



Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Endoscopic resection of a periampullary gangliocytic paraganglioma of the duodenum: A case report

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ARTICLE INFO

Article history:

Received 28 September 2016

Accepted 25 October 2016

Available online 27 October 2016

Keywords:

Gangliocytic paraganglioma

Histomorphology

Immunohistochemistry

Tumour

Neuroendocrine tumours

Case report

ABSTRACT

INTRODUCTION: The gangliocytic paraganglioma (GP) is an extremely rare neuroendocrine tumour originating from the second part of the duodenum. Generally GP shows benign clinical behaviour. The GP is typically characterized by consisting of three tumour components: the epithelioid, the spindle-shaped and the ganglion-like cells.

PRESENTATION OF CASE: We present a female patient at the age of 65, who underwent a routine gastroscopy due to known gastro-oesophageal reflux. Accidentally a 2 × 1,5 cm sized mass of unknown entity was revealed in the duodenum. The magnet resonance tomography neither detected distant metastasis nor any local lymphadenopathy. After endosonographically guided punctures of the submucosal mass, a malignant tumour could not be diagnosed thus the decision to perform an endoscopic resection was made and successfully conducted. Immunohistochemical examination revealed a total resected GP.

DISCUSSION: In literature malignant transformation with distant metastasis and local recurrences has been described. Furthermore the clinical manifestation and location varies. The GP has often been misdiagnosed as a neuroendocrine tumour (NET) G1.

CONCLUSION: Due to published cases of metastasising GPs, we recommend a long term follow-up. In a non-metastatic stage the endoscopic resection should be the therapy of choice in order to prevent unnecessary major surgical interventions. In this case report we will discuss the clinical appearance, behaviour and differential diagnosis of GP.

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1. Introduction

The GP is a rare submucosal gastrointestinal tumour with mostly benign clinical behaviour. The tumour was first described by Dahl et al. in 1957 [1]. Generally the GP is characterized by consisting of three tumour components: spindle-shaped cells with Schwannian differentiation, ganglion-like cells and neuroendocrine cells [2,3]. These cells can be immunoreactive to pancreatin, synaptophysin, chromogranin and are able to express S-100, NSE, pancreatic polypeptide, somatostatin and vasoactive intestinal peptide [2]. The spindle cells are characterized by forming fascicles or envelope nerve cells and axons expressing the S100 protein. The ganglion cells are scattered as single elements or appear aggregated in clusters [4].

The GP typically occurs in the 5th decade of life [5]. Men are more often affected than women (1,8: 1). A literature review by Okubo et al. revealed that approximately 90% of the published cases of

GPs originate from the duodenum [2]. These tumours are classified as neoplasia of the digestive tract by the World Health Organisation [4]. In literature the GP has been described as appearing in the esophagus, jejunum, pylorus and even in the retroperitoneal space [6]. The main symptoms are mostly abdominal pain, discomfort, anaemia and upper gastrointestinal bleeding. To diagnose a GP can be extremely challenging. Due to the submucosal location of GP, an endoscopic biopsy often times presents negative results [4]. Moreover a GP can be confirmed by computer tomography, ultrasound imaging, magnet resonance tomography and detection of neuronal and neuroendocrine markers such as synaptophysin, chromogranin A, CD56, S-100, NSE and somatostatin in the blood [7]. Additionally the diagnosis can be supported by immunohistochemical findings such as oestrogen, progesterone receptors, pancreatic polypeptide, somatostatin and vasoactive intestinal polypeptide (VIP) [7]. Therapy of choice is the local endoscopic excision. In case of lymph node or distant metastasis, a pancreaticoduodenectomy is recommended [8].

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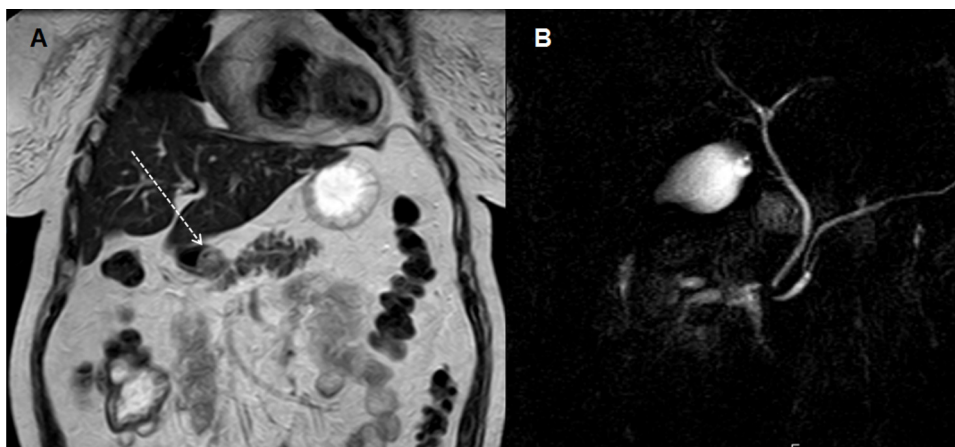


Fig. 1. Magnetic resonance imaging of the upper abdomen (A). Visible is a 2 × 1,5 cm sized mass in the duodenum (marked by white dashed arrow). Distant metastasis and lymphadenopathy are not visible. The magnetic resonance cholangiopancreatography does not reveal a tumour originating from the biliary duct system (B).

