CASE REPORT

TJG
The Turkish Journal of Gastroenterology

Plexiform schwannoma of the duodenum accompanying pyloric stenosis: Report of a case

Ali AKTEKİN[1], Selvinaz ÖZKARA[2], Kaan MERİÇ[3], Meryem GÜNAY GÜRLEYİK[1], Fügen AKER[2], Abdullah SAĞLAM[1]

Departments of [1]General Surgery, [2]Pathology and [3]Radiology, Haydarpaşa Numune Education and Research Hospital, İstanbul

Plexiform schwannoma is a benign peripheral nerve sheath tumor and is composed of Schwann cells arranged in a plexiform pattern. Most plexiform schwannomas are skin tumors, and there has been no case report of this tumor originating in the duodenum. We describe the first known case of plexiform schwannoma of the duodenum. A 60-year-old man presented with a short history of food intolerance, epigastric discomfort, fullness and bloatedness, sometimes vomiting, and weight loss, without any clinical picture of neurofibromatosis. Upper gastrointestinal endoscopy revealed pyloric stenosis with normal mucosal lining. The computed tomography demonstrated circumferentially and concentrically thickened pylorus up to 18 mm with narrowed lumen and limited contrast passage. Antrectomy and gastrojejunostomy were performed due to unknown etiology of the obstruction. The cut surface of the lesion revealed thickened pylorus up to 15 mm in a circumferential manner. It contained a 5 mm tumor consisted of multiple white nodules in the submucosal and subserosal layers with overlying duodenal mucosa. Microscopic examination revealed nodular structures composed of spindle cells within fascicular pattern without any atypia or mitosis. Immunohistochemical examination revealed that the cells diffusely and strongly expressed S100 proteins in a nuclear and cytoplasmic pattern, but not CD117, smooth muscle actin, desmin, or CD34, confirming plexiform schwannoma.

Key words: Duodenum, gastrointestinal stromal tumors, nerve sheath tumor, plexiform schwannoma

Pilor stenozuna eşlik eden duodenumda pleksiform schwannoma: Olgu raporu

Pleksiform schwannoma, periferik sinir kılıfının bir tümörü olup plexiform yapıda dizilmiş schwan hücrelerinden oluşur. Çoğunlukla cilt tutulumu ile kendini gösterir. Duodenumun pleksiform schwannoması şimdiye kadar henüz rapor edilmemiştir. Biz bu yazımızda ilk olarak duodenumun pleksiform schwannomasını rapor ettik. Altmış yaşında erkek hasta oral alımda güçlük, epigastrik ağrı, karında dolgunluk ve gaz, kusma ve kilo kaybı ile hastanemize başvurdu. Hastada klinik olarak neurofibromatosis bulgusu bulunmamaktadır. Endoskopik incelemede normal mukoza ile kaplı pilor stenozu olduğu görüldü. Bilgisayarlı tomografi incelemesinde pilorun 18 mm kadar ulaşan konsantrik olarak kalınlaştığı, kontrast geçişinin azaldığı rapor edildi. Hastaya antrektomi ve gastrojejunostomi ameliyatı yapıldı. Lezyon patolojik olarak incelendiğinde pilorun çepecevre olarak 15 mm kadar kalınlaştığı, pilorun duodenum yüzünde yüzeyi sağlam duodenum mukozası ile kaplı 5 mm çaplı beyaz renkte sert nodüler yapı olduğu görüldü. Mikroskopik incelemede bu nodüler yapının fasiküler yapıda dizilmiş atipi ve mitoz göstermeyen iğsi hücrelerden oluştuğu görüldü. İmmünohistokimyasal incelemede ise bu hücrelerin yaygın ve kuvvetli olarak S100 ile boyanırken, CD117, düz kas aktin, desmin ve CD34 ile boyanmadığı görülerek pleksiform schwannoma olduğu teşhis edildi.

