SCRIPTA MEDICA (BRNO) – 75 (1): 7–14, *February 2002*

A RARE CASE OF MALIGNANT EPITELIOID GASTROINTESTINAL STROMAL TUMOUR. CASE REPORT

STAFFA R.[1], STUPKA Z.[1], KRPENSKÝ A.[2], HUŠEK K.[2], ČERNÝ E.[2], JELÍNKOVÁ I.[2]

[1] Second Department of Surgery and [2]First Department of Pathological Anatomy, St. Anne's Teaching Hospital, Faculty of Medicine, Masaryk University, Brno.

A b s t r a c t

A middle-aged men suffering from protracted dyspepsia was admitted to the Second Department of Surgery. Endosonography revealed an exophytic gastric tumour. Partial gastrectomy with enteroanastomosis (Billroth II) was performed. Histological and immunohistological examination revealed a malignant epithelioid gastrointestinal stromal tumour (GIST), which is a neoplasm rarely reported in the literature. Two years later the patient showed no signs of tumour recurrence or metastatic dissemination and his dyspeptic complaints disappeared. He was regarded as a potentially healed patient.

K e y w o r d s

Gastrointestinal stromal tumour, Chronic dyspepsia, Surgical treatment

INTRODUCTION

Although the incidence of malignant gastric tumours has a tendency to decrease, patients suffering from protracted dyspepsia may be affected with a latent neoplastic process that can remain undetected in spite of modern invasive diagnostic methods. In such cases, surgical intervention is indicated because it is the only way of how to prevent dissemination of the tumour and to save the patient. This procedure also confirms the clinical suspicion and facilitates exact diagnosis by means of intraoperative biopsy (*1,2*). In chronic dyspepsia, a comprehensive approach, involving close and early cooperation of internists and surgeons, is necessary. The case presented in this paper demonstrates the importance of early surgical intervention.

CASE REPORT

The patient S.O., born in 1945, suffered from long-lasting dyspepsia; he underwent a series of routine tests without a definite diagnosis being made. Utrasonography revealed some signs indicative of a neoplastic process in the epigastric region. To confirm the diagnosis, the patient was admitted to our department on 12th April 1999. CT examination showed the presence of a neoplastic lesion with a suspected localisation in the left liver lobe.