2. Presentation of case

In November 2015 a 65 year old women was treated in our hospital due to a submucosal tumour of uncertain origin. Her medical history included a diverticulosis and rheumatoid arthritis. The patient suffered from a hiatus hernia and gastro-oesophageal reflux. Therefore she routinely underwent a gastroscopy at her local hospital, where a 2 × 1,5 cm sized mass was found in the second part of the duodenum. After performing magnetic resonance tomography with cholangiopancreatography distant metastasis and lymphadenopathy were not detected (Fig. 1).

Due to the submucosal location several attempts to obtain a histopathological diagnose of the tumour were unsuccessful. Under suspicion of a malignant tumour such as carcinoma of the duodenal papilla the patient was advised to undergo an open surgery exploration. She rejected the proposal, yet she came to our hospital for a second opinion and further diagnostic.

The physical examination did not reveal any pathological findings. Furthermore the laboratory test demonstrated normal levels of C-reactive protein, leucocytes, CA72-4 and CA19-9. The tumour marker CEA was slightly increased (4,1 µg/l). We performed a gastroduodenoscopy with ultrasound guided puncture and endosonographic imaging of the submucosal tumour. The mass was anechoic and seemed to be located in all wall layers. Swollen lymph nodes were not detected. The histopathological examination revealed a tumour arising from the submucosa, covered by mucous membrane and surrounded by smooth muscle. Due to the lack of distant metastasis and lymphadenopathy, the decision for an endoscopic removal of the tumour was made. Under analgesic sedation we performed a gastroduodenoscopy. The mass of unclear entity was located approximately 2 cm above the *Papilla Vateri*. After submucosal injection with adrenalin and mucosal dissection the tumour was successfully removed via cautery snare. The mass seemed to originate from the *muscularis propria*. The defect of the mucosa was covered with two clips (Fig. 3). The post interventional course was without complications and the patient left the hospital within the next 2 days (Fig. 2).

2.1. Pathological findings

The histopathological examination reveal a GP. Fig. 4 shows a 19 × 15 × 10 mm large tumour with yellow-grey cut surface as a submucosal mass with trilinear differentiation.

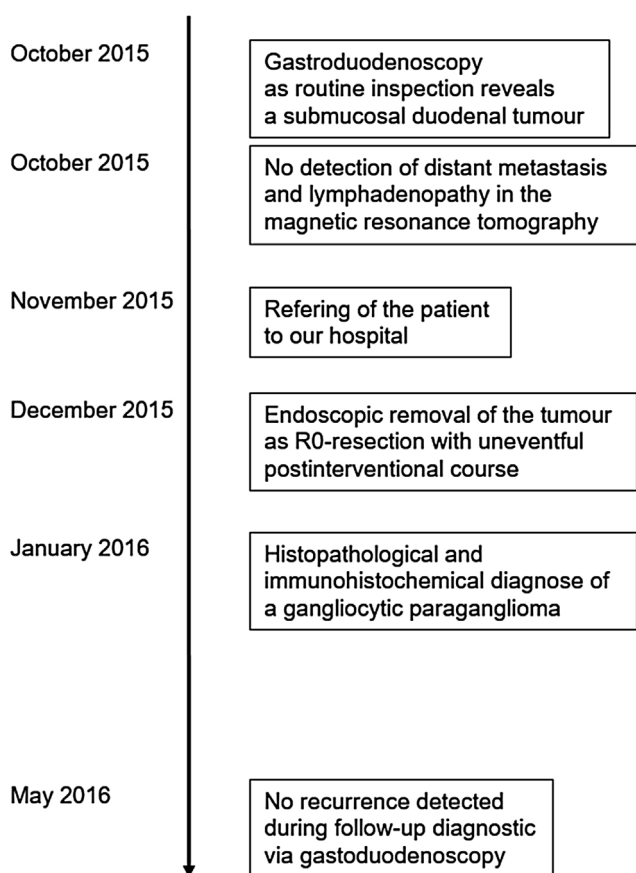


Fig. 2. The Figure indicates the timeline of this case report.

2.2. Immunohistochemical findings

Immunohistochemical findings are imaged in Fig. 5. Neuronal, gangliocytic and nest-like epitheloid structures are imaged in Picture 1. The S100 expression of spindle cell with Schwann-like differentiation is imaged in Picture 2. The neuroendocrine cell and ganglion cells show synaptophysin expression is demonstrated in Picture 3. The VIP expression is shown in Picture 4.

As follow-up diagnostic we performed a gastroduodenoscopy 3 month after the endoscopic removal of the GP. No recurrence was

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