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ScienceDirect
Pathology – Research and Practice 203 (2007) 667–670
PATHOLOGY RESEARCH AND PRACTICE
www.elsevier.de/prp

TEACHING CASES

Synchronous GIST with osteoclast-like giant cells and a well-differentiated neuroendocrine tumor in Ampula Vateri: Coexistence of two extremely rare entities

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Received 4 October 2006; accepted 25 April 2007

Abstract

Mesenchymal tumors of the gastrointestinal system with variable histopathological appearances and constant expression of CD117 are known as gastrointestinal stromal tumors (GISTs). Neuroendocrine tumors may be seen in the gastrointestinal system and other organ systems of the body. We report a 44-year-old male patient with a $6.5 \times 3 \times 6$ cm mass located in the Ampulla of Vater. Histopathologic examination revealed a GIST with a marked nuclear pleomorphism and a high mitotic rate, and that was rich in osteoclast-like giant cells (OGC). Immunohistochemically, GIST was positive for CD117, while OGCs were negative for CD117 and positive for CD68 and α_1-antitrypsin. There was also found a well-differentiated neuroendocrine tumor near the GIST, in the serosal aspect of the duodenum at the point of the Ampulla of Vater. This second tumor was 20 mm in diameter, and was relatively well circumscribed with few glands invading the GIST. This tumor was positive for synaptophysin and chromogranin. Neither mitosis nor vascular invasion was observed. The patient had no familial history or clinical manifestations of neurofibromatosis. This case presents the unique synchronous existence of two extremely rare entities, a GIST with OGC and a well-differentiated neuroendocrine tumor, both located in the Ampulla of Vater.

Keywords: Gastrointestinal stromal tumor; Osteoclast-like giant cells; Neuroendocrine tumor; Ampulla of Vater

1. Introduction

The term gastrointestinal stromal tumor (GIST) refers to the mesenchymal tumors of the gastrointestinal system other than the gastrointestinal counterparts of the specific soft tissue tumors. Since the first description of GIST as a distinct entity, numerous case reports have appeared in the literature, and a number of variants have been described. GIST with osteoclast-like giants cells is one of them, and since its first description by Leung et al in 2001, only two cases have been reported [4,5]. Synchronous GIST and neuroendocrine tumors have also been described [1]. The Ampulla of Vater is a very rare localization both for GISTs and for well-differentiated neuroendocrine tumors [9]. To the knowledge of the authors of this manuscript, the case presented here is unique for the simultaneous occurrence of a GIST with osteoclast-like giant cells (OGC) and a neuroendocrine tumor in the Ampulla of Vater.

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doi:10.1016/j.prp.2007.04.012

2. Case report

A 44-year-old male patient was admitted to the hospital with lack of appetite and 7 kg weight loss in 3 months. The patient's abdominal pre- and post-contrast CT scans revealed a primary tumor in the second part of the duodenum. It was of soft tissue density and measured $6.5 \times 3 \times 6$ cm (Fig. 1). The mass was compressing the pancreatic head. The intrahepatic biliary tree, the main duct channel (8 mm), and the pancreatic duct (3 mm) were minimally dilated. The pancreas was otherwise normal. ERCP revealed an ulcerated mass in ampulla. An endoscopic biopsy was taken and reported as stromal tumor. The patient underwent pancreatico-duodenectomy. No complication occurred, and the patient was discharged on the 10th postoperative day.

In the pancreatico-duodenectomy material, there was a $9 \times 8 \times 8$ cm mass in the duodenum surrounding the Ampulla of Vater. The mass had an expansile growth pattern through the wall of the duodenum, and the overlying mucosa was ulcerated. The cut surface of the mass was white-yellow, lobulated, and contained foci of hemorrhage. Other parts of the duodenum were normal in appearance. The mass located around the Ampulla of Vater pushed the pancreas but did not infiltrate it. No mass was observed in the cut surface of the pancreas, and the pancreatic duct was minimally dilated. Extensive sampling was done.

