***Case Report* / Olgu Sunumu**

A Rare Case of Schwannoma Arising From a Diverticulum in the First Portion of Duodenum

Duodenum Birinci Kısım Divertikülünden Kaynaklanan Nadir Bir Schwannom Olgusu

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A case of duodenal schwannoma, a rare tumor arising from schwannian cells of peripheral nerves is presented. A 57-year-old woman was admitted with abdominal pain and fever during investigation of anemia. Physical examination revealed tenderness in the right side of the upper abdomen. Laboratory tests showed no abnormality except for leucocytosis of 15300/l. All tumor markers were negative. Ultrasonography revealed a hypoechoic mass adjacent to the duodenum. Computed tomography and magnetic resonance imaging showed a 4x3 cm mass in the same region and an abscess with dimensions of 8x11x12 cm in the right lobe of liver. Hepatic abscess drainage and duodenal diverticulectomy was performed. Histopathological examination revealed a tumorous mass composed of interlacing bundles of spindle cells, showing mild variations in nuclear shape and size. Immunohistochemically tumor cells were positive for S100 protein, thus, schwannoma of the duodenum was diagnosed. The postoperative course was uneventful and the patient was discharged on the eighth day of operation.

Key words: Schwannoma; duodenum; diverticulum.

Bu yazıda periferal sinir schwann hücrelerinden köken alan nadir bir duodenal schwannoma olgusu sunulmuştur. 57 yaşında bayan hasta anemi tetkiki sırasında ateş ve karın ağrısı şikayeti ile başvurdu. Fizik muayenesinde karın sağ üst kadranda hassasiyet tespit edildi. Laboratuar değerlerinde 15300/l olan lökositoz haricinde anormallik yoktu. Tüm tümör belirteçleri normal değerlerdeydi. Ultrasonografide duodenuma yapışık hipoekoik kitle tespit edildi. Bilgisayarlı tomografi ve manyetik rezonans incelemede aynı lokalizasyonda 4x3 cm'lik kitle ve 8x11x12 cm'lik karaciğer sağ lobda abse bulundu. Hepatik abse drenajı ve duodenal divertikülektomi yapıldı. Histopatolojik incelemede şekil ve büyüklükte hafif derecede değişiklikler gösteren kesişen iğsi hücre kümeleri içeren tümöral kitle görüldü. İmmünohistokimyasal olarak tümör hücrelerinin S100 protein ile kuvvetli pozitiflik vermesi nedeni ile duodenal schwannoma tanısı kondu. Ameliyat sonrası dönemi sorunsuz geçen hasta 8. gün taburcu oldu.

Anahtar sözcükler: Schwannoma; duodenum; divertikül.

Tumors of the small bowel are uncommon, representing 3-6% of all gastrointestinal neoplasms. Less than 30% of these tumors are located in duodenal region. Gastrointestinal schwannomas are extremely rare neoplasms accounting for less than 1% of all neoplasms of the gastrointestinal tract.[1] Most of the schwannomas are benign tumors. Hemorrhage and bowel obstruction are the most common presenting features and as the tumor is peripherally located, enlarging mass associated with abdominal pain is the main complaint.[1] They have almost the same inci-

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dence in men and women and a similar median age of presentation, fifth to sixth decades.[2] Immunohistochemistry plays a central role at the diagnosis of schwannoma as in many other types of cancer. Strong positive reaction for S-100 protein turns out to be supportive for the diagnosis of schwannoma.[3] Radical excision with margins free of disease is the treatment of choice, since their response to both chemotherapy and radiotherapy remains uncertain.[4,5] We recently treated a woman who presented with abdominal pain, fever, and anemia which was subsequently diagnosed as a schwannoma located in a diverticulum of the first portion of duodenum. We describe the clinical and pathological features of this unusual tumor.

CASE REPORT

A 57-year-old woman was admitted to our hospital with abdominal pain and fever during investigation of anemia. Physical examination revealed tenderness at the right side of the upper abdomen. Laboratory tests showed no abnormality except for leucocytosis of 15 300/l. All tumor markers were negative. A review of her family history was unremarkable. Ultrasonography (data was not shown) revealed a hypoechoic mass adjacent to the duodenum and a computed tomography showed a 4x3 cm mass in the same region and an abscess in 8x11x12 cm in the right lobe of liver (Figs. 1a, b). Also magnetic resonance imaging (MRI) revealed a hypointense mass in the region of gastrohepatic ligament and a hepatic abscess (Fig. 2). The patient was referred to the surgical department with a preliminary diagnosis of hepatic abscess and duodenal mass. Based on the clinical and radiological findings, an operation decision was given. A midline laparotomy revealed a hepatic abscess with dimensions of 8x11x12 cm in the right lobe and a diverticulum in the first portion of the duodenum. A 3x3 cm mass was palpated in the diverticulum. The duodenal diverticulum was resected carefully and drainage was performed for hepatic abscess. Macroscopically, the resected mass was an elastic mass of 3.1x3x2.2 cm. The cut surface of the tumor mass was yellowish-white. Histologically, hematoxylin-eosin stained slides revealed a tumor which was composed of spindle-shaped cells arranged in a palisading pattern forming cellular and hypocellular areas (Figs. 3a, b). The nuclei of the tumor cells showed mild variations in size and shape. Mitotic figures were scarce. The findings were consistent with a schwannoma. Immunohistochemically positive staining for S-100 protein, and negative reaction for desmin, CD34, and c-kit (CD117) confirmed the histopathological diagnosis. The patient's postoperative course was uneventful and she was discharged on the eighth day after operation and she is currently free from disease in the four-year period.

