Intraabdominal Schwannomas: single-center experience

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OBJECTIVE: Intraabdominal schwannomas are rare benign tumors. In this study, we aimed to present our clinical experience in patients with intrabdominally located schwannoma.

MATERIAL-METHOD: Patients who received the diagnosis of intrabdominal schwannoma between 2011-2019 were retrospectively examined. Demographic and clinical characteristics, treatment methods, short- and long-term results and immunohistochemical characteristics of the patients were analyzed.

RESULTS: A total of 7 patients were included in the study. Four patients were female and three were male. The mean age was 51.5 (31-63) years. The most common clinical presentation was abdominal pain (57.1%). Tumor location was stomach (n=2), pelvic region (n=2), rectum (n=1), retropancreas (n=1), and left juxtadrenal space (n=1). Postoperative wound infection developed in one patient and pancreatic fistula complication was seen in one patient. Re-admissions to the hospital were due to anemia and pleural effusion in two patients. The mean tumor diameter was 6 cm (0.3-13 cm). All patients were S 100 strongly positive Mitoses / 50 HPFs (high power field), <2 Ki67 <3%. The mean follow-up period was 60 months. Currently, 5 patients are being followed without disease, 1 patient survives despite recurrence and 1 patient has died due to non-cancer reasons.

CONCLUSION: Intraabdominal schwannomas are rare tumors which most commonly exhibit gastrointestinal involvement. Since these tumors are mostly benign, the long-term prognosis of patients is good. Schwannoma should be kept in mind in the differential diagnosis of intrabdominal masses. Radical resections with high morbidity and mortality should be avoided if preoperative diagnosis is made.

KEY WORDS: Abdominal tumor, Mesenchymal tumor, Nerve sheath tumor, Schwannoma.

Introduction

Schwannoma is a benign tumor originating from Schwann cells in the peripheral nerve sheath 1. Schwannoma is a homogenous tumor and can occur in any tissue of the body. Although the most common localization is the head and neck region, it is rarely seen in the intraabdominal region. In the intraabdominal region, it is mostly seen in the gastrointestinal tract 1) GIS (Gastrointestinal System) schwannomas were first reported by Daimaru et al. in 1988 2. Gastrointestinal schwannomas are most commonly seen in the stomach (83%), small intestine (12%) and rarely localized in the colon and rectum. However, intraabdominal, pancreatic and retroperitoneal schwannomas are extremely rare 3-5. Gastrointestinal schwannomas are equally common in both sexes. It is most common between the ages of 50-60 and has the highest incidence in the sixth decade of life 6,7. Although most of the Schwannoma cases are benign and asymptomatic, there is a potential for malignant transformation and this is related to the size of the tumor 6.
Pancreatic schwannoma is a slowly growing, encapsulated, benign neoplasm, typically presenting in the peripheral epineurium of the sympathetic or parasympathetic autonomic fibers of the pancreas or branches of the Vagus nerve.

Schwannomas in the adrenal medulla are very rare. Retroperitoneal localized schwannomas occur especially in the juxta-adrenal space and differential diagnosis should be made with adrenal masses. It has been shown in the literature that approximately 0.5% to 5% of schwannomas are retroperitoneal.

Preoperative diagnosis of soft tissue schwannomas is difficult. Complete resection with negative surgical margin should be performed for definitive treatment.

A limited number of series have been reported in the literature on intrabdominal schwannoma. Our knowledge of these rare tumors is often based on case reports.

In this study, we aimed to present our clinical experience in patients operated on with the diagnosis of intraabdominal schwannoma in a tertiary referral center.

**Material-Method**

Patients with a diagnosis of schwannoma with intrabdominal localization, who were diagnosed between January 2011 and January 2019 in the General Surgery Clinic of Çukurova University Medical Faculty based on the combination of histological and immunohistochemical features, were included. Approval was obtained from the Local Ethics Committee 89/1 numbered and 14.06.2019 dated. Patient files and hospital information system records were examined and a database was created. The cases were analyzed retrospectively. Follow-up data were supported by telephone interviews with patients. Patients under 18 years of age and patients whose records could not be reached were excluded from the study.

