Schwannoma of the Duodenum Causing Melena

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A rare case of duodenal schwannoma is reported. A 69-year-old man was admitted for evaluation of melena. Endoscopy and hypotonic duodenography showed a submucosal tumor in the third part of the duodenum. Biopsy findings were suggestive of leiomyosarcoma, therefore pancreateoduodenectomy was performed. Hematoxylin-eosin staining of the resected specimen showed interlacing bundles of spindle-shaped cells with palisading nuclei. Immunohistochemical staining showed positivity for S-100 protein and neuron-specific enolase, but desmin was negative, thus a diagnosis of schwannoma was made. Schwannoma is often difficult to distinguish from leiomyogenic tumors by standard staining, but immunohistochemical staining proved useful in this case.

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Key words: duodenal tumor, S-100 protein, neuron-specific enolase (NSE)

Introduction

Schwannomas are rare tumors arising from Schwann cells which cover the peripheral nerves, and are difficult to distinguish from leiomyogenic tumors. They are extremely rare in the gastrointestinal tract with the stomach being involved 10 times more frequently than the small intestine (1). This report describes a very rare case of schwannoma of the duodenum which was finally diagnosed by immunohistochemical staining.

Case Report

A 69-year-old man was transferred to our hospital for detailed investigation of melena. He had had two previous admissions for the same problem, but upper gastrointestinal tract endoscopy and radiography had failed to detect the source of bleeding. He was pale on admission, with a hemoglobin of 10.9 g/dl. He had no symptoms, abdominal swelling or tenderness. Careful endoscopy revealed a smooth protrusion in the third part of the duodenum. The tumor was covered by normal mucosa and had a shallow depression on its surface, with the base of the depression being slightly congested (Fig. 1). Hypotonic duodenography demonstrated a protrusion in the third part of the duodenum, with a smooth surface, bridging folds, and a shallow collection of barium on its surface (Fig. 2). Selective angiography of the gastroduodenal artery showed hypervascularity of the tumor vessels. As biopsy specimens obtained from the depressed area of the lesion suggested that it was a leiomyosarcoma, pancreateoduodenectomy was performed. The resected specimen showed a submucosal tumor (4.0x3.5x3.0 cm) with a central depression (1.7x1.2 cm) in the third part of the duodenum (Fig. 3a). The surface of the lesion was covered by normal mucosa. The tumor without capsule grew extraluminally for the most part but had no invasion to the neighboring tissue (Fig. 3b). Although hematoxylin-eosin staining (HE) of the tumor revealed interlacing bundles of spindle-shaped cells with nuclei arranged in a vaguely palisading fashion, it was difficult to determine whether the tumor was derived from smooth muscle cells or was neurogenic in origin. The pattern of the cells corresponded to Antoni’s A type with no regressive degeneration, little nuclear mitosis and cellular atypia (Fig. 4a). Immunohistochemical staining for S-100 protein (Fig. 4b) and neuron-specific enolase (NSE) was positive, while staining for desmin was negative. Accordingly, the lesion was finally diagnosed as a schwannoma of the duodenum.

Discussion

Neurogenic tumors of the duodenum are extremely rare. River et al (2) found 90 neurogenic tumors (6.4%) in a series of 1,399 benign tumors of the small intestine, and reported that only 6 (0.43%) of them were located in the duodenum. Sivak et al (3) reported 44 neurogenic tumors (3.2%) in a series of 1,481 small bowel tumors, and stated that 10 of the 28 neurogenic tumors...
Fig. 1. Endoscopy revealed a submucosal tumor in the third part of the duodenum with a shallow depression on its surface.

Fig. 2. Hypotonic duodenography in the prone position demonstrated a smooth protrusion with bridging folds and a shallow collection of barium in the third part of the duodenum.

Fig. 3. a) Photograph showing the resected duodenal tumor and its surface depression. b) Cross-sectional viewing showing that the tumor without capsule grows extraluminally for the most part being mainly located in the muscle layer (×16).

Examined in detail were of duodenal origin. Nilsson and Jonsson (1) reviewed 43 cases of schwannoma of the small intestine, and 8 of the tumors were located in the duodenum. In addition Biese et al (4) reviewed 45 cases of small bowel schwannoma and reported that 10 were located in the duodenum. Duodenal schwannomas are mostly located in the second or third part of this organ (5). Benign schwannomas have been suggested to undergo malignant changes (1, 5), but it is difficult to diagnose malignant schwannoma on a histological basis alone and the diagnosis has to be made from the clinical and macroscopic findings. An indistinct border, invasion of neighboring organs, and metastasis are all signs of malignancy (5). Our case was diagnosed as borderline malignancy, because the tumor did not have capsule and its size was larger than 3 cm in diameter. However, angiography showed no signs of malignancy, such as tortuous irregular vessels, dense inhomogenous opacification, or early venous phase.

The symptoms of neurogenic tumors of the small intestine are vague and nonspecific, with the most frequent symptom being intestinal hemorrhage (2, 4). River et al (2) reported
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Fig. 4. a) Photomicrograph of the tumor revealing interlacing bundles of spindle-shaped cells with the nuclei arranged in a vaguely palisading fashion (HE, x400). b) Immunohistochemical staining of the tumor cells for S-100 protein is positive (x160).

intestinal hemorrhage in 32 out of 90 patients with neurogenic tumors of the small intestine, while Biese et al (4) reported that 23 out of 45 small bowel schwannomas caused intestinal hemorrhage. The hemorrhagic symptoms persisted for 1–15 years before a diagnosis was established and the average delay until operation was around 3 years. In the present case, the tumor was identified 18 months after the episode of melena. Thus, thorough examination of the small intestine should be undertaken in patients with a history of multiple episodes of massive upper gastrointestinal hemorrhage of unknown etiology.

Also in our patient, the source of bleeding might be from the central depression, because histologic examination disclosed a small part with thin epithelium and without lamina muscularis mucosae.

On histological examination, it is often difficult to distinguish schwannoma from a leiomyogenic tumor using hematoxylin-eosin staining alone. The immunohistochemical detection of S-100 (6) and NSE (7) may be useful for making an accurate diagnosis. S-100 protein is a nervous tissue-specific protein (8), while NSE is a molecular marker of peripheral and central neuroendocrine cells (9). All benign tumors arising from Schwann cells are S-100 protein positive, while malignant schwannoma is either positive or negative depending on the level of differentiation (10). In contrast, fibrosarcoma and leiomyosarcoma are negative for S-100 protein (10). In the present case, S-100 protein and NSE were positive, while desmin as a marker for smooth muscle cells (11) was negative. Therefore, the tumor was diagnosed as a duodenal schwannoma.

In this report, we presented a very rare case of duodenal schwannoma and emphasized the usefulness of the immunohistochemical staining methods for an accurate diagnosis.

References