# Gastric Schwannoma in a Female Patient with Pulmonary Tuberculosis — A Clinicopathological Assessment and Diagnosis

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## Abstract

Schwannomas, or neurinomas, are generally benign, slow-growing, asymptomatic neoplasms originating from the Schwann cells of a nerve sheath. As a part of spindle cell mesenchymal tumours, schwannomas arising from the gastrointestinal tract (GIT) are unusual; however, when they occur, the most common site involved is the stomach, which represents 0.2% of all gastric tumours. We report the case of a 35-year-old female patient with a history of pulmonary tuberculosis presenting with a large palpable abdominal mass reaching up to the peritoneal cavity. The initial clinical impression was a tuberculous abdominal mass, a cyst, or a teratoma. However, intra-operative findings during a subtotal gastrectomy revealed an exophytic gastric serosal mass, which suggested a gastrointestinal stromal tumour (GIST). Post-operative histopathological findings showed a fascicular arrangement of neoplastic spindle cells with pallisading nuclei that showed intense positivity for S-100 protein, and were negative for CD117 and desmin in immunohistochemistry studies. These results confirmed the final diagnosis of a gastric schwannoma.

*Keywords:* gastrointestinal stromal tumours, immunohistocytochemistry, neurilemmoma, stomach, medical sciences

## Introduction

Gastrointestinal mesenchymal tumours are a group of tumours that originate from the mesenchymal stem cells of the gastrointestinal tract (GIT) and consist of gastrointestinal stromal tumours (GIST), leiomyomas or leiomyosarcomas, and submucosal schwannomas (1). Histologically, these tumours form a spindle cellular pattern and were traditionally considered to be of smooth muscle origin; however, the immunohistochemical studies by Sarlomo-Rikala and Christopher showed the differences between these tumours by demonstrating positive desmin and muscle actin immunostaining in leiomyoma or leiomyosarcoma (2,3). No evidence of this positive desmin and muscle actin immunostaining was found in GIST because most of these tumours showed positive staining for CD117 and CD34, whereas S-100 positivity indicated schwannomas (2,3). Diagnosis of primary GIT schwannomas based on the S-100 positivity had been dubious until Daimaru et al. in the year 1992, presented a

series of well-documented cases, in which 24 out of 306 GIT spindle cell tumour cases were found to be schwannomas through immunohistochemistry (IHC). In contrast, only 9 of these cases were diagnosed as schwannomas when stained only with hematoxylin-eosin (H&E) (4). Some of the non-specific histological features supporting schwannomas include the presence of spindle cell tumours lacking epithelioid features and skeinoid fibres that have a peripheral cuff of lymphoid tissue and specific intracellular needle-shaped PAS-positive crystalloids (5,6). This marked difference shown by Daimaru demonstrates the high likelihood for misdiagnosing schwannomas as GISTs when IHC is not used (4). Gastric involvement, though most common among the GIT schwannomas, represents only 0.2% of all gastric tumours and 4% of all benign gastric neoplasms (7). Here, we present the case of a 35-year-old woman with a gastric schwannoma manifesting as a painless palpable mass in the abdomen, which was diagnosed using different immunohistochemical markers.