DISCUSSION

Gastrointestinal stromal tumors constitute the largest group of mesenchymal tumors in the

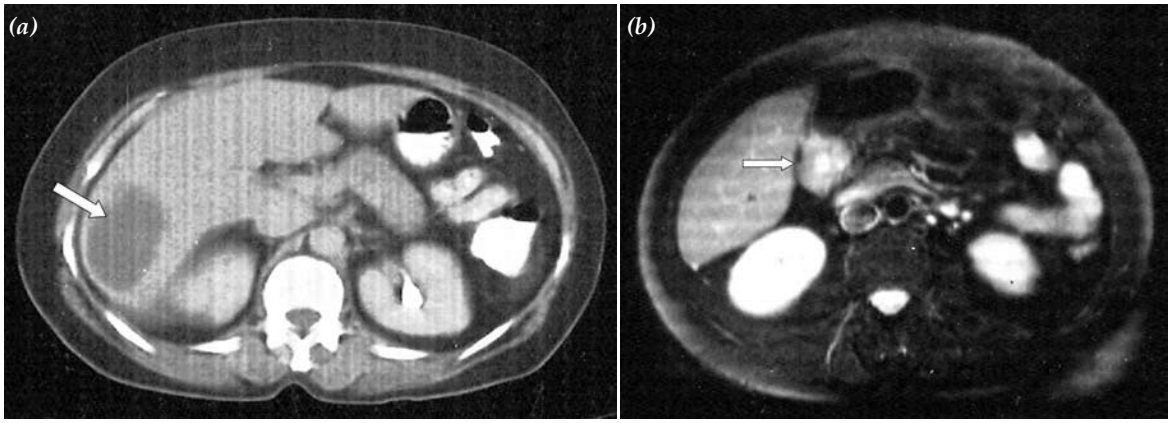

Fig. 1. ***(a)*** *Computed tomography of the abdomen demonstrated a round, 8x11x12 cm diameter collection of fluid and air in the undersurface of the right hepatic lobe (arrow).* ***(b)*** *Computed tomography demonstrates a 3x3 cm diameter lesion (arrow) adjacent to the duodenum.*

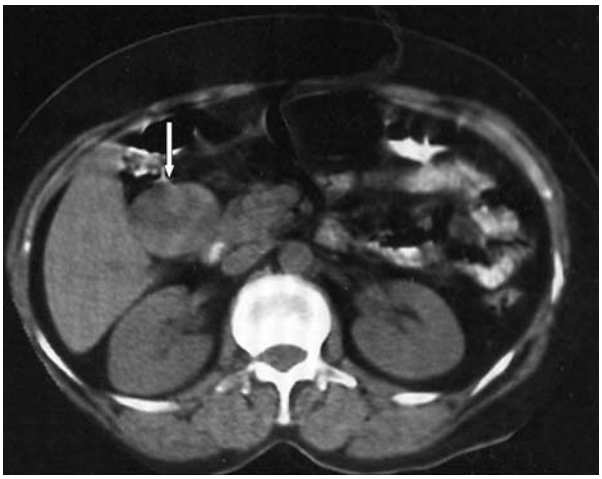

Fig. 2. *Magnetic resonance imaging revealed a 4 cm of spheric lesion near by the duodenum (small arrow).*

digestive tract. Schwannomas are the smallest group of these tumors.[6] Schwannomas are rare tumors arising from Schwann cells which cover the peripheral nerves, and are difficult to distinguish from leiomyogenic tumors. Most of the gastrointestinal tract schwannomas are involved within the stomach and schwannoma in the duodenal region is extremely rare.[7] They are mostly located in the second or third portion of duodenum.[8] Nilsson and Jonsson[9] reviewed 43 cases of schwannoma of the small intestine, and eight of the tumors were located in the duodenum. Miettinen et al.[5,7] reported two cases of schwannoma in the duodenum among approximately 3000 gastrointestinal mesenchymal tumors from the Armed Forces Institute of Pathology and 250 from the Haartman Institute. There are small series of schwannomas which occur in fifth to sixth decades, equally in men and women.[1,3,5,7] Our patient was in the fifth decade and opposite to the literature the tumor was located in a diverticulum in the first portion of the duodenum. Botos et al.[10] reported only one case of a gastrointestinal stromal tumor arising from a duodenal diverticulum and our patient is the first schwannoma case originated from a diverticulum in the first portion of the duodenum. Schwannomas can be asymptomatic for years and the symptoms may be nonspecific. However, they may present with the symptoms of abdominal pain or discomfort.[11] Intestinal bleeding, intussusception, ulceration and obstruction are the clinical manifestations of schwannomas of gastrointestinal tract.[3,8,12] Gallo and Sagatelian[11] and Seno et al.[12] reported schwannoma of duodenum presented with gastrointestinal bleeding. Recurrent gastrointestinal bleeding with silent periods are the most common symptoms associated with tumor size, and erosion of the overlying mucosa. The diagnosis of patients is based on their symptoms. Endoscopy may be inefficient without mucosal involvement or when the tumor size is small. Also this investigation may be normal or may only show nonspecific secondary findings such as extrinsic mass effect or ulceration, if the schwannoma is mainly exophytic. Furthermore, endoscopic biopsy may not be adequate for definite diagnosis because schwannomas are submucosal tumors and mucosal abnormality may be minimal.[13] Computed tomography is the