Patients’ demographic characteristics, Body Mass Index (BMI), American Society of Anesthesiologists (ASA) score, presenting symptoms, tumor localization, histopathological and immunohistochemical features of the tumor, treatment procedures, intraoperative complications, postoperative complications, postoperative hospital stay, unplanned re-operation, postoperative 90-day mortality, 90-day unplanned re-admission, mean follow-up times and current clinical status were analyzed.

Anastomotic leakage was defined as the deterioration of anastomotic integrity detected by clinical and radiological imaging methods.

Wound infection was defined as superficial or deep surgical site infection in the incision according to the definition of Centers for Disease Control and Prevention (CDC).

Unscheduled re-operation was accepted as a surgical intervention under general, spinal or epidural anesthesia within 30 days after primary surgery, according to the definition of the National College of Surgeons National Surgical Quality Improvement Program (ACS-NSQIP).

Data were analyzed using IBM SPSS Statistics for Windows, version 24 (IBM Corp., Armonk, N.Y., USA).

<table>
<thead>
<tr>
<th>Case no</th>
<th>Age/Sex</th>
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<th>ASA</th>
<th>Clinical presentation</th>
<th>Tumor location</th>
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</thead>
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<tr>
<td>1</td>
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<td>49/F</td>
<td>28.1</td>
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<td>Stomach small curvature</td>
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<tr>
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<td>Pelvic</td>
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<td>59/M</td>
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<td>Stomach small curvature</td>
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<td>Retropancreas</td>
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<tr>
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<td>58/M</td>
<td>26</td>
<td>1</td>
<td>Constipation</td>
<td>Rectum</td>
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<table>
<thead>
<tr>
<th>Case no</th>
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<th>Intraoperative complications</th>
<th>Postoperative complications</th>
<th>Postoperative Length of Hospital stay(day)</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Partial Pancreatectomy+R-Y</td>
<td>–</td>
<td>Pancreatic Fistula</td>
<td>32</td>
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<tr>
<td>2</td>
<td>Mass excision + pancreojejunostomy</td>
<td>Sudden hypotension</td>
<td>–</td>
<td>13</td>
</tr>
<tr>
<td>3</td>
<td>Proximal Gastrectomy + Esophagogastrosomy</td>
<td>–</td>
<td>–</td>
<td>9</td>
</tr>
<tr>
<td>4</td>
<td>Cytoreductive Surgery + HIPEC</td>
<td>–</td>
<td>Wound infection</td>
<td>11</td>
</tr>
<tr>
<td>5</td>
<td>Wedge Resection</td>
<td>–</td>
<td>–</td>
<td>75</td>
</tr>
<tr>
<td>6</td>
<td>Debulking</td>
<td>–</td>
<td>–</td>
<td>1</td>
</tr>
<tr>
<td>7</td>
<td>Transanal Excision</td>
<td>–</td>
<td>–</td>
<td></td>
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</tbody>
</table>
Categorical measurements were summarized as numbers and percentages, and continuous measurements were summarized as mean and standard deviation (median and minimum-maximum where necessary).

**Results**

Seven patients, 4 female and 3 male, were included in the study. The mean age of the patients was 51.5 ± 10.7 (31-63) and the body mass index was 24.1 ± 3.4 (17.5-28.1). The most common clinical presentation was abdominal pain (57.1%). The tumor was mostly located in the stomach (28%) and the pelvic region localization was 28%. Demographic and clinical characteristics of the patients are shown in Table I.

Six patients underwent laparotomy and resection of tumor. One patient underwent transanal resection of rectal schwannoma. Intraoperative sudden hypotension developed in one patient and the operation was continued after treatment. One patient developed pancreatic fistula and one patient developed wound infection. The mean length of hospital stay was 11 ± 10 (1-32) days. Operative procedures and surgical morbidity of the patients are shown in Table II. None of the patients required reoperation or developed postoperative mortality. One patient had an unplanned re-admission due to pleural effusion and one patient due to anemia. Mean follow up was 60±23.7 (37-98) months. Five patients were followed-up without disease and one patient was still alive despite recurrence. One patient died of unrelated causes. Clinical outcomes of the patients are shown in Table III.