## Case Report

A 35-year-old female with pulmonary tuberculosis presented to the Sharif Medical College Hospital Lahore with a 6-month history of a progressively enlarging but painless abdominal mass. There were no associated symptoms except those related to tuberculosis, including loss of appetite and weight. She had been on anti-tuberculosis therapy for the previous 8.5 months. Her general physical examination was unremarkable, although crepitations and ronchi were heard in the left lung. Upon radiological assessment, both lung fields showed moderate pleural effusion and foci of calcification as a result of pulmonary tuberculosis. The abdominal examination showed a large, freely mobile, firm but smooth-surfaced, non-pulsatile mass that measured 12 x 10 cm and was dull on percussion. Laboratory investigations revealed an erythorcyte sedimentation rate titre three times higher than normal and a haemoglobin level of 8.2 g/dL. Radiographical survey using ultrasound followed by CT scan of the abdomino-pelvic area showed an enlarged, 15 x 13 x 8 cm, well-defined, solid, heterogeneous mass in the abdomen with foci of necrosis and a 15 mm area of dense calcification seen posteriorly. The mass was connected to the stomach but had no link with the bladder or the uterus. The first clinical impression of the surgeon was a disseminated lesion in the abdomen from pulmonary tuberculosis, a mesenteric cyst, or a teratoma. Neurofibromatosis 1 (NF-1) was ruled out by following the criterion suggested by the National Institute of Health (NIH), which specifies the identification of at least two of the seven "Cardinal Clinical Features". These features are as follows: 1) 6 or more café-au-lait macules over 5 mm in largest diameter in pre-pubertal individuals and over 15 mm in largest diameter in post-pubertal individuals, 2) 2 or more neurofibromas of any type, or 1 plexiform neurofibroma, 3) freckling in the axillary or inguinal regions, 4) optic glioma, 5) 2 or more Lisch nodules (iris hamartomas), 6) a distinctive osseous lesion, such as sphenoid dysplasia or thinning of the long bone cortex with or without pseudarthrosis, and 7) a first degree relative (parent, sibling, or offspring) with NF-1 by the above criteria. Initial manifestations most often occur in childhood. Clinical signs may be apparent at birth, however, in some affected individuals, signs may not develop until adulthood. Exploratory laparotomy was planned for the patient, and the pre-operative findings showed a well-circumscribed mass arising from the serosal surface of the distal stomach wall close to the greater curvature but separated from the liver, pancreas, and major abdominal vessels. The mass was protruding into the peritoneal cavity. A subtotal gastrectomy was carried out to remove the tumour capsule, which was gently and carefully dissected from the fascicles of the nerve. Additionally, adherent omentum and 13 enlarged adjacent draining lymph nodes were resected. Gastro-jejunostomy was performed, and the post-operative course was uneventful. The patient was discharged in stable condition within a week's time. The surgical specimen received by our pathology department was fixed overnight. Gross inspection the next day revealed an exophytic, yellow-tan, bosselated mass measuring 13 x 12 x 7 cm. The cut surface of the specimen was similar to fish flesh with whirling trabeculations (Figure 1). The mucosa overlying the tumour was unremarkable with a tiny, less than 1 cm, focus of ulceration over the tumour area. The tissue sections from the resected tumour specimen were embedded in paraffin blocks. Eleven sections of 4–7µm (five for H&E staining and 2 each for three immunohistochemical markers) were cut from the tumour tissue and collected on poly-L-lysine-coated slides for IHC. Sections from the lymph nodes were subjected to H&E staining only. Histological observations revealed dense lymphocytic infiltration at the periphery of the tumour with predominantly Antoni type A areas. These areas consisted of fascicular arrangements of neoplastic spindle cells bearing pallisading nuclei with occasional pleomorphism interspersed within a loose collagenous matrix. Few foci exhibiting Antoni type B changes were also observed. No evidence of necrosis, haemorrhage, cystic degeneration, or any malignant change was seen, along with moderate to severe reactive hyperplasia of all lymph nodes

These findings supported the diagnosis of a GIST or a gastric schwannoma (Figure 2a & 2b). Differential diagnosis was confirmed by applying immunohistochemical markers using the standard 'Avidin Biotin Peroxidase' method. The primary antibodies (AbD Serotec, Germany) employed were purified concentrated mouse monoclonal antibodies to Desmin (HCA071A), CD117 (MCA2598), and the polyclonal antibody to S-100 (AHP385T) protein. The morphologic and immunohistochemical features of the tumour, namely an intense S-100 protein positivity and negativity for desmin and CD117, resemble the diagnostics used in previously reported gastric schwannomas (Figure 3).