Anahtar kelimeler: Duodenum, gastrointestinal stromal tümörler, sinir kılıfı tümörleri, pleksiform schwannoma

INTRODUCTION

Plexiform schwannoma (PS) is a rare benign peripheral nerve sheath tumor composed of Schwann cells arranged in a plexiform pattern. It was first described by Harkin et al. (1) in 1978,

Address for correspondence: Ali AKTEKİN
Department of General Surgery, Haydarpaşa Numune Education and Research Hospital, İstanbul, Turkey
E-mail: draliaktekin@yahoo.com

Manuscript received: 08.06.2011 **Accepted:** 08.08.2011

Turk J Gastroenterol 2012; 23 (4): 385-389
doi: 10.4318/tjg.2012.0403

and the first case of a solitary PS in a visceral organ was described in 1997 by Hirose et al. (2). PS predominantly affects young adults and occurs most commonly as a solitary, slow-growing, asymptomatic nodule on the head, neck, trunk, or upper extremities. It usually arises in either dermis or subcutaneous tissue, but has also been described in skeletal muscle or other deep somatic soft tissue (3). Occurrences of intra-abdominal and retroperitoneal schwannomas are rare. Schwannomas have been reported to represent only 2.9%-6% of all gastrointestinal (GI) mesenchymal tumors. Intra-abdominal schwannomas occur most frequently in the alimentary tract, and the most common site is the stomach. Other intra-abdominal sites are even rarer and have been reported in the esophagus, small intestine, colon, greater omentum, lesser sac, and the biliary tree (4). Sporadic cases of PS usually present as solitary tumors. A few reports have presented multiple PS in patients with neurofibromatosis (NF) (3). We describe herein the first known case of PS of the duodenum.

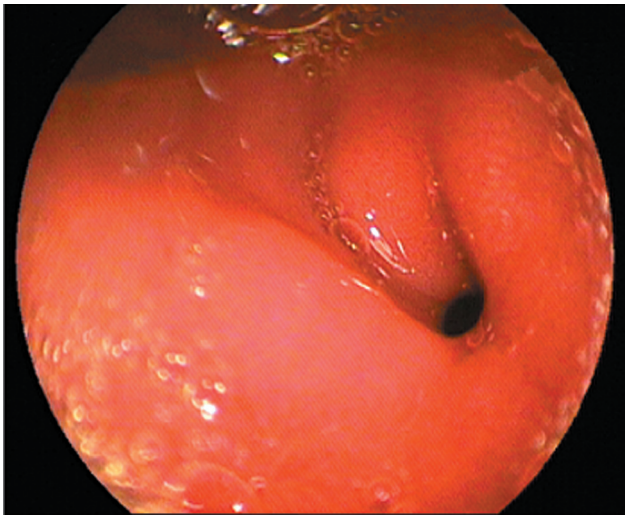

Figure 1. Upper gastrointestinal endoscopy revealed pyloric stenosis with normal mucosal lining.

CASE REPORT

A 60-year-old male presented with a history of food intolerance, epigastric discomfort, fullness and bloatedness, sometimes vomiting, and weight loss, but with normal blood chemistry. Upper GI endoscopy revealed pyloric stenosis without any ulceration. The endoscopic view of the mucosal lining of that part was normal (Figure 1). Endoscopic biopsy revealed normal duodenal mucosal epithelium. The computed tomography (CT) demonstrated a circumferentially and concentrically thickened pylorus and proximal duodenal wall up to 18 mm with narrowed lumen and limited contrast passage (Figure 2). The patient was operated due to obstructive duodenal pathology, and antrectomy with duodenal resection was performed. The patient had an uneventful postoperative course. The clinical examination did not fit with any signs or symptoms of NF.

