AN UNUSUAL CASE OF LUMBAR PAIN: GANGLIOCYTIC PARAGANGLIOMA

(Received 19 August, 1996)

O. Övünç, M.D.* / E. Övünç, M.D.* / O. Bayramiçli, M.D.****
N. Ülgen, M.D.***** / M. Barlan, M.D.******
Ü. İnce, M.D.** / Ö. Peker, M.D.****

* *Associate Professor, Department of Gastroenterology, Haydarpaşa Numune Hospital, Istanbul, Turkey.*
** *Associate Professor, Department of Pathology, Haydarpaşa Numune Hospital, Istanbul, Turkey.*
*** *Specialist, Department of Neurosurgery, Göztepe Social Security Hospital, Istanbul, Turkey.*
**** *Specialist, Department of Gastroenterology, Haydarpaşa Numune Hospital, Istanbul, Turkey.*
***** *Specialist, Department of General Surgery, Göztepe Social Security Hospital, Istanbul, Turkey.*
****** *Specialist, Department of Radiology, Haydarpaşa Numune Hospital, Istanbul, Turkey.*

ABSTRACT

We report a case of duodenal gangliocytic paraganglioma presenting with an atypical lumbar pain torturing the patient for fifteen years. This middle aged woman received symptomatic treatment during this period without any effect, but a dramatic relieve of pain did follow the excision of the tumor.

Key Words : Lumbar pain, paraganglioma, duodenal tumors.

INTRODUCTION

Paraganglioma is an exceptional tumor composed of neural elements, ganglion cells and endocrine cells. It can also be found very rarely in the duodenum presenting as many clinical pictures like abdominal pain, gastrointestinal hemorrhage. Until now approximately 100 cases of duodenal paragangliomas have been published and we report here a patient with paraganglioma who had atypical back and abdominal pain for fifteen years and her complaints have been attributed to many diverse diseases like lumbar hernia, gastritis, gastric ulcer and even psychiatric disease.

CASE REPORT

A forty-six-year-old housewife presented with 15 years history of back and abdominal pain. Her pain originated from lumbar spinal region and had a belt-like radiation to epigastrium. She was treated with various types of analgetics without any success. In the last three months she began to have attacks of vomiting some 20 minutes after the meals and lost 5 kilogramms of body weight during this period of time. She had also bloating and abdominal distention. Her pain was intensified with the meals and never relieved even after vomiting and belching. Her pain did last 12-18 hours and subsided gradually but did recur almost everyday. Esophagogastroduodenoscopy and abdominal ultrasound was performed and a gastric ulcer, plus duodenitis was identified and the patient was treated with Omeprazol and Sucralphate. The vomiting resolved but the pain continued.

The patient was admitted to emergency room of the local hospital two weeks after the initial examination because of indurable pain. An abdominal CT examination revealed a mass in the second part of the duodenum protruding into the lumen just cranial to Ampulla Vateri. These findings were believed to be compatible with a polyp or enteric duplication cyst and the patient was referred to a neurosurgeon proposing that the pain had a neurological origin. Having found no pathological findings after the neurological examination the patient was referred to the gastroenterology department to clarify the histological nature of the duodenal lesion and non-healing gastric ulcer.

Her past medical history was remarkable only for an appendectomy 20 years ago and subtotal hysterectomy 3 years ago. She had three live births. She neither smoked nor consumed alcoholic drinks, but took several analgesics, nonsteroidal antiinflammatory agents and sedatives regularly. Physical examination was normal except for obesity, anemiae and a blood pressure of 160/90 mmHg.