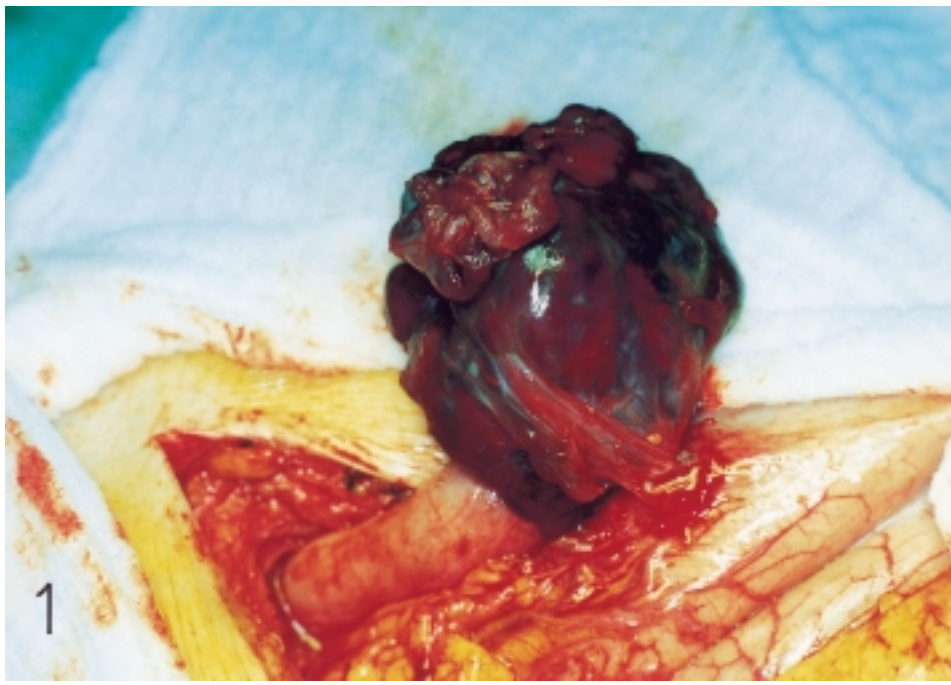

Fig. 1
Gastrointestinal stromal tumour growing on the outside of the stomach wall;
an intraoperative finding.

Gastrofibroscopy was performed but gave a negative result. A repeated sonographic examination failed to confirm the diagnosis, it only indicated an association of the suspected lesion with the pancreas. In the end, endosonography located the tumour to the region of the lesser stomach curvature without any evidence of spread into other organs.

Surgical treatment was indicated and further tests were made, including aspiration cytology under sonographic control. In the aspiration sample, there were fusiform cells whose characteristics were difficult to define. Routine examination for neoplastic blood markers was negative.

The patient was operated on one week after admission (April, 19, 1999). Important data from the operation record are as follows: from the external surface of the stomach, an exophytic tumour protruded in the region of the lesser curvature; a short, thick stalk connected the tumour with the stomach wall; the size of the tumour was equal to a child's fist; its surface was uneven and the tissue showed hemorrhagic foci (*Fig.1, 2*). A intraoperative biopsy sample was taken and was indicative of malignancy. Partial resection of the stomach was performed,

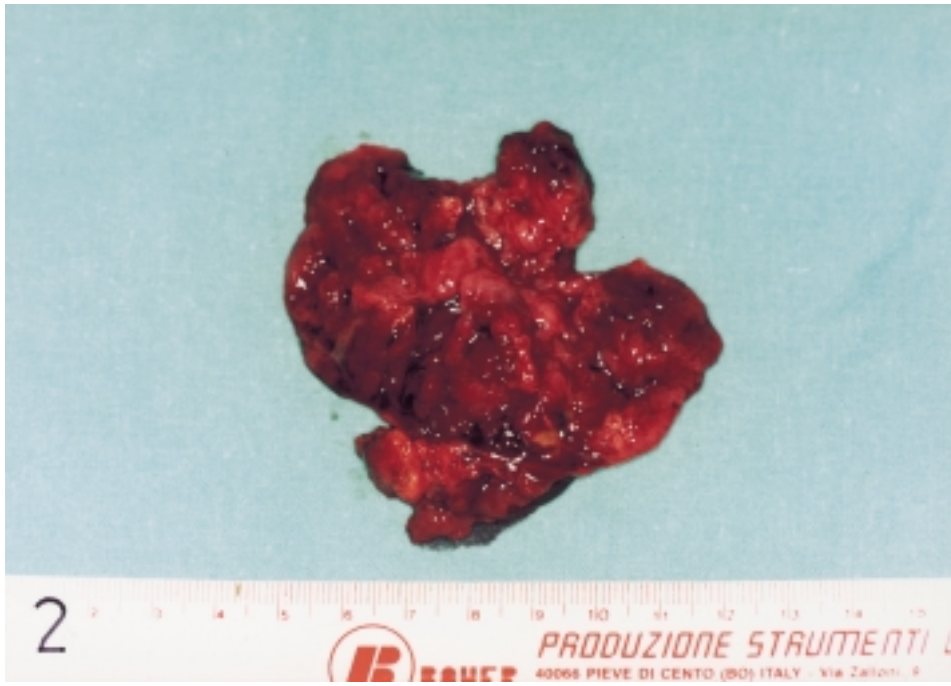

Fig. 2
Macroscopic findings after excision of the tumour

using a gastroenteroanastomosis according to Billroth II. A careful inspection of the abdominal cavity failed to reveal any signs of metastatic dissemination or other pathological lesions. All tissues removed by surgery were sent for histological examination to the Department of Pathology.