The cut surface of the lesion revealed a thickened pylorus up to 15 mm in a circumferential manner. Histological examination by hematoxylin–eosin staining of the specimen revealed a 5 mm tumor consisting of multiple white nodules in the submucosal and subserosal layers with overlying normal-appearing duodenal mucosa. It was un-encapsula-

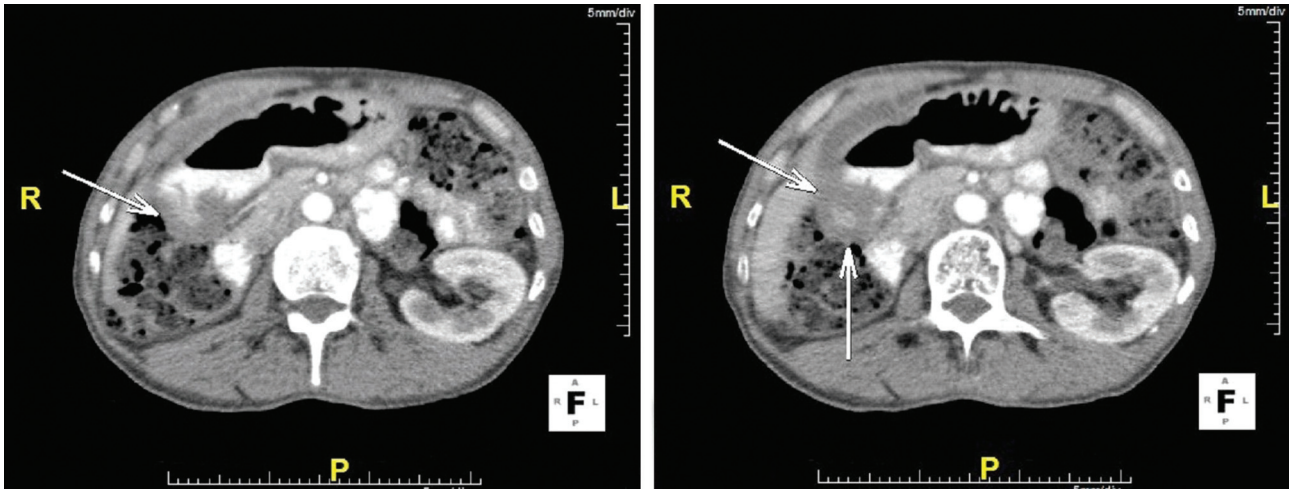

Figure 2. The axial section of the CT demonstrated a circumferentially and concentrically thickened pylorus and proximal duodenal wall up to 18 mm (1 layer) with narrowed lumen and limited contrast passage.

ted but well circumscribed, firm, and homogeneous with rubbery nodular development in the submucosal and muscularis propria of the duodenum in plexiform pattern. The tumor consisted of microscopic multiple nodules of varying size demonstrating the obviously plexiform tumor. Neither degenerative changes as necrosis or hemorrhage nor cystic changes were observed (Figure 3a). Nodular structures were composed of spindle cells within fascicular pattern without atypia or mitosis (Figure 3b). The tumor was surrounded by a cuff of lymphoid aggregates (Figure 3a). The Verocay bodies and Antoni A and B areas were absent. Immunohistochemical examination revealed that the cells diffusely and strongly expressed S100 protein in a nuclear and cytoplasmic pattern, but not CD117, smooth muscle actin, desmin, or CD34 (Figure 3c, 3d). These results confirmed the diagnosis of a multi-nodular PS and eliminated leiomyoma, neurofibroma, and gastrointestinal stromal tumor (GIST). Surgical margins were clear. The surgical specimen included 11 lymph nodes without any metastasis.

DISCUSSION

Gastrointestinal tract (GIT) schwannomas have been reported to occur in patients over a wide range of age groups, with a median age of 50-60 years. Most series reported a female preponderance. These tumors range in size from 0.5 to 11 cm (4,5). The tumor was only 5 mm in size in our case. Although most of the reported cases involved a solitary lesion, there are a few cases of multiple lesions (1,3,6). Because of its rarity, it is not diagnosed preoperatively. The most common presenting symptom is an episode of intermittent GIT blee-

