behavior if followed, and metastatic cases can be effectively treated with imatinib mesylate, which improves survival from <50% to >90% at 1 year.\(^{14,15}\) In comparison, schwannomas remain benign in nearly all cases, and are rarely symptomatic.\(^{3}\) As the tumor enlarges, it displaces the nerve to the periphery, preserving its function.\(^{16}\) Symptoms can result from the enlarging submucosal tumor restricting circulation to the mucosa, leaving it ischemic and prone to ulceration, bleeding, and damage by gastric acidity.\(^{17}\)

Imaging modalities such as CT, ultrasound, and MRI can provide limited but useful information. Schwannomas often appear uniform on CT, which can distinguish it from leiomyomas and leiomyosarcomas; ultrasound with a sufficient resolution can determine the tumor’s layers of origin, and MRI can map the precise location of the tumor and the displacement of its surrounding organs.\(^{18,19,20}\) These findings may also be helpful in explaining the cause of symptoms as well as determining the best approach to treatment, particularly in surgical cases.

Due to the similar spindle-shaped appearance of schwannomas and other gastric mesenchymal tumors, it can be difficult to distinguish between them using light microscopy.\(^{21}\) Grossly, they resemble leiomyomas and other stromal tumors: solitary, firm, smooth, white in color, and well circumscribed.\(^{3}\) Advances in immunohistochemistry allowed for a more precise differentiation: in one study, 24 out of 306 gastric spindle-cell tumors were diagnosed as schwannomas using immunohistochemistry, but only 9 were diagnosed as schwannomas using hematoxylin-eosin.\(^{22}\)

Immunohistochemical diagnosis of schwannoma is based on positivity for the S-100 protein, and negativity for CD117, CD34, desmin, and muscle specific actin.\(^{23}\) CD117 and CD34 are positive in GIST, and muscle specific proteins such as actin, desmin, and caldesmon are positive in smooth muscle tumors such as leiomyoma and leiomyosarcoma; all of which are negative in schwannomas.\(^{23}\) Our patient's results are congruent with this guideline and a definitive diagnosis of schwannoma can be made.

Surgical treatment for gastric schwannomas would involve complete resection of the tumor.\(^{24}\) For our patient, this would mean a gastrectomy: the complete or partial excision of the stomach. The post-operative prognosis is excellent\(^{5,3}\) and most patients return to their pre-operative quality of life.\(^{25,26}\) however, it is debatable whether or not surgery should be deemed necessary for benign, asymptomatic schwannoma patients. In our opinion, surgery should be reserved for either malignancy or for symptomatic cases, especially for difficult operations such as in our patient. Most of the literature on surgical outcome appears to be based on cases were diagnosed as schwannomas post-operatively; therefore, the benefits of surgery for known asymptomatic cases are unclear.

Taking into account the available information, we felt that it was best to follow our patient with annual CT-scans, and to defer any surgical intervention until her symptoms become unmanageable; or until the improbable event that her tumor becomes malignant.

REFERENCES

11. Blechman AN, Mann WJ. Evaluation and management of a ruptured ovarian cyst. Up2date. 2010