Histological findings reported by Krpenský. 1. Tumour: the histological presentation was essentially the same as that of the intraoperative sample. It consisted of fusiform and polymorphous cells with scanty mitotic figures. The cells showed neurogenic and myogenic differentiation so that, with regard to its location, the tumour was classified as a gastrointestinal stromal tumour. 2. Tissue bordering the resection lines: showed mild, focal mesothelial hyperplasia of the peritoneum without the presence of neoplastic cells. 3. Wall of the stomach: no signs of tumour cell infiltration. 4. Other samples were examined and showed monomorphic, medium-size epithelioid cells with some glycogen inclusions. The nuclei were ovoid with fine chromatin structures and lacked visible nucleoli. Mitotic activity varied. Tumour cells were examined by immunohistochemistry; desmin gave a strong positive reaction but smooth-muscle actin and S 100 were

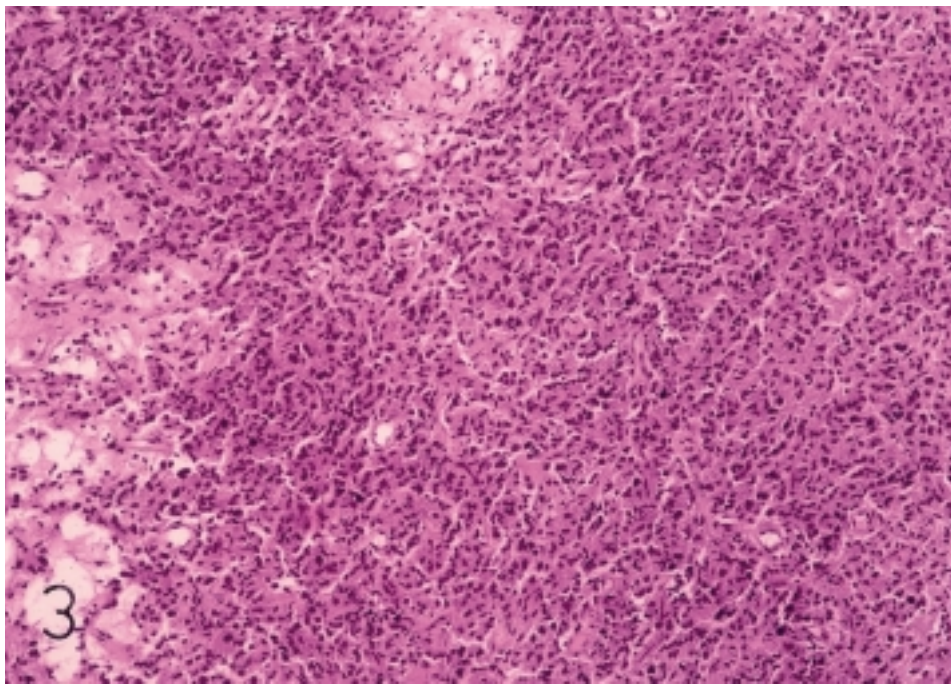

Fig. 3
Microscopic findings with the predominant epithelioid cell type. Stained with haematoxilin and eosin. Magnification, x 10

negative. The histological presentation and immuno- profiles of the tumour cells allowed us to classify the malignancy as an epithelioid stromal gastrointestinal tumour (*Fig 3, 4*).

The patient's postoperative course was favourable with *per primam* healing. The patient was on a complete parenteral regimen. His inner environment was stabilised and thromboembolism was prevented (*5*). He was gradually converted to solid food and, when his gastrointestinal passage was found to be normalised, he was discharged on April 28, 1999. Rehabilitation and an appropriate dietary regimen were recommended. Consultation with oncologists resulted in a mere follow-up since no specific therapy seemed to be indicated. At regular check-ups at both our department and the cancer hospital, the patient did not present with any signs of either disease recurrence or its dissemination. He reported to tolerate food intake well and without any dyspeptic problems. At 2 years after surgery we could consider this patient to be potentially healed.