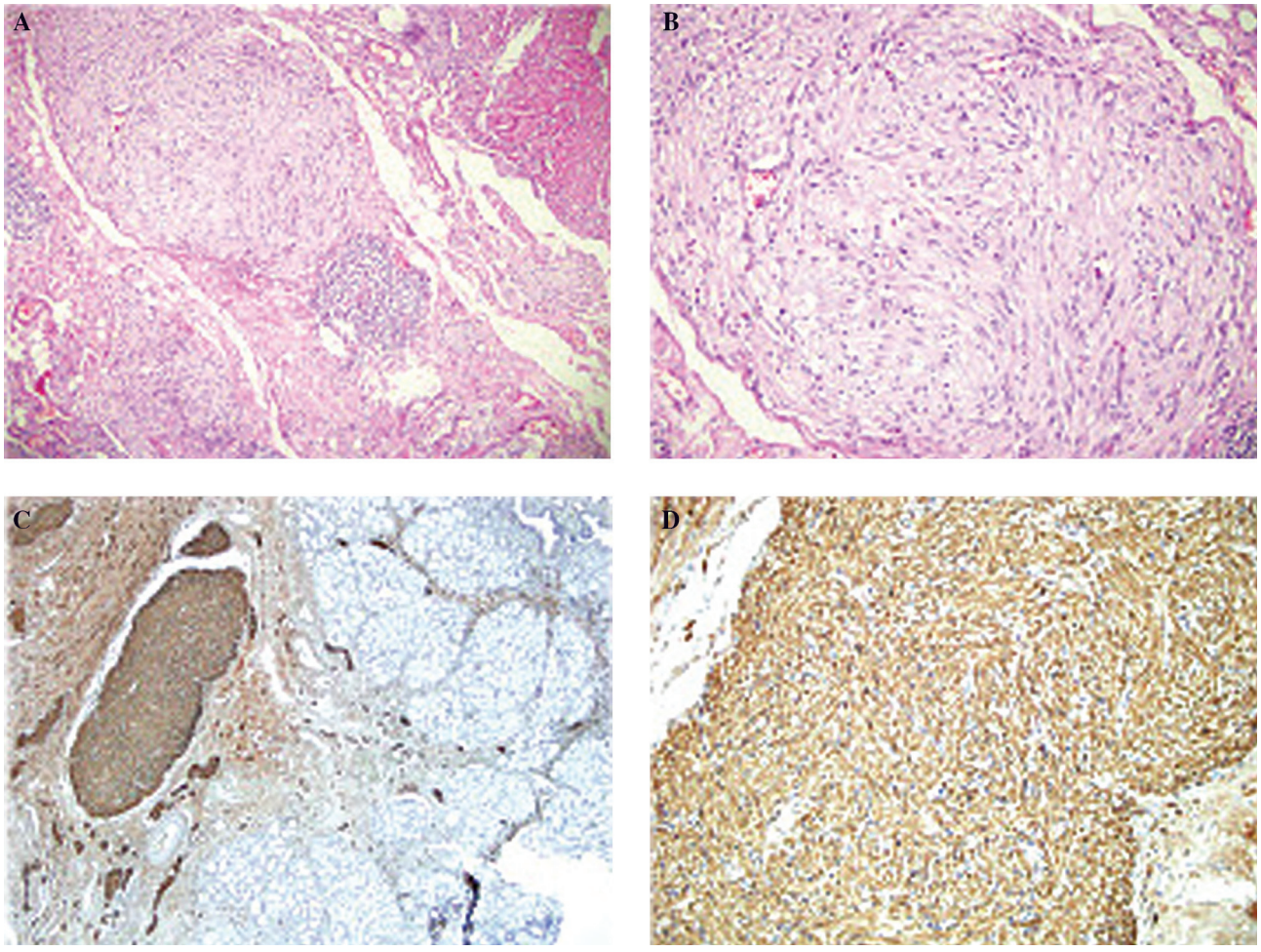

Figure 3. A – Plexiform pattern of growth of tumor between submucosa and fibers of muscularis propria with lymphoid aggregates accompanying the tumor (H&E, x200); **B –** Tumor composed of spindle cells without any atypia or mitosis (H&E, x400); **C & D -** Immunohistochemical examination revealed that the cells diffusely and strongly expressed S100 protein in a nuclear and cytoplasmic pattern (4x100 & 5x400).