**Figure 1:** Cross-section of the surgical specimen, which shows an enlarged yellow-tan solid mass with whirl like trabeculations originating from the serosal surface of the stomach.
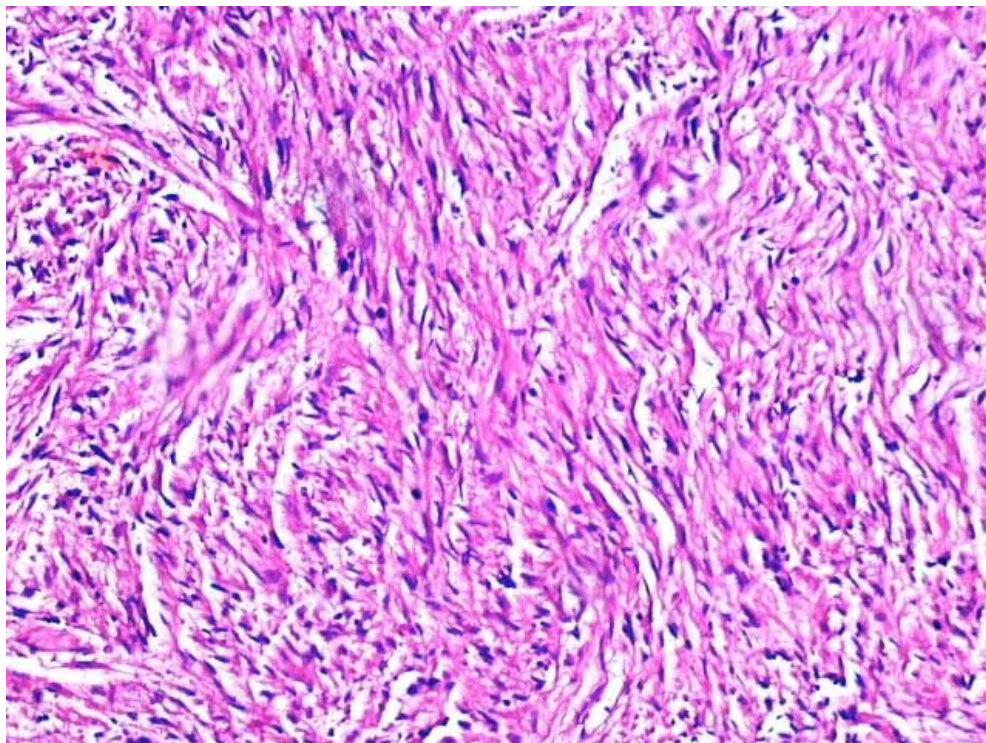


**Figure 2a:** Photomicrograph of the gastric tumour, which shows interlacing bundles of spindle cells, pallisading nuclei with nuclear atypia and no mitosis, interspersed with collagenous strands (H&E, x200).
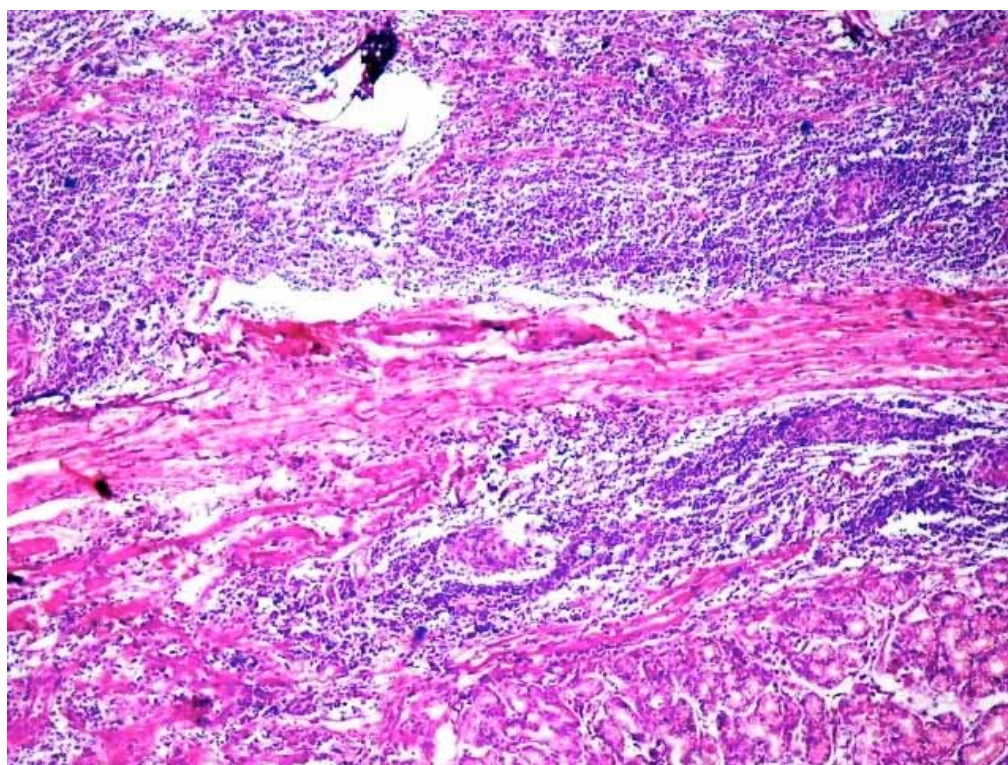
**Figure 2b:** Dense lymphocytic cuffing at the peripheral part of the tumour, which resembles a tumour of lymphoid origin (H&E, x100).

