Neurofibroma of the stomach without Recklinghausen's disease: a case report

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Abstract

Neurofibromas of the stomach can occur in the course of Recklinghausen's disease. Sporadic gastric neurofibroma appears rarely. This tumour may look like an ulcer and can be a cause of abdominal pain, nausea, and bleeding from the gastrointestinal tract. We reported a 61-year-old women complaining of stomachache for several months. Gastroscopy revealed a tumour with ulceration in the prepyloric part of the stomach. *Helicobacter pylori* infection was also present. *Helicobacter pylori* eradication and prolonged treatment of proton pump inhibitors did not decrease the ailments or the size of the tumour. It was not possible to determine the nature and origin of the tumour by carrying out examinations such as endoscopic ultrasound and computed tomography of the abdomen. Only after surgery and histopathological examination with immunohistochemistry was this tumour identified as a neurofibroma. In order to differentiate the tumour the following immunohistochemical examinations were carried out: CD34 (slightly +), CD117 (–), S-100 (+), desmin (–), NSE (+), GFAP (–), SMA (–), bc12 (–), CD99 (–), ALK1 (–), and MiB (1–1.5%). In such cases excision of the tumour is the preferred treatment.

Introduction

Neurofibromas of the stomach can occur in the course of Recklinghausen's disease-neurofibromatosis type 1 (NF-1) and are diagnosed in about 52% of such cases [1]. Neurofibromatosis type 1 is a multisystemic disorder that can affect several organs [2]. Sporadic gastric neurofibroma occurs considerably less often. It may have a course of abdominal pain, nausea, dyspepsia, bleeding from the gastrointestinal tract, and sometimes stenosis of the pylorus [3]. The recognition of this tumour is difficult and requires many diagnostic procedures. Gastroscopy is ineffective in differentiation of ulcer, gastrointestinal stromal tumour (GIST), and cancer. Other examinations (radiological and laboratory tests) did not give a proper diagnosis.

Case report

We present the case of rare stomach tumour. A 61-year-old women presented to our outpatient clinic in May 2009. She complained of stomachache for the preceding several months. Burning and gnawing pain was localised in the upper abdomen and occurred before meals. Physical examination revealed pain in the upper abdomen under palpation and no other abnormalities. The patient looked slim, but she had not noticed any loss of weight recently. Gastroscopy was performed. This examination showed atypical changes in the prepyloric part of the stomach. It looked like two tumours with a depression in the middle and with ulceration. The diameter of first tumour was about 3 cm, the second about 2 cm, and part of the ulceration was covert due to normal looking mucosa (Figure 1). A biopsy was taken and the description was as follows: chronic acute gastritis middle degree with focal lymphoplasia and hyperplasia. Necrotic-inflammatory masses were described. A large number of *Helicobacter pylori* (*H. pylori*) were presented. The women obtained *H. pylori* eradication but it failed to decrease her symptoms. Also, prolonged

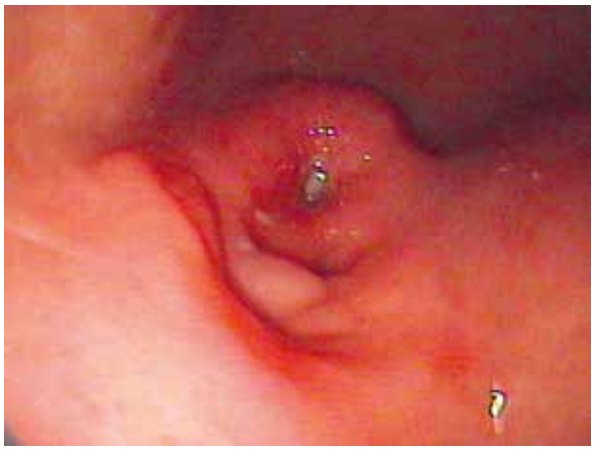

Figure 1. The tumour in gastroscopy. There is ulceration in the middle of the tumour

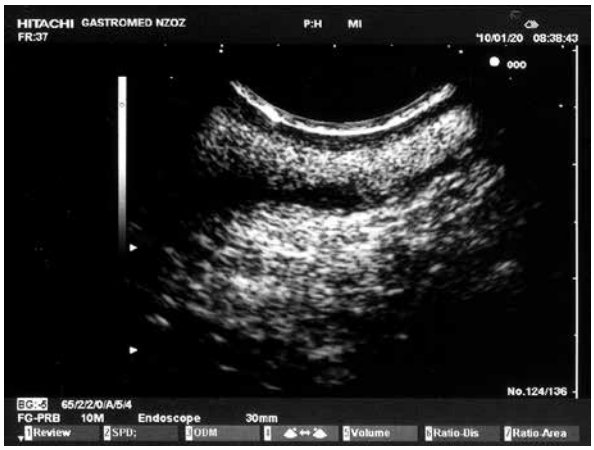

Figure 2. EUS examination. Soft, hyperechogenic, homogenous mass with hypoechogenic border

treatment of proton pump inhibitors did not decrease her ailments or the size of the tumour. Standard laboratory examination did not reveal any deviations apart from a slight increase in insulin level. Endoscopic examination performed after 6 months showed the same picture as the first one. Repeated biopsy showed the same results. *Helicobacter pylori* infection was still present. In subsequently performed endoscopic ultrasonography (EUS) a soft, hyperechogenic, homogenous mass with a hypoechogenic border thickness of about 5 mm was observed. Under the pressure of the endoscope head this change was flattened out to prevent assessment of the layer from which it originated (Figure 2). Due to an unclear picture in gastroscopy and endoscopic ultrasonography, computed tomography (CT) was performed, which showed thickening of the stomach wall in the previously described place. It was not possible to determine the nature and origin of the tumour by carrying out an examination. Due to serious suspicion of neoplastic process the woman was referred to the Surgical Department. Her abdomen was opened by medial section and an intramural tumour of 4 cm diameter was found. Enlarged lymph nodes were also noticed in the lesser omentum. The remaining abdominal organs were normal. A performed intra-operative histological examination did not reveal carcinoma cells. The tumour was removed and a Billroth I operation was performed. A histological examination made after the surgery revealed a gastric tumour, immunohistochemically differentiated as a neurofibroma (Figure 3). The tumour occupied both submucosa and mucosa. For differentiation of the tumour the following immunohistochemical examinations were performed: CD34 (slightly +), CD117 (–), S100 (+), de-

