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Case Report

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DUODENAL MALIGNANT SCHWANNOMA: A NEW CASE REPORT

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ABSTRACT

Summary: Schwannomas are very infrequent tumors originating from Schwann cells of the neural sheath.[3] Gastrointestinal schwannomas are among the few mesenchymal tumors of the gastrointestinal tract, and only a few cases have been reported to date. Duodenal schwannomas are usually discovered accidentally and making a preoperative diagnosis remains difficult. We report a case of duodenal schwannoma discovered during abdominal pain assessment. In imaging, this tumor was large and locally advanced, at the endoscopy, the appearance was ulcero-budding.

KEYWORDS: duodenal schwannoma, Malignant Peripheral Nerve Sheath Tumors, Malignant Digestive Neurilemmoma

INTRODUCTION

Schwannomas, also called "neurilemmomas", are benign tumors derived from Schwann cells of the neural sheath. These are among the rare mesenchymal tumors of the gastrointestinal tract (GI), their incidence is estimated at 2% - 6% of all subepithelial tumors of the gastrointestinal tract.[1,2] GI schwannomas occur most often in the stomach, followed by the colon and rectum.[2] While the grelic localization of schwannomas is rarely reported.3. The clinical course of gastrointestinal schwannomas is not specific and their preoperative diagnosis remains difficult.[4] Schwannomas can be distinguished from other subepithelial tumors by endoscopic ultrasound (EUS)[5] and immunohistochemistry. A positive reaction for the S-100 protein is favorable for the diagnosis of schwannomas.[6] We report here the case of a patient with duodenum schwannoma confirmed by immunohistochemistry

OBSERVATION

This is a 54-year-old patient who consulted for abdominal pain that had been evolving for 3 months, associated with a deterioration of the general condition. The thoraco pelvic abdominal CT scan showed a large tumor mass centered on duodenal light (D2-D3) measuring 90 / 60mm, tissue, necrotic in the center, raised after injection of contrast medium heterogeneously and having an intimate contact with the hepatic pedicle judged therefore unresectable. Oeso-gastroduodenal fibroscopy revealed a non-stenosing ulcero-budding tumor in the second portion of the duodenum that was 2 cm long. After multiple biopsies, the pathological study was in favor of a fusocellular tumor proliferation first evoking a stromal tumor (GIST) (fig 1), an immunohistochemical complement was made in favor of a tumor proliferation of schwanian nature without atypia marked, without mitoses and without foci of necrosis in favor of the malignant character of the tumor (immuno-reactivity S100prot) (fig 2). After carrying out a molecular biology study, there was no mutation in the different exons of the Kit and PDGFR genes, the molecular profile has therefore made it possible to eliminate a mutated gastrointestinal stromal tumor Kit or PDGFR. Because the patient was not eligible for chemotherapy, she died after two courses of doxorubicin alone.

DISCUSSION

Schwannomas are rare tumors resulting from Schwann cells, which cover the peripheral nerves, and are difficult to distinguish from other mesenchymal tumors. The most common gastrointestinal location of schwannomas is the stomach, whereas duodenal waiting remains extremely rare.[7] Duodenal schwannomas are mainly located in the second or third part of the duodenum.[8] Nilsson and al. Reported that, of 43 schwannomas of the small bowel, eight were localized in the duodenum.[9] There was no difference in incidence between men and women, especially in the fifth and sixth decades of life.[6] Our patient was in the 5th decade and the tumor was located

in the 2nd-3rd duodenum, which joins the cases reported in the literature. Duodenal schwannomas are usually asymptomatic and can be discovered by chance. If the tumor is symptomatic, the most common symptoms are often digestive bleeding or abdominal pain.[8] Duodenal schwannomas are usually benign, the malignant form is very rare[10] The main means of diagnosis of schwannomas are; endoscopy, abdominal echo-endoscopy, CT scan and / or abdominal magnetic resonance imaging (MRI) to determine tumor localization, locoregional and distant extension.[12,13] However, schwannomas can not be distinguished from other neurogenic malignancies only by imaging means. The histological study is essential for a definitive diagnosis. Schwannomas are covered by intact mucosa and usually involve the submucosa and muscularis. Immunohistochemistry is essential to distinguish between schwannomas and GIST or leiomyomas.[11] The schwannoma cells are 100% immunoreactive with S-100 protein, GISTs generally express c-kit and CD34, but without reactivation with S-100 protein, whereas leiomyomas are positive for smooth muscle fibers actin and desmin, and negative for the S-100 protein.[1] The optimal treatment for a malignant schwannoma has not been fully established.[6] The role of chemotherapy and radiotherapy remains unclear. Incomplete resection may be associated with local recurrence, so surgical margins are considered the primary prognostic factor.