Initial laboratory studies revealed a mild hypochrom anemiae, but other biochemical tests, urine analysis, radiological exams and ECG were normal. Gastroscopy was performed for the second time. A hiatal hernia was present. In the antral region where in the prior endoscopy a gastric ulcer was described, a superficial ulcer like horseshoe was noted which was surrounded with a hiperemic area. An external polipoid compression on lateral wall of postbulbar duodenum was noted; there was no ulceration on it when examined with a laterally viewing duodenoscope. A cytological material was obtained from this area with a needle. Presumptive diagnosis was submucosal duodenal tumor. Pathological examination showed benign intestinal surface epithelium. The biopsy was repeated with a thin needle under CT guidance; the specimen was defined as a tumor originating from peripheric nerve sheaths or leiomyoma but there was also the concern of reactive fibrosis. Exploratory laparotomy was performed and in the second part of duodenum a capsulated, 3x2 cm measuring, submucouse mass was found. The mass was excised totally after duodenotomy. There was no metastasis in paraaortic and hepatic hilar lymph nodes. Histochemical examination revealed a gangliocytic paraganglioma with abberrant pancreatic tissue in duodenal muscle fibers surrounding the tumor. Immunhistochemical staining with chromogranin-A was strongly positive in epitheloid poligonal cells and the staining with S-100 was strongly positive in fusiform stromal cells. Pancytokeratin, desmin and smooth-muscle-actin were found to be negative. The margins of excised specimen were tumor-free. The postoperative follow-up was eventless and the patient has been completely asymptomatic for three months.

DISCUSSION

Neurogenic tumors of small intestine are rare entities and gangliocytic paraganglioma is an extremely rare benign neurogenic tumor nearly exclusively located in the second portion of the duodenum(1). It has been referred by different names like ganglioneuroma, paraganglioneuroma, paraganglioma, gangliocytic paraganglioma and nonchromaffin paraganglioma, but Kepes and Zacharias(2) coined the term gangliocytic paraganglioma and the term has become widely used since then. Until now approximately 100 cases have been published, most of them corresponding to benign tumors(3,4), but there are also three documented cases showing regional lymph node metastasis(4). Tumor consists of epithelial cells and gangliocytic elements. Immunhistochemically there is strong reactivity for S-100 protein in spindle cells, characteristics of staining with other immunoreactive dyes like desmin, cytokeratin, chromogranin-A is still conroversial(5). Histopathogenetically it is regarded either as a hamartoma or as a true neoplasm(5). The patients present mostly with abdominal pain or gastrointestinal bleeding(6). Gangliocytic paraganglioma may be seen in conjunction with von Recklinghausen's disease(7) or be an element of the Carney triad.(8) These tumors are excised locally and recurrence is uncommon(9). Our case had an atypical presentation with lumbar pain for fifteen years mimicking lumbar hernia, gastric diseases and many other neurologic diseases and she had many divers treatment options for such a long period of time. After the operation the pain resolved promptly which was unresponsive to any analgetic drug before the excision of the tumor.

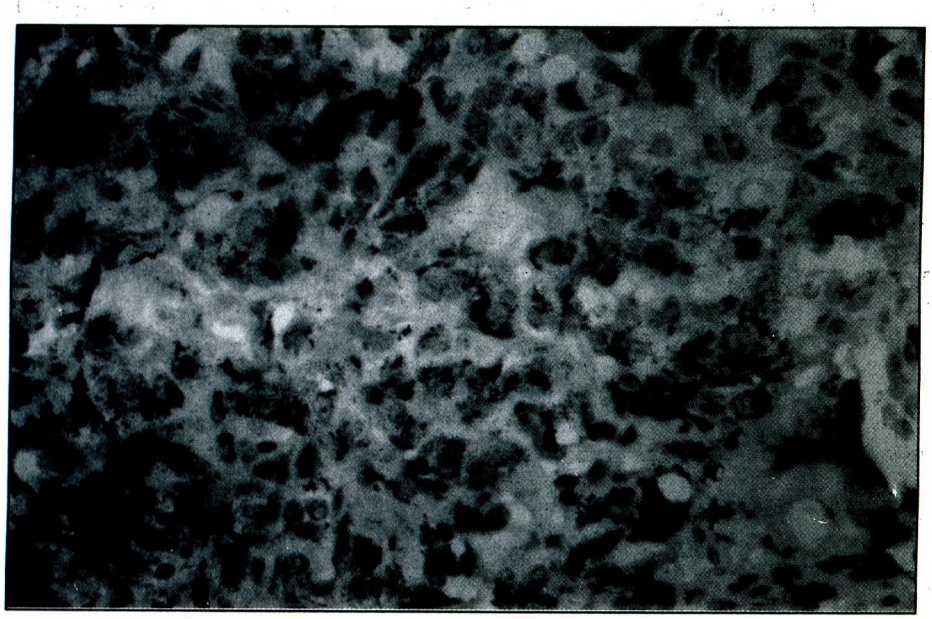

Fig. 1: Immunhistochemical staining with chromogranin-A was strongly positive in epitheloid poligonal cells.