Mean tumor diameter was 6±4.5 (0.3-13) cm. Mitoses/50 (HPFs), <2 Ki67 <3% in all patients. The histopathological and immunohistochemical properties of the patients are shown in Table IV. Pathological examination of the stomach schwannomas is shown in Figs. 1, 2.

**Discussion**

Schwannomas are slow-growing encapsulated tumors originating from Schwann cells in the collagen matrix. Schwannoma was first reported by Verocay in 1910, and in 1935 Stout described its clinical and pathological features in detail. In 1988, Daimaru et al. first described the concept of gastrointestinal benign schwannoma and demonstrated its clinical, morphological and phenotypic features. Intraabdominal schwannomas are rare tumors and constitute 2.9% to 5.6% of mesenchymal tumors. Since intraabdominal schwannomas outside the gastrointestinal tract are rare, this study mainly focuses on the clinicopathological features of GIS schwannomas. Gastrointestinal schwannomas are most commonly seen in the stomach (83%), small intestine (12%) and rarely in the colon and rectum. According to the World Health Organization (WHO) classification of digestive system tumors, intestinal schwannoma belongs to the class of gastrointestinal mesenchymal tumors (GISTs). Some authors have classified intestinal schwannoma as a category of gastrointestinal autonomic neurogenic tumors (GANT). Gastrointestinal Schwannomas are more common in the sixth decade. Although some series in the literature indicate that incidence in both sexes are equal, there are also series in which female gender is more dominant. The mean age of our series was lower than the average age in the literature and female gender was dominant. Schwannomas are generally asymptomatic and the symptoms are not diagnostic. Mekras et al. found most of the cases in their series incidentally.
reported by Antonio et al., Schwannoma cases can be incidentally detected during radiological imaging conducted for other diseases 20. However, when symptomatic, it causes non-specific clinical symptoms. Bruneton et al. reported in their series that the most common symptoms were bleeding and abdominal pain 21. Shu, Zhenbo et al. reported that small bowel-derived Schwannomas presented with melena 22. In our series, the most common presenting symptoms of the patients were nonspecific symptoms such as abdominal pain, dyspepsia and constipation. The patient who had a mass of 13 cm presented with swelling in the right lower quadrant and had a palpable mass. Goh, Brian KP et al. reported in their series that intrabdominal Schwannomas most commonly had gastric origin, and less frequently colon, rectum and small intestine origin 11. Retroperitoneal and pancreatic cases have also rarely been reported in the literature 4,5. In our series, intrabdominal schwannomas were located in the small curvature of the stomach in 2 patients, rectum in 1 patient, pelvic in 2 patients, retropancreatic in 1 patient and left juxtadrenal region in 1 patient. Radiological image of retropancreatic tumor is shown in Fig. 3.

Table III - Clinical outcomes

<table>
<thead>
<tr>
<th>Case no</th>
<th>Reoperation</th>
<th>Postoperative 90-day mortality</th>
<th>90-day readmission</th>
<th>Mean follow up (month)</th>
<th>Current Status</th>
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<tr>
<td>1</td>
<td>–</td>
<td>–</td>
<td>Pleural effusion</td>
<td>98</td>
<td>DUC</td>
</tr>
<tr>
<td>2</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>88</td>
<td>DF</td>
</tr>
<tr>
<td>3</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>58</td>
<td>DF</td>
</tr>
<tr>
<td>4</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>53</td>
<td>AWD</td>
</tr>
<tr>
<td>5</td>
<td>–</td>
<td>–</td>
<td>Anemia</td>
<td>47</td>
<td>DF</td>
</tr>
<tr>
<td>6</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>40</td>
<td>DF</td>
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<tr>
<td>7</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>37</td>
<td>DF</td>
</tr>
</tbody>
</table>

