CASE REPORT

Solitary facial metastasis of an ileal carcinoid

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Abstract. We report an unusual case of solitary facial metastasis as first clinical manifestation of an ileal carcinoid. Only one similar case has been reported in the literature. The patient, a 79 years old man, was referred for the excision of a right facial subcutaneous nodule. Pathology showed a soft tissue metastasis of mid-gut carcinoid. After a complete radiological investigation, a small carcinoid of the terminal ileum was found and the patient underwent a right emicolectomy. Pathology confirmed a typical EC cells carcinoid identical to that of the facial tumor. At 2 years of follow-up no clinical or radiological signs of others metastases were found.

Key words: carcinoid tumor, solitary metastasis

Introduction

Mid-gut carcinoid tumors usually metastatize to liver, lungs, and bones. Unusual metastatic sites like mammary gland, spleen, orbital tissue and heart are described as first manifestation of an occult ileal tumor (1-5). We present a case of solitary facial metastasis of an ileal carcinoid tumor in an asymptomatic patient. Cervical and facial soft tissue metastases of ileal carcinoids are extremely rare, as confirmed by the single reference in literature(6).

Case report

A 79 years old man was referred for evaluation of a right facial subcutaneous nodule. He experienced recurrent intermittent abdominal pain. A barium meal performed 5 years before had been suggestive for Crohn's disease, but the patients was not medically treated.

Neck ultrasound and ORL examination were negative. Serum calcitonin was normal. A CT scan showed a 20 mm subcutaneous mass with firm adhesion to the right masseter muscle (Fig. 1). Complete excision of the mass was performed. Pathology showed a soft tissue metastasis of a carcinoid with a type A histologic pattern (7) (Fig. 2) and intense serotonin immunostaining and argentaffin reaction (Fig. 3), suggesting the ileal neuroendocrine origin of the primary tumor. Further investigations in the attempt to locate the primary neoplasm were planned. No

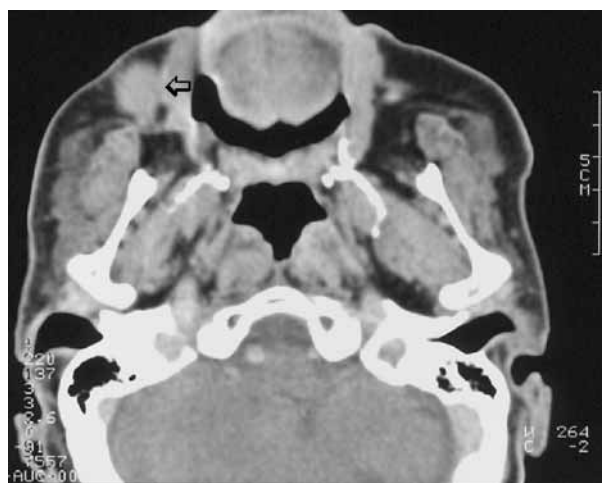

Figure 1. CT scan: Subcutaneous facial metastasis (18x23 mm) with regular margins and firm adhesion to the right masseter muscle.

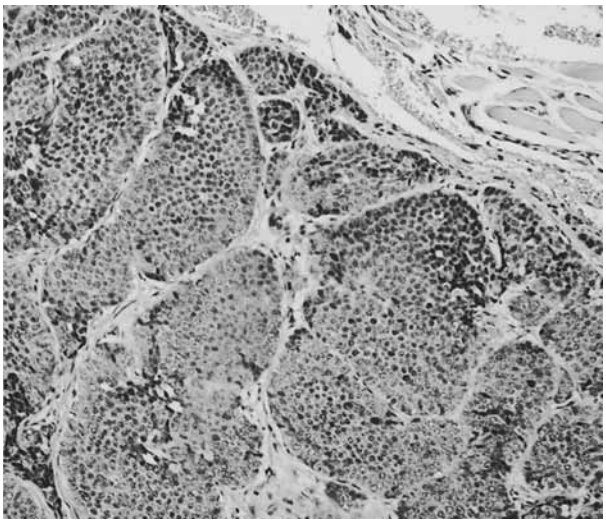

Figure 2. Histology of the facial tumor with peritumoral skeletal muscle fibres(upper right corner).

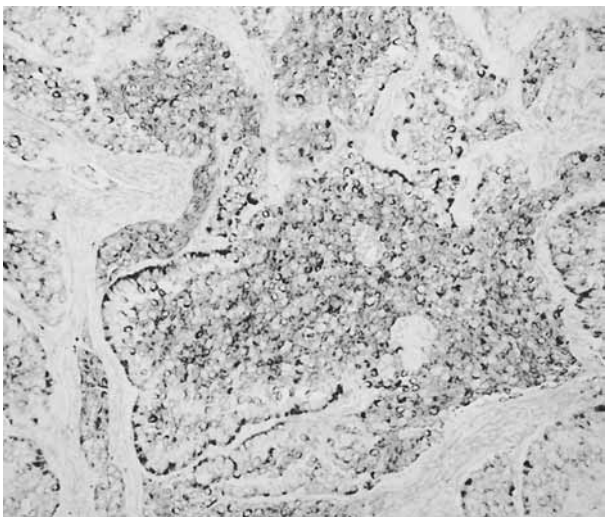

Figure 3. Argentaffin reaction of tumor cells typical of ileal carcinoid.