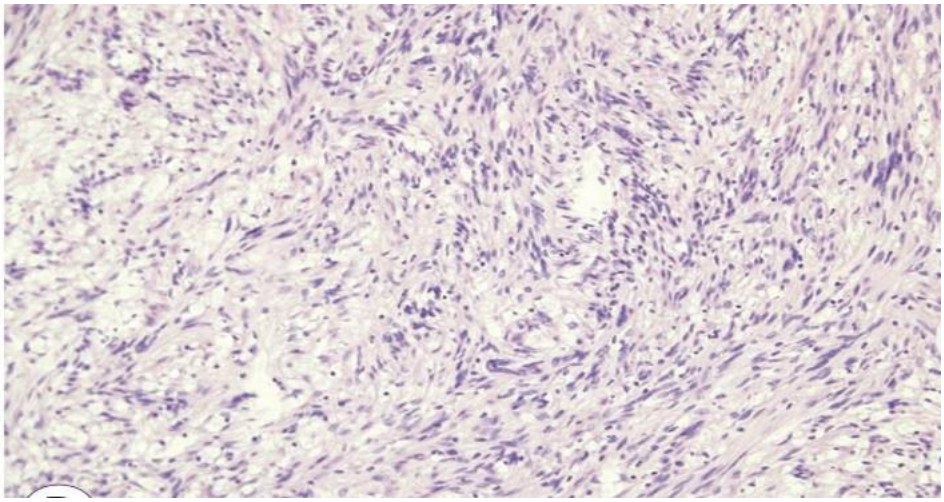

Figure1: Elongated fusiform cells with undulating tapered nuclei, with abundant clear cytoplasmic vacuoles.

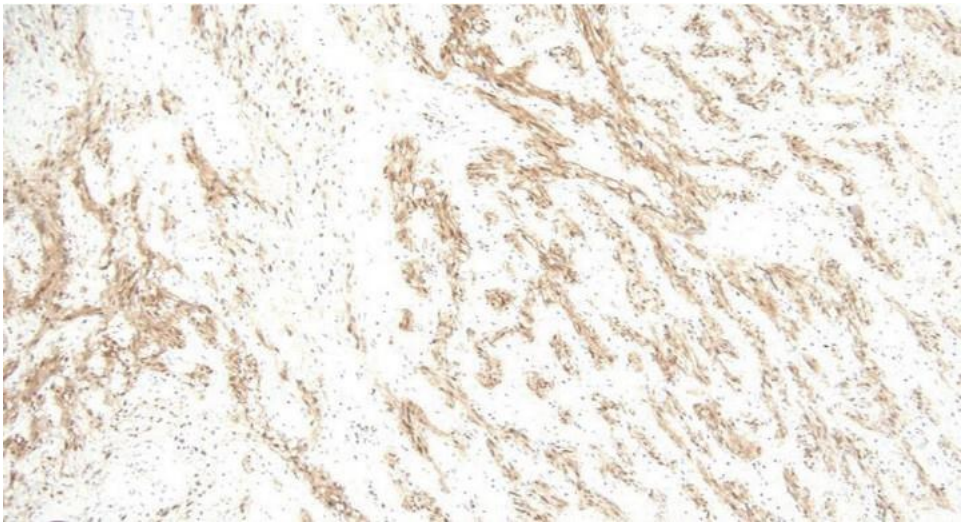

Figure 2: Intense and diffuse nuclear staining of cells after immunohistochemistry with anti PS100 antibody.

In conclusion, we report a very rare case of schwannoma located at the duodenal level. Due to the difficulty of distinguishing schwannomas from other mesenchymal tumors, the diagnosis as well as the treatment of this tumor can be delayed. The definitive diagnosis can be established based on radiological (abdominal CT, EUS and MRI) and histological (IHC) studies before any surgical resection.

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