DUC, died of unrelated causes; DF=disease free; AWD Alive with disease

Table IV - Histopathological and immunohistochemical characteristics

<table>
<thead>
<tr>
<th>Case no</th>
<th>Diameter(cm)</th>
<th>Mitoses/50 HPFs</th>
<th>Immunohistochemistry</th>
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<tr>
<td>1</td>
<td>5</td>
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<td>S100(+),Ki67&lt;%1,SMA(-),DESM N(-),CD34(-),CD117(-)</td>
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<tr>
<td>2</td>
<td>10</td>
<td>0</td>
<td>S100(+),Ki67 %2</td>
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<td>3</td>
<td>7</td>
<td>0</td>
<td>S100(+),Ki67 %2, SMA(-),DESM N(-),CD34(-),CD117(-),Dog1(-),EMA(-),NFP(-)</td>
</tr>
<tr>
<td>4</td>
<td>13.3</td>
<td>2</td>
<td>S100(+),Ki67%2,SMA(+),Vimentin(+),NSE(+),GFAP(+),CD34(+),CD117(+),Dog1(-)</td>
</tr>
<tr>
<td>5</td>
<td>3</td>
<td>1</td>
<td>S100(+),Ki67%2,SMA(+),DESM N(-),CD34(+),CD117(+),Dog1(-)</td>
</tr>
<tr>
<td>6</td>
<td>3.3</td>
<td>0</td>
<td>S100(+),Ki67 %3</td>
</tr>
<tr>
<td>7</td>
<td>0.3</td>
<td>0</td>
<td>S100(+),Ki 67 %1, DESM N(-),CD117(-)</td>
</tr>
</tbody>
</table>

Preoperative diagnosis of GIS schwannomas is often difficult because there is no specific clinical symptom of the tumor, and there are no diagnostic methods show any pathognomonic features 18. In computed tomography (CT) examination, these tumors are often detected as homogenous exophytic masses; cystic changes, cavity formation, necrosis or calcification are rare. In endoscopy, gastric schwannomas are seen as large as high submu-
resection with negative margins is the standard treatment for benign schwannomas. Radical surgery and extended resection are not recommended. The surgical method may vary depending on the location and size of the tumor and minimally invasive procedures are recommended for surgical treatment. When the tumor is small, endoscopic resection can be performed and transanal tumor resection can be considered for rectal tumors. There is a reported risk of recurrence after complete resection of benign schwannomas. Radical surgery is recommended for malignant schwannomas. Particularly for asymptomatic patients, some controversy remains as to whether tumors should be removed. Resection is necessary in cases where definitive diagnosis cannot be made preoperatively, even if asymptomatic. Schwannomas are usually less than 2 cm in diameter and most of these patients have no significant symptoms. Gastrointestinal Stromal Tumors (GIST) are borderline tumors with a relatively high risk of malignant transformation. According to the National Comprehensive Cancer Network (NCCN) guidelines, surgical resection of GISTs with a diameter of less than 2 cm is not recommended if there are no high-risk EUS features (irregular borders, cystic spaces, ulcers, ulceration, echogenic foci). Periodic endoscopic or radiographic surveillance should be considered in determining treatment modality. Since the risk of malignant transformation of intestinal schwannoma is lower than that of GISTs, it is thought that these tumors can be followed. Resection should be prioritized when differential diagnosis of Schwannomas is difficult with GISTs. GANTs and malignant peripheral schwannomas. Surgical treatment is applied in our clinical practice for the treatment of schwannoma cases. In our series, wedge resection and proximal gastrectomy were performed in one of the gastric origin schwannoma patients. Debulting was performed in the retroperitoneal localized schwannoma case. Partial pancreatectomy +-py pancreateojunostomy was performed in the case of schwannoma originating from the pancreas in the left juxta-drenal space. Resection was performed in one of the pelvic Schwannoma cases and cytoreductive surgery + Hyperthermic Intraperitoneal Chemotherapy (HIPEC) was performed in the other one because of peritoneal carcinomatosis. The patient who underwent pelvic mass excision as an intraoperative complication developed sudden hypotension. Hypotension was considered as an anesthesia-related complication in the patient who did not have hemorrhage and the operation was continued. Postoperative pancreatic fistula developed in the patient who underwent partial pancreatectomy. The fistula completely resolved with Somatostatin 3.5 mcg/kg/hour IV infusion therapy and parenteral nutrition. Cytoreductive surgery + HIPEC patient developed wound infection and was controlled with regular dressing without surgical intervention. The mean length of hospital stay was 11 days and the patient with the longest hospitalization was followed up for pancreatic fistula. None of the patients required re-operation. There was no postoperative 90-day mortality. After discharge, the patient who developed pancreatic fistula was re-hospitalized because of pleural effusion and thoracentesis was performed. The effusion fluid was transudate and when investigated for the presence of intrabdominal collection, no collection was detected in the operation site. The patient who had undergone cytoreductive surgery was admitted for anemia and underwent blood transfusion. Gastrointestinal schwannomas are usually submucosal and hypervascular. The lesions typically protrude into the lumen. Endoscopic examination shows small mucosal erosions. Schwannomas are submucosal and may not be diagnosed by superficial biopsies. It is usually clinically silent and may be discovered incidentally during upper gastrointestinal endoscopy, eroding the mucosa, causing gastrointestinal bleeding. It may present with luminal obstruction and bleeding as a result of mass growth.