Microscopic examination revealed a highly cellular mass that was composed of fusiform cells forming short fascicles and scattered epitheloid cells. A moderate to marked degree of nuclear pleomorphism, and $\frac{30}{50}$ hpf mitosis were observed (Fig. 2). Invasion of the tumor led to ulceration of the mucosa, but the borders of the tumor were relatively well demarcated in the deeper layers of the intestinal wall. There were groups of multinucleated osteoclast-like giant cells, each containing 20–30 nuclei, scattered throughout the tumor (Fig. 3). These cells had no mitosis or nuclear pleomorphism. PAS reaction revealed skeinoid fibers. Immunohistochemically, fusiform and epitheloid cells were positive for *c*-kit (CD117), negative for S-100, NSE, sinaptophysine, chromogranine, HMB-45, smooth muscle actin (SMA), desmin, CD68, and lysosime, while OGC were positive for CD68 and lysosime, and negative for the rest (Fig. 4).

In one sample taken from the lesion, we noted another tumor that consisted of glands and cribriform formations, and that was composed of bland appearing cells with small round nuclei and eosinophilic cytoplasm. This second tumor was located in the serosal aspect of the duodenum at the point of the Ampulla of Vater near the GIST (Fig. 5). No mitosis was observed. The lesion was relatively well circumscribed and was 20 mm in diameter. Few glands of this second tumor invaded the GIST. In immunohistochemical study, the tumor cells were stained with synaptophysin and chromogranin (Fig. 6).

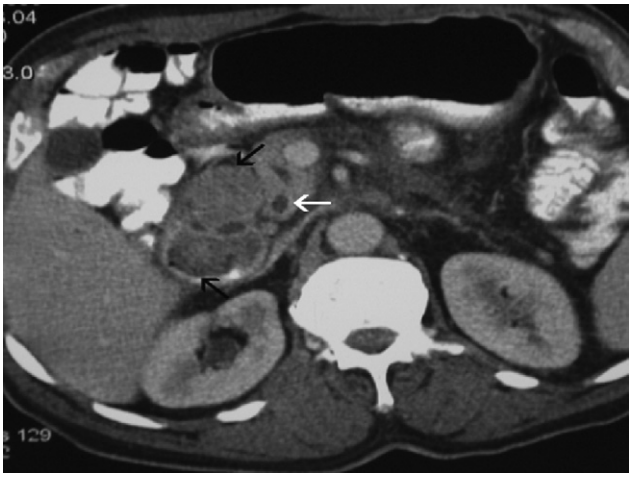

Fig. 1. CT image of the tumor located in the second part of the duodenum.

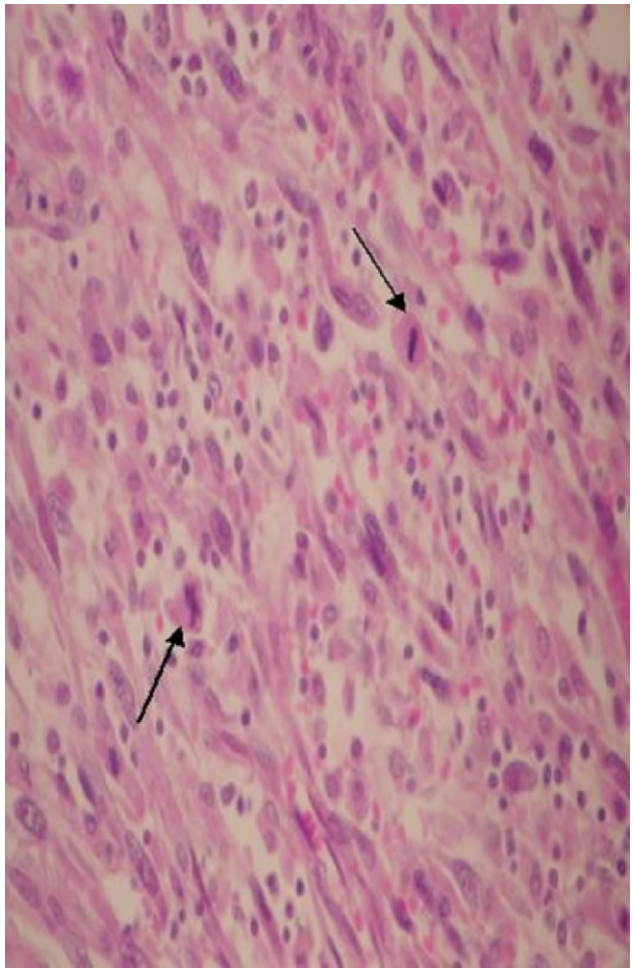

Fig. 2. A marked degree of nuclear pleomorphism and numerous mitoses were observed (arrows: mitosis) (HE $\times$ 400).