Carcinoid Tumour of Caecum: an Unusual Palpable Mass in the Right Iliac Fossa

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Abstract Carcinoid tumour represents 0.8–1.5 % of malignant digestive tumours. Gastrointestinal (GI) carcinoids account for 95 % of all the carcinoids, and caecal carcinoids account for 5 % of all the carcinoids. These tumours are frequent in women (2–4:1) as reported by Spallitta and Termine (Minerva Chir 55:77–87, 2002). In order of frequency, they may occur in the appendix (35 %), ileum (28 %), rectum (13 %) and bronchi. Incidence is less than 1 % in the pancreas, gall bladder, liver, larynx, testes and ovaries. The colon and ileocaecal region are rare sites of origin for gastrointestinal carcinoids as reported by Soga (J Exp Clin Cancer Res 17:139–48, 1998). They remain asymptomatic for years and many a times are diagnosed endoscopically, intraoperatively or during autopsy, based on their histopathological findings. We present a rare case of caecal carcinoid.

Keywords Caecum · Carcinoid · Tumour · GIST

Introduction

The carcinoid tumour arises from the enterochromaffin cells of Kulchitsky at the base of the crypts of Leiberkuhn. Carcinoid tumours are non-pancreatic digestive neuroendocrine tumours, generally well differentiated. Most of them are localized in the ileum, rectum, stomach and appendix. Only the bronchi are frequent extra-digestive localization. Most of these tumours are

asymptomatic and discovered incidentally during a laparotomy or secondary to the exploration of a carcinoid syndrome. CT, ultrasonography and angiography play a primary role in the diagnosis of this tumour. Radical surgery is the only treatment in carcinoids to prevent metastasis risk. Despite the fact that they are usually silent and diagnosed with advantages, carcinoids have good prognosis and offer long survival after effective treatment.

Case Report

A 67-year-old male presented with a lump in the abdomen in the right iliac fossa, with dull aching, continuous pain and loose watery stools since 45 days. There was no history of previous such episodes, fever, melaena, tuberculosis, vomiting, fatigue, dizziness or palpitations. General examination was unremarkable while per abdominal examination revealed a tender lump of size 4 × 4 cm in the right iliac fossa which was firm in consistency and freely mobile. Ultrasonography showed a complex bowel mass of size 5 × 4 × 3 cm, round to oval in shape in the right iliac fossa with collapsed central lumen and poor vascularity. Colonoscopy revealed fullness with ulcerative growth at the ileocaecal junction suggestive of neoplastic etiology. A contrast-enhanced CT scan of the abdomen and pelvis showed circumferential wall thickening of the caecum causing luminal narrowing with few subcentimetre-sized lymph nodes in the right iliac fossa.

Exploratory laparotomy revealed a mass in the caecal wall without serosal involvement and pericecal and mesenteric lymph node enlargement with normal appendix. Radical right hemicolectomy with side to side ileo-transverse colon anastomosis was performed. Postoperative period was uneventful.

The resected specimen included a 5-cm-diameter tumour, which had narrowed the lumen by 80 % and infiltrated the ileocecal valve while the rest of the bowel was normal (as shown in Fig. 1). Histopathology revealed a well differentiated carcinoid

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Fig. 1 Gross specimen showing cut section of the caecum with 80 % luminal narrowing with CT scan and colonoscopic view of the tumour (shown by the arrows)

tumour of caecum infiltrating up to the muscularis propria (as shown in Fig. 2). Serosa was free of tumour infiltration and 3 out of 12 lymph nodes were involved.

Discussion

The carcinoid tumours arise from neuroendocrine cells, which are widespread in the human body, especially in the organs derived from the primitive intestine. These tumours usually secrete excessive amounts of hormone serotonin. These tumours originate from any cell of amine precursor uptake and decarboxylation (APUD) system and therefore produce several intestinal hormones. These tumours are frequent in women (2–4:1) [1]. In order of frequency, they may occur in the appendix (35 %), ileum (28 %), rectum (13 %) and bronchi. Incidence is less than 1 % in the pancreas, gall bladder, liver, larynx, testes and ovaries. The colon and ileocaecal region are rare sites of origin for gastrointestinal carcinoids [2].

These vascularized tumours follow the so called ‘rule of one third’ which states that one third of these tumours are multiple, one third of those in the GI tract are in the small bowel, one third metastasize and one third have second malignancy.

Fig. 2 Histopathology picture showing a well differentiated carcinoid tumour of the caecum

Approximately 1 in 200 appendectomies results in the discovery of carcinoid [3, 4].

Carcinoid tumour presents considerable problems of diagnosis because symptoms are aspecific. Diagnosis is possible in patients with high urinary levels of 5-HIAA, in the presence of carcinoid syndrome and by endoscopic biopsy when tumour infiltrates the gastrointestinal mucosa. Carcinoid syndrome is the syndrome complex due to hormones secreted from metastasis from carcinoid tumours [5].

Determinant risk factors are primary size, localization and serosal penetration. In patients with any of these risk factors, resection with regional lymphadenectomy is recommended. Neither adjuvant chemotherapy nor radiotherapy may play a significant role in this neoplasm.

Incidence of metastasis depends upon tumour size. Tumour less than 1 cm has 15–25 % incidence of metastasis, 1–2 cm has 60–80 % incidence of metastasis and tumour >2 cm has more than 70 % incidence of metastasis [6].

Chemotherapeutic agents such as adriamycin, streptozotocin, 5-fluorouracil and cisplatin can be used. Symptomatic treatment is also given for diarrhoea, hypertension and bronchial constriction with interferon alpha and octreotide.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no competing interests.

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