Wedge resection, subtotal gastrectomy or total gastrectomy are the preferred treatment methods in gastric Schwannoma cases. The most important point in surgical treatment is complete resection of the tumor with a negative surgical margin. Gastric schwannomas rarely metastasize to lymph nodes like other soft tissue sarcomas and therefore surgical lymphadenectomy is not routinely recommended in gastric schwannomas. In our series, gastric schwannoma patients presented with nonspecific symptoms such as abdominal pain and dyspepsia. Patients with a non-diagnostic biopsy result underwent surgery with a preliminary diagnosis of stromal tumor and wedge resection was performed in one patient and proximal gastrectomy was performed in the other patient. Both patients are followed with disease-free survival.

Pancreatic schwannomas are most commonly localized to the head of the pancreas (40%), followed by the trunk (21%), neck (6%), tail (15%) and uncinate process (13%), respectively. Pancreatic schwannoma patients are usually asymptomatic or have nonspecific symptoms such as abdominal pain, nausea and vomiting. Complete, but conservative resection is the treatment of choice for pancreatic Schwannomas. Peripancreatic schwannomas originating from pancreatic nerve plexuses can be confused with pancreatic tumor and preoperative diagnosis in these patients may prevent unnecessary pancreatectomy. Retroperitoneal schwannomas may be malignant and potentially fatal. Therefore, it is important to completely remove the retroperitoneal tumors. Local recurrence has been reported in 5-10% of retroperitoneal schwannomas. In our series, one patient had schwannoma located in the tail of the pancreas and partial pancreatectomy was performed. The other patient had retroperitoneal region localization and in this patient the pancreas was preserved and mass excision with negative surgical margin was performed.
Colorectal schwannomas are less common than other GI schwannomas. In the literature, it was reported that colorectal schwannoma was more common in the female gender (59%) and the mean age at presentation was 61.5 years. More than 98% of colorectal schwannomas are benign and have a low mitosis rate and a low Ki-67 proliferation index. The best treatment option is complete surgical resection with negative margins. Radical surgery is not usually necessary.

In our study, one patient had schwanna located in the rectum and transanal excision was performed. Mitotic activity was not observed in the tumor and Ki-67 proliferation index was 1%. At 37 months postoperatively, the patient continues to be monitored without any disease.

The rate of diagnosis of intestinal schwannoma in the preoperative period is relatively low. Inagawa et al. showed that preoperative diagnosis rate was only 15.2%. Diagnosis is mainly based on postoperative pathology specimen examination and immunohistochemistry studies.

Immunohistochemical examinations are necessary to differentiate between different spindle cell tumor types. Desmin and SMA positivity showed smooth muscle lesions such as leiomyoma or leiomyosarcoma; CD34 and CD117 positivity indicate GIST. S100 strongly positive staining supports the diagnosis of schwannoma.

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tumori rari che presentano più comunemente un coinvolgimento gastrointestinal. Poiché questi tumori sono per lo più benigni, la prognosi a lungo termine dei pazienti è buona. Lo Schwannoma dovrebbe essere tenuto presente nella diagnosi differenziale delle masse intraaddominali. Le resezioni radicali con elevata morbidità e mortalità dovrebbero essere evitate se viene fatta una diagnosi preoperatoria.

References