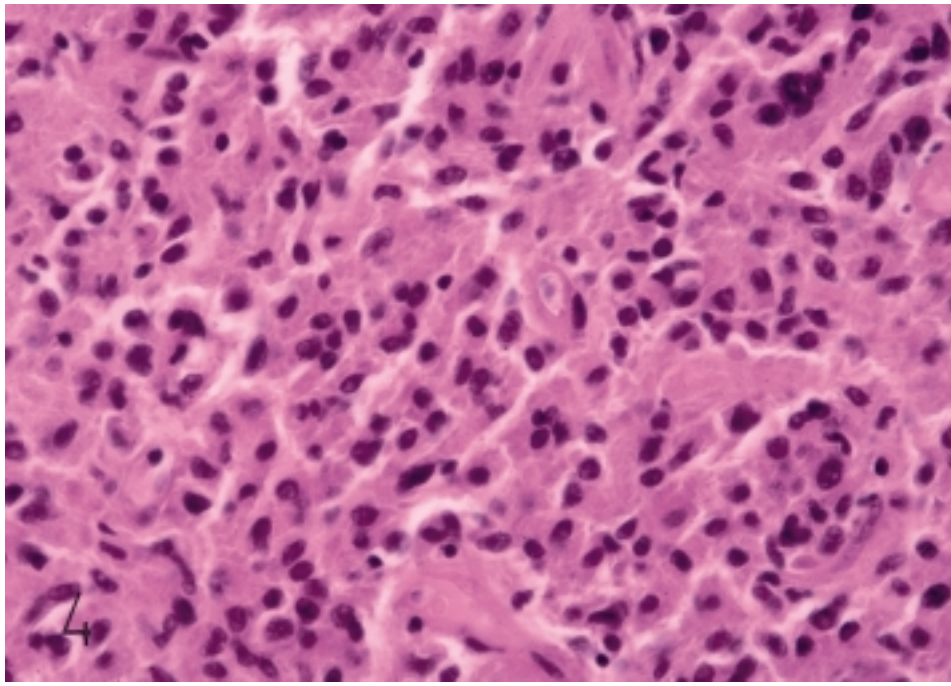

Fig. 4
Detail of Fig. 3. Magnification, x 40

DISCUSSION

The malignant epithelioid stromal tumor is regarded as a rare malignancy that should be distinguished from leiomyomas and leiomyosarcomas as well as from neurinomas. In cases in which histological findings are not clear, the unambiguous evidence is provided by immunohistochemical methods, e.g., detection of CD 34 or CD 117 antigens. The histological findings in this case were convincing enough to make the use of sophisticated immunohistochemical procedures unnecessary. The diagnosis made on the basis of perioperative tissue sample examination was essentially the same as that resulting from postoperative tissue examination. There are reports suggesting that the correct diagnosis of gastrointestinal stromal tumours can be made even from very little material harvested by fine needle aspiration (*6*). However, in tumours with ambiguous histological evidence, the diagnosis of gastrointestinal stromal tumour largely relies on immunohistochemical techniques (*7, 8,9,10*).

Gastrointestinal stromal tumours can also occur in the omental and mesenteric tissues (*11*), the duodenum (*4*) and at other sites of the gastrointestinal tract. As for the prognosis of this tumour development, there are reports suggesting a positive relationship between tumour anatomical site and its biological characteristics. Tumours located to the stomach seem to have a better prognosis than those affecting the small intestine (*3*). Our observations on the course of disease and its convalescence in our patient can support this view. However, the prognosis in our case may be related to the exophytic growth of this tumour.