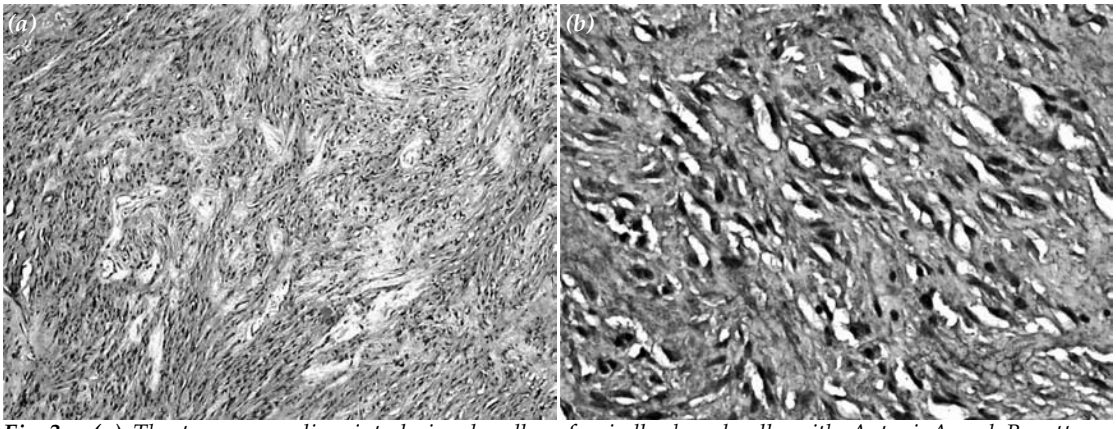

Fig. 3. ***(a)*** *The tumor revealing interlacing bundles of spindle-shaped cells with Antoni A and B patterns (H-E x 50).* ***(b)*** *Diffuse strong positive cytoplasmic reaction for S-100 protein (S-100 x 200).*

common method to evaluate these patients. It is also useful in suggesting the benign or malignant nature of the neoplasm and preoperative staging. Computed tomography demonstrates round or oval homogeneously attenuating, well-defined mass with frequent signs of degeneration, such as cysts and calcifications.[2,13,14] The MRI appearance of schwannomas arising from cranial or spinal nerves has been well described in the literature but very little is known about MRI findings of gastrointestinal tract. Magnetic resonance imaging reveals these lesions as sharply demarcated, strongly enhancing tumors, having low to medium signal intensity on T1-weighted images and high signal intensity on T2-weighted images.[15] There is also limited information about positron electron tomography (PET) findings of schwannomas. Positron electron tomography with fluorodeoxyglucose (FDG) uptake is widely used to quantify the metabolism of malignant bone and soft-tissue tumors, whereas little is known about FDG uptake in benign lesions. Positron electron tomography with fluorodeoxyglucose is of limited value for the preoperative differentiation of schwannoma from sarcoma.[16,17] Macroscopically schwannomas are solitary, well-circumscribed, encapsulated, homogeneous, firm and yellow-white tumors. On microscopy, schwannomas are composed of Schwannian cells with regions of high and low cellularity termed Antoni A and Antoni B areas, respectively, with intense staining for S-100 protein. In the present case, immunohistochemical staining for S-100 protein showed strong positivity and there was no reaction for desmin, CD34, and c-kit.[18] Surgery remains the only potentially curative therapy for these tumors. The surgical approach depends on the tumor size, tumor localization and histological features. Besides the commonly used surgical techniques consisting of resection and enuclation, minimally invasive modalities such as laparoscopy is the operational treatment alternatives of these tumors. Additional extended procedures may be added due to peroperative pathology consultations.[19] Schwannomas are almost benign tumors but may rarely undergo malignant transformation. Radical excision with margins free of disease is the treatment of choice for suspected cases. The response to chemotherapy and radiotherapy remains uncertain. Nevertheless, despite aggressive surgical management, these tumors appear with a high rate of local recurrence and malignant degeneration.[20,21] In summary, we report a very rare case of schwannoma which arises from a diverticulum in the first portion of the duodenum. The treatment period must be organized with a multidisciplinary approach consisting of a pathologist, a radiologist, and a surgeon.

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