## Discussion

Schwannomas are the most common solitary, encapsulated, and slow-growing peripheral nerve-sheath tumours. Gastric schwannomas originate within the nerve sheath of Auerbach plexus or, less commonly, from Meissner plexus. Histologically, gastric schwannomas are composed of Schwann cells dispersed in a collagenous matrix (7). These tumours arise from the fundus, body, or antrum of the stomach and are commonly intramural, however, they can be extraluminal or endoluminal. Tumours vary from 0.5 cm to 11 cm in diameter and are spherical or ovoid with an occasional multinodular pattern (4,5,8,9). As the tumour enlarges, it displaces the nerve to the periphery of the tumour, preserving neural function. In our case, the tumour was characteristically exophytic, extending from the gastric wall to the abdominal cavity, and the differentiation of the schwannoma from other submucosal tumours was very difficult on pre-operative assessment.

These tumours occur more frequently in females in the fifth to sixth decade of life, although the patient in our study was quite young. These tumours are often asymptomatic and can be discovered incidentally. The most common presenting symptom is an episode of upper GIT bleeding with or without abdominal pain, which may be secondary to the growing submucosal mass. This mass compromises the blood supply to the gastric mucosa, which then ulcerates secondary to ischemia or a reduced tolerance to the gastric acidity (4,8,9).

Although conventional radiographical procedures can demonstrate the presence or extent of invasion, they cannot provide enough information in the differential diagnosis (10). Similarly, our preliminary clinical impression was different from the actual diagnosis. Surgical resection is the treatment of choice and the prognosis is excellent because malignant transformation is rare (4).

Concordant with our provisional microscopic diagnosis of GIST without employing immunostaining, Fujii reported about 4% gastric schwannomas among 150 GISTs when subjected to IHC (11). Prevot and colleagues also reported three cases (1 male, 2 females; age range of 56–74 years) of gastric schwannomas in which the tumours were well circumscribed but not encapsulated and had sizes ranging from 2 to 11 cm in diameter. A diffuse and intense positivity for vimentin and S-100 protein was detected in all cases along with a variable, sometimes focal positivity for glial fibrillary acidic protein and neuron-specific enolase (4).
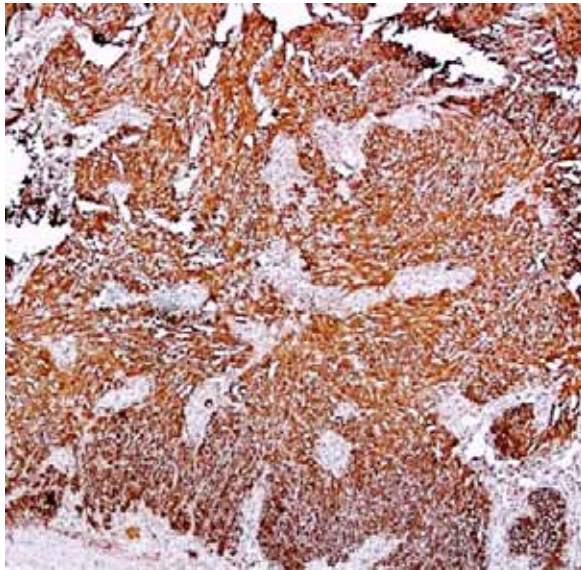
**Figure 3:** Immunochemistry shows the bundles of spindle cells that have a strong positivity for S-100 protein staining (brown colour) (S-100 stain, x200).

## Conclusion

Gastric schwannomas, though rare among the spindle cell tumours of GIT, can occur even at a young age and should always be confirmed by IHC, especially to rule out GISTs. A retrospective or a prospective series of GISTs should be analysed using different immunohistochemical markers in our population.

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