ding, abdominal pain, epigastric discomfort and bloatedness, epigastric mass, and rarely, obstruction (4, 7). In this patient, the pylorus was covered with normal mucosal epithelium without ulceration but obstruction may have been the result of the secondary effect of the 5 mm PS. At present, there are limited data reporting the CT features of GIT schwannomas. Neither preoperative cross-sectional imaging nor endoscopy has shown any pathognomonic features unique to this tumor and both are unhelpful. On CT scan, these tumors have a homogeneous pattern of attenuation on both unenhanced and contrast-enhanced scans with tumor enhancement occurring in the equilibrium phase. The main differential diagnoses of GIT schwannomas are GISTs. These tumors most frequently have a heterogeneous appearance on CT because of hemorrhage, necrosis or cystic changes. Other neoplasms such as lymphomas and GI adenocarcinomas may also have overlapping features with GIT schwannomas. Similar to cross-sectional imaging, the endoscopic features of GI schwannomas are non-pathognomonic because these tumors are submucosal lesions. Endoscopy and CT demonstrated the narrowed lumen, and these are not pathognomonic features. Endosonography can be helpful in defining the borders of the tumor and localization within the GI wall, but no typical endosonographic features of GI schwannoma have been defined. Because of their plexiform pattern of growth, it is relatively common for benign schwannomas to traverse several layers of bowel wall and into the surrounding adipose tissue. Endoscopic biopsies are usually not representative of the deeper submucosal tissue. If samples from the deeper tissues are obtained, they usually demonstrate nonspecific spindle cells and will be insufficient for a definite diagnosis (4). In our case, the duodenum was lined by normal epithelium, and so endoscopic biopsy could not achieve the diagnosis and revealed the normal duodenal epithelium.

Pathologically, GIT schwannomas are regarded as distinct tumors from conventional schwannomas (4). They arise from the nerve plexus of the gut wall (8). Degenerative changes such as necrosis or hemorrhage and cystic changes, which are frequently found in soft tissue schwannomas and also in the retroperitoneum, are seldom present. Microscopically, unlike conventional schwannomas, GIT schwannomas are not encapsulated but most are well- circumscribed. They are frequently surrounded by a cuff of lymphoid aggregates, highly cellular, and composed mainly of bipolar spindle cells. Verocay bodies, vascular hyalinization, Antoni A and B areas, and a typical palisading structure are typically absent, unlike in conventional schwannomas (4,8). On the other hand, the lesser sac schwannoma demonstrates the typical pathologic features of peripheral and soft tissue. The pathologic appearance of extra-GI intra-abdominal schwannomas is variable, with some cases in the omentum or lesser sac having the typical appearance of conventional schwannomas, but those in the liver and biliary tree reported as having features similar to GIT schwannomas (4,9). Immunocytochemical studies are required to distinguish schwannoma from neurofibromas, GISTs, and leiomyomas. Cells from schwannomas are usually 100% immunoreactive with S-100 protein in contrast to 30%-40% immunoreactivity in neurofibromas. GISTs are positive for c-kit and negative for S-100 protein, whereas leiomyomas are positive for smooth muscle actin and desmin, and negative for S-100 (4). In our case, duodenal lesion diffusely and strongly expressed S100 protein in a nuclear and cytoplasmic pattern, but not CD117, smooth muscle actin, desmin, or CD34, similar to the reported cases in the literature.

Although GIT schwannomas are benign tumors, isolated case reports of “malignant schwannomas”, also termed malignant peripheral nerve sheath tumors, have been reported (10). PS is distinguished from malignant peripheral nerve sheath tumors in the pathologic examination by the plexiform pattern of growth, by the shape of the Schwann cells, the nuclei, a low mitotic index, and noninvasive growth (3). Most pathologists regard these malignant tumors with neural differentiation as distinct tumors from GIT schwannomas, giving them the term GI autonomic nerve tumors (11). In our patient, the properties of the PS were plexiform pattern of growth, low mitotic index and absence of lymph nodal metastasis, which confirmed it as a benign lesion.