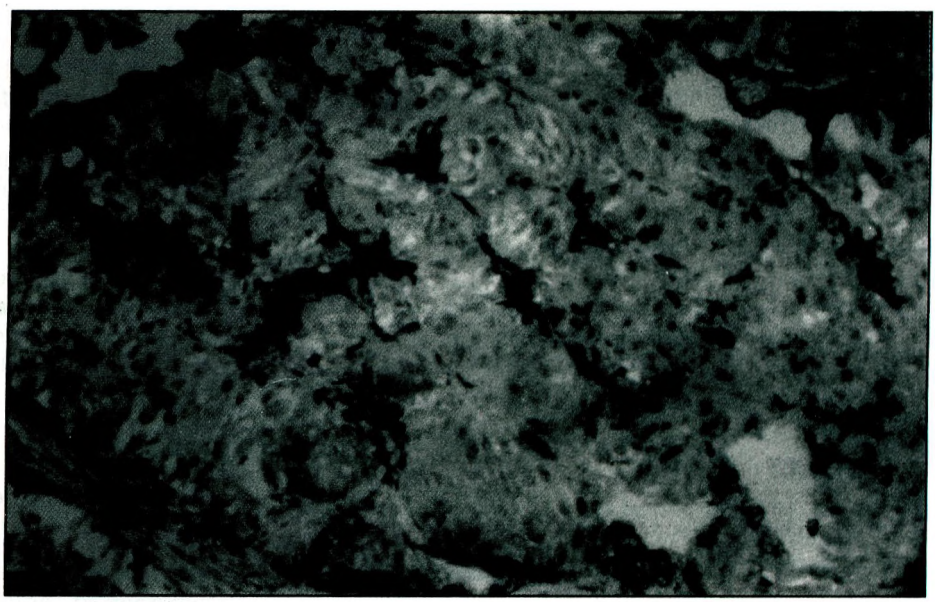

Fig. 2: Immunhistochemical staining with S-100 was strongly positive in fusiform stromal cells.

Therefore we emphasize the importance of bearing in mind that in differential diagnosis of longlasting and unexplained lumbar pain, duodenal neural tumors may be of importance.

REFERENCES

1. *Dennis S. Small bowel neoplasms (other than carcinoid and lymphoma). In: Sleisenger MH, and Fordtran S, eds. Gastrointestinal disease 5th edition. Philadelphia: W.B.Saunders, 1993:1397.*
2. *Kepes JJ, Zacharias DL. Gangliocytic paraganglioma of the duodenum. A report of two cases with light and microscopic examination. Cancer 1971;27:61-70.*
3. *Korbi S, Kapancı Y. Widgren S. Malignant paraganglioma of the duodenum. Immunhistochemical and ultrastructural study of a case. Ann Pathol 1987;7:47-55.*
4. *Hashimoto S, Kawasaki S, Matsuzawa K, Harada H, Makuuchi M. Gangliocytic paraganglioma of the papilla of Vater with regional lymph node metastasis. Am J Gastroenterol 1992;87:1216-1218.*
5. *Sando WC, Mills SE, Rodgers BM. Duodenal gangliocytic paraganglioma occurring in adolescence. J Pediatr Gastroenterol Nutr 1986;5:659-664.*
6. *Barbareshi M, Frigo B, Aldovini D, Leonardi E, Cristina S, Falleni M. Duodenal gangliocytic paraganglioma. Report of a case and review of the literature. Virchows Arch A Pathol Anat Histopathol 1989;416:81-89.*
7. *Stephens M, Williams GT, Jasani B, Williams ED. Synchoronous duodenal neuroendocrine tumors in von Recklinghausen's disease-a case report of coexisting gangliocytic paraganglioma and somatostatin-rich glandular carcinoid. Histopathology 1987;11:1331-1340.*
8. *Perez-Atayde AR, Shamberger RC, Kozakewich HW. Neuroectodermal differentiation of the gastrointestinal tumors in the Carney triad. An ultrastructural and immunhistochemical study. Am J Surg Pathol 1993;17:706-714.*
9. *Scheithauer BW, Nors FE, LeChago J, et al. Duodenal gangliocytic paraganglioma. Clinicopathologic and immunocytochemical study of 11 cases. Am J Clin Pathol 1986;86:559-565.*