symptoms of carcinoid syndrome were present. The urinary excretion of 5-hydroxyndol acetic acid (5HIAA) was normal. A thoraco-abdominal scan showed a 20x15 mm exophitic mass close to the ileocecal valve with mesenteric retraction and an abdominal aortic aneurysm (Fig. 4). A barium meal confirmed the presence of a small lesion of the terminal ileum with evident deformation of the cecum profile secondary to a diffuse mesenteric invasion (Fig. 5). A somatostatin receptor scintigraphy was negative for metastases at

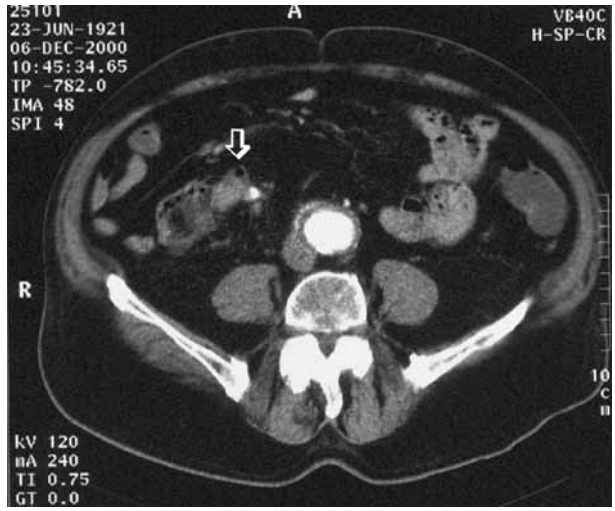

Figure 4. CT scan: Exophitic mass (20x15mm) of the last ileal loop and cecum with mesenteric retraction. Abdominal aortic aneurysm.

other locations. At laparotomy we found the mesentery of the last ileal loop massively retracted and fibrotized, firmly connected to, but not infiltrating, the third portion of duodenum. Intraoperative ultrasounds were negative for liver metastases. The patient underwent a right emicolectomy with resection of the terminal ileum and recovered uneventfully. Gross examination showed a 0.8 cm soft, pink polypoid lesion, located at 7 cm from the ileal valve. Moreover a 2x1 cm, partially calcified tumor mass infiltrated the underlying mesentery. Histologically a typical EC cells

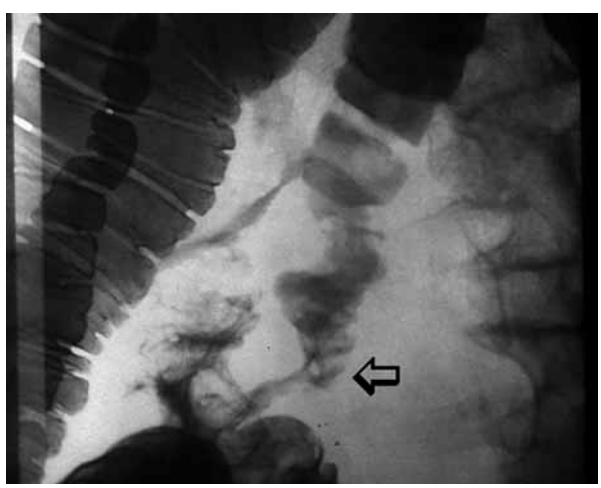

Figure 5. Small bowel enema: Irregular profile of the last ileal loop and cecum with mesenteric retraction.

carcinoid tumor was found in both lesions with serotonin and argentaffin staining as in facial tumor (Fig. 6).

At 2 years of follow-up no clinical or radiological signs of other metastases have been found.

Discussion

Metastases of carcinoid tumors of the small intestine usually involve liver, lungs and bones. Other sites like mammary gland, spleen, orbital tissue and heart are described as uncommon, but in a few cases represent the first manifestation of an occult tumor (1-5). This is the first case reported in literature of facial soft tissue metastasis as first disclosure of an ileal carcinoid. Only a similar case were reported by Naschitz in 1992, but the soft tissue metastasis was located at the distal insertion of the sternocleidomastoid muscle and the primary tumor was discovered only four years later after the onset of a carcinoid syndrome with liver metastases (6). In our case the primary tumor was only 0.8 cm in diameter but it was diagnosed thanks to the mesenteric retraction, visualized by the CT scan, due to tumor infiltration. The locally advanced disease and the valuation *ex post* of the early barium enema were probably suggestive of the tumor occurrence five years before, when a Crohn's disease was suspected. The carcinoid tumor was composed of argentaffin EC cells which are the predominant cell type in jejuno-ileal carcinoids. Late recognition of carcinoids of this intestinal region is not uncommon, resulting in an advanced stage at detection with high incidence of distal metastases. Over 70% of the entire jejuno-ileal carcinoids are small lesions (20 mm or less), difficult to detect with standard radiological investigations (8). The fact that the tumor was well differentiated can explain its slow growth rate. Soft tissue metastasis of ileal carcinoid is remarkably rare expecially without the previous metastatic involvement of the liver. Therefore this case highlights an