Synchronous nonfunctional duodenal carcinoid and high risk gastrointestinal stromal tumour (GIST) of the stomach

Dear Editor,

Simultaneous finding of gastrointestinal stromal tumour (GIST) and primary tumour arising in another site is an always more frequent event in the clinical practice. A 76-year-old man came to our Hospital for abdominal pain. Esophagogastroduodenoscopy was negative. Contrast-enhanced computed tomography (CT) showed an inhomogeneous mass (7.0 cm in maximum diameter) along the gastric lesser curve (arrows), extending into the perigastric adipose tissue (Figure 1). A subtotal gastrectomy showed a 8x5x5 cm. serosal mass with intact mucosa. On sectioning the tumour was composed by soft, tan tissue with foci of haemorrhage and necrosis. Histologically, the neoplasm showed fascicular proliferative pattern of spindle cells with diffuse perinuclear vacuolization. The mitotic activity was 12 mitoses/50 hpf. The spindle cells were positive for c-KIT (CD117), CD34 and DOG 1 (Figure 2A). Infiltration of the muscularis propria and of the lesser omentum was found. The diagnosis of high malignant potential GIST was performed. The duodenal resection margin showed a small lesion of 5 mm. within submucosa with an intact mucosa. The lesion was composed by round-ed nests of closely packed tumour cells, showing positive stainings for Grimelius Silver, chromogranin, neuron – specific enolase and anti-serotonin antibodies (Figure 2B). The diagnosis of non angioinvasive serotonin-producing carcinoid was made. After a 12 months follow-up the patient is free of disease.

The present case of synchronous duodenal carcinoid and high risk gastric GIST is the first description in the English literature. Agaimy et al[1] conducted a review of the literature, regarding cases with simultaneous sporadic GISTs and other malignancies and reported an overall frequency of second tumours in different series varying from 4.5% to 33% (mean, 13%). In the same study they examined 518 cancers in 486 GIST patients, excluding Neurofibromatosis 1 and Carney triad associated tumours: a total of 29 patients had multiple malignancies. Gastric GISTs were most commonly involved with other neoplasms, reflecting their overall high frequency (60%) of all GISTs. In Agaimy et al’ study were found 8 carcinoid tumours: 4 gastric, 2 of the ileum, 1 of the small intestine and 1 of the colon. Ferreira et al[2] examined 43 GISTs and found 6 cases with other primary tumours. In 3 patients the GIST was located in the stomach. Liszka et al[3] found other malignancies in 22 of the 82 (26,8%) patients with GIST. Goncalves et al[4] discovered a 13.8% incidence of non GIST tumours in a series of 101 GISTs cases. No neuroendocrine tumour was found in all the three reported studies.

Only 3 cases of concomitant gastric carcinoid and GIST have been reported in the literature[5-7]. Non-functioning duodenal carcinoid usually consists of serotonin-producing enterochromaffin cells or calcitonin-producing cells. They do not behave aggressively unless they extend beyond the submucosa. Duodenal carcinoid may coexist with different types of cancer, either synchronously or metachronously. Burke et al[8] studied 99 patients with duodenal carcinoids and found 28 patients with another malignancy. No GIST was found coexisting with duodenal carcinoid. Simultaneous GISTs and other tumours
are frequently incidentally discovered. An accurate clinical examination should be performed in patient with gastric GIST. We read with great interest the paper of Cirillo who reported the association of gastric carcinoid and low risk GIST and emphasized that this finding is very unusual. In this paper the Author described the association of GIST with other neoplasms. Our case is the first description of synchronous nonfunctional duodenal carcinoid and high risk GIST of the stomach and supported the consideration that in the management of GIST other asymptomatic neoplasms in other sites should be accurately investigated.

Figure 2. A, The gastric tumour showed fascicular proliferative pattern of spindle cells with diffuse perinuclear vacuolization (H&E 100 x). Insert: the neoplastic cells were diffusely positive for c-KIT (CD117) (200 x). B, The duodenal tumour was composed by rounded nests of closely packed tumour cells (H&E 100 x). Insert: the tumor cell showed positivity for anti-serotonin antibodies (200 x).
References


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