Schwannomas may occur in the setting of NF types 1 (von Recklinghausen's disease) and 2. Most reported cases of PSs were solitary lesions, while a few studies reported that tumors can multiply in NF2 patients. It is critical to differentiate a PS from a plexiform neurofibroma because the latter is pathognomonic of NF1 and carries significant risk of malignant transformation (3,12). Our patient did not have any signs or symptoms of NF type 1 or 2 so our patient is sporadic.

Nonetheless, although benign, the treatment of choice of GIT schwannomas is complete surgical excision in fit, healthy patients, both to distinguish these tumors from other GIT mesenchymal tumors and for symptom relief. Because of the obstructive symptoms and the unrecognized etiology, we performed surgery with a clear margin with an uneventful postoperative course. The outcome after surgical resection is excellent, and no report in the literature has shown malignant potential (4,5,8,11).

In conclusion, although PS of the GIT is very rare, it could also affect the duodenum. Preoperative endoscopy, CT and endoscopic biopsy are unrevealing. Preoperative endosonography may be helpful, but it is not used on a widespread basis. Pyloric stenosis without ulcer is also an interesting point. The pylorus was covered with normal mucosal epithelium, but the obstruction may have been the result of the secondary effect of the 5-mm PS. Its low mitotic activity and absence of atypia and lymph node metastasis showed its benign nature, but long-term follow-up is still necessary.

REFERENCES

1. Harkin JC, Arrington JH, Reed RJ. Benign plexiform schwannoma, a lesion distinct from plexiform neurofibroma (abstract). J Neuropathol Exp Neurol 1978; 37: 622.
2. Hirose T, Scheithauer BW, Sano T. Giant plexiform schwannoma: a report of two cases with soft tissue and visceral involvement. Mod Pathol 1997; 10: 1075-81.
3. Ko JY, Kim JE, Kim YH, Ro YS. Cutaneous plexiform schwannomas in a patient with neurofibromatosis type 2. Ann Dermatol 2009; 21: 402-5.
4. Goh BKP, Chow PKH, Kesavan S, et al. Intraabdominal schwannomas: a single institution experience. J Gastrointest Surg 2008; 12: 756-60.
5. Kwon MS, Lee SS, Ahn GH. Schwannomas of the gastrointestinal tract: clinicopathological features of 12 cases including a case of esophageal tumor compared with those of gastrointestinal stromal tumors and leiomyomas of the gastrointestinal tract. Pathol Res Pract 2002; 198: 605-13.
6. Iida A, Imamura Y, Katayama K, et al. Plexiform schwannoma of the small intestine: report of a case. Surg Today 2003; 33: 940–3.
7. Tahir TM, Anwar S, Naseem N, et al. Gastric schwannoma in a female patient with pulmonary tuberculosis - a clinicopathological assessment and diagnosis. Malaysian J Med Sci 2010; 17: 45-50.
8. Hou YY, Tan YS, Wang XN, et al. Schwannoma of the gastrointestinal tract: a clinicopathological, immunohistochemical and ultrastructural study of 33 cases. Histopathology 2006; 48: 536-45.
9. Khan AA, Schizas AM, Cresswell AB, et al. Digestive tract schwannoma. Dig Surg 2006; 23: 265–9.
10. Bees NR, Ng CS, Dicks-Mireaux C, Kiely EM. Gastric malignant schwannoma in a child. Br J Radiol 1997; 70: 952–5.
11. Prevot S, Bienvenu L, Vaillant JC, de Saint-Maur PP. Benign schwannoma of the digestive tract. A clinicopathologic and immunohistochemical study of five cases, including a case of esophageal tumor. Am J Surg Pathol 1999; 23: 431-6.
12. Woodruff JM, Marshall ML, Godwin TA, et al. Plexiform (multinodular) schwannoma. A tumor simulating the plexiform neurofibroma. Am J Surg Pathol 1983; 7: 691-7.