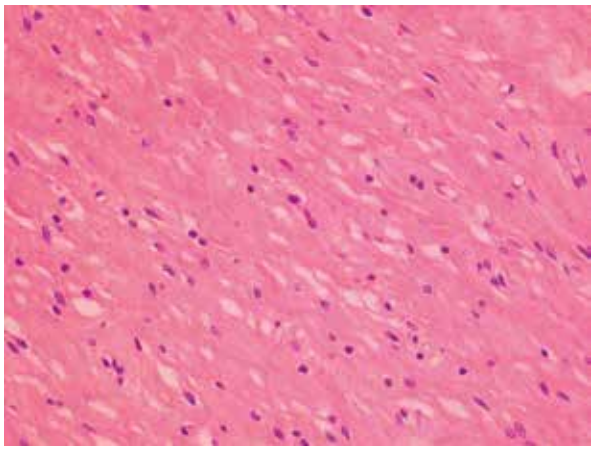

Figure 3. Histological examination shows spindle-shaped cells with elongated nuclei set in fibromyxoid stroma. H + E, 10×

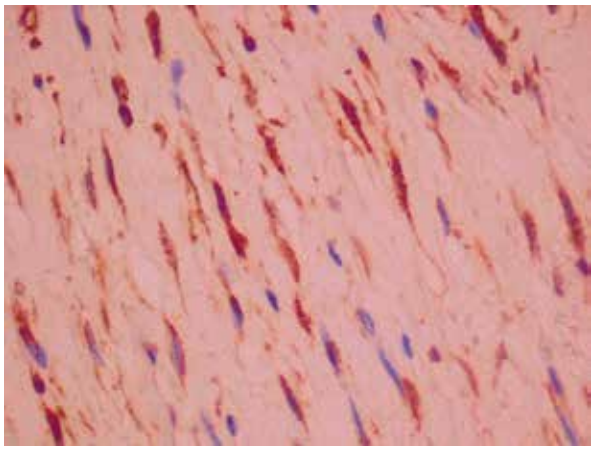

Figure 4. S-100 20×. Immunohistochemical staining: tumour cells are positive for S-100 protein

smin (–), neuron specific enolase-NSE (+), glial fibrillar acidic protein-GFAP (–), smooth muscle actin-SMA (–), bc12 (–), cluster of differentiation-CD99 (–), and MiB (1–1.5%). Tumour cells are strongly positive to S-100 protein (Figure 4). This tumour is likely to be benign. Histological examination of the lymph nodes showed only reactive lymphadenitis. The recovery after operation was typical. Two years later the patient feels good, she has no stomach pain, and looks normal.

Discussion

In the literature we found only a few described cases of stomach fibroma, but most of them were connected with the Recklinghausen's disease, which makes it easier to present a diagnosis [4]. Sometimes that tumour is disclosed by gastrointestinal bleeding and sometimes it is a cause of pyloric stenosis. Neurogenic gastric tumours are usually benign and only 10% of neurofibromas can undergo malignant transformation [5]. We reported that a neurofibroma of the stomach without Recklinghausen's disease endoscopically looked like an ulcer or GIST and required detailed diagnosis before surgical treatment (gastroscopy, EUS, CT). Wide excision of the tumour seems to be the treatment of choice [6]. Histological examination confirmed the diagnosis. The large amount of collagenous fibrous tissue that distinguishes them from neurinomas was found. The tumours are essentially composed of proliferated Schwann cells [7]. Schwannomas and neurofibromas arise from peripheral nerves. Schwannomas consist of Schwann cells, while neurofibromas contain several cellular components, including Schwann cells, perineurial-like cells, and endoneurial fibroblasts. S-100 protein has been used as an adjunct in the differential diagnosis between schwannomas and neurofibromas [8]. Immunohistochemical examination for S100 protein and neurofilaments are usually positive due to the presence of residual myelinated nerve fibres. The presence of S100 proteins also distinguishes neurofibromas from intramuscular myxomas. Gastrointestinal stromal tumours are increasingly being recognised due to their characteristic expression of CD117. However, our tumour did not express this protein. Sometimes a cluster of differentiation 34 (CD34)-positive cells may also be seen. We diagnosed fibroma by three immunostaining methods. Positive CD34 immunostaining and negative S100 distinguish dermatofibrosarcoma protuberance from diffuse neurofibroma. On the other hand, gastric glomus tumour is strongly positive for SMA but CD34, CD117, and desmin negative.

Liu *et al*. [9] reported GIST of stomach morphologically resembling a neurofibroma. The research describes the GIST tumour as being negative for CD117 expression and composed of bland spindle cells reminiscent of a neurofibroma. They proposed detection of a new platelet-derived growth factor receptor α mutation in exon 18 (D842F) to identify CD117-negative GISTs. It may be a diagnosis of the future. Our case report is interesting because of its rarity, appearance and clinical features like an ulcer, and difficulties with diagnosis.

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