The genetic aspects of gastrointestinal stromal tumours have so far been poorly understood. The progenitor cells of these tumours (Cajal's interstitial cells) can undergo various mutations that lead to diverse differentiation including the epithelioid type regarded as a rare tumour (*4,12*).

It can be concluded that any protracted dyspepsia requires early attention from both internists and surgeons, regardless of the fact that laboratory and other diagnostic results may be negative or conservative therapy has a good outcome (*13*).

Staffa R., Stupka Z., Krpenský A., Hušek K., Černý E., Jelínková I.:

VZÁCNÝ PŘÍPAD MALIGNÍHO EPITELOIDNÍHO GASTROINTESTINÁLNÍHO TUMORU

S o u h r n

U muže středního věku s déletrvající dyspepsií byl při hospitalizaci na II. chirurgické klinice Fakultní nemocnice u sv. Anny v Brně zjištěn a úspěšně operován objemnější exofytický maligní epiteloidní gastrointestinální stromální tumor žaludku, jehož výskyt, jak potvrzují údaje v zahraniční literatuře, je možno považovat za vzácný. Po resekci dle Billrotha II. je pacient s dvouletým odstupem při kontrolách u nás i na onkologii bez potíží a známek recidivy či generalizace tumoru.

REFERENCES

1. *Duda M, Herman J, Stehlík D*. The role of a surgeon in palliative treatment of tumours. Acta Univ Palacki Olomouc, Fac Med 2000; 143: 80–81.
2. *Herman J, Duda M, Krč I, Pohanka J*. Malignant lymphoma of the stomach and small intestine. Čes Slov Gastroenterol 1999; 53: 111–113.
3. *Emory TS, Sobin LH, Lukes L et al.* Prognosis of gastrointestinal smooth-muscle (stromal) tumors. Dependence on anatomic site. Am J Surg Pathol 1999; 23: 82–87.
4. Goldblum JR, Appelman HD. Stromal tumors of the duodenum. Am J Surg Pathol 1995; 19: 71–80.
5. *Leypold J, Přívara M, Štěpánková J*. Prevence pooperačních trombembolických příhod malými dávkami heparinu [Prevention of postoperative thromboembolism by low heparin doses]. Rozhl Chir 1984; 62: 611–615.
6. *Li QS, O_Leary TJ, Buchner SB et al*. Fine needle sspiration of gastrointestinal stromal tumors. Acta Cytologica 2001; 45: 9–17.
7. *Hurlimann J, Gardiol D*. Gastrointestinal stromal tumours: an immunohistochemical study of 165 cases. Histopathology 1991; 19: 311–320.
8. *Ueyama T, Guo KJ, Hashimoto H et al*. A Clinicopathologic and immunohistochemical study of gastrointestinal stromal tumors. Cancer 1992; 69: 947–955.

9. *Cheuk W, Chung Lee K, Chan J et al.* C-kit immunocytochemical staining in the cytologic diagnosis of metastatic gastrointestinal stromal tumor. A report of two cases. Acta Cytologica 2000; 44: 679–685.
10. *Miettien M, Virolainen M, Maarit-Rikala MS*. Gastrointestinal stromal tumors – value of CD 34 antigen in their identification and separation from true Leiomyomas and Schwanomas. Am J Surg Pathol 1995; 19: 207–216.
11. *Miettien M, Monihan JM, Sarlamo-Rikala M et al*. Gastrointestinal stromal tumors (smooth muscle tumors GISTS) in the omentum and mesentery. Am J Surg Pathol 1999; 23: 1109–1118.
12. *Seidal T, Edvardsson H.* Expression of c-kit (CD117) and Ki 67 provides information about the possible cell of origin and clinical course of gastrointestinal stromal tumours. Histopathology 1999; 34: 416– 424.
13. *Piskač P, Riebel O, Leypold J*. Chirurgická komplikace endoskopických výkonů [Surgical complications of endoscopic procedures]. Čes Slov Gastroenterol 1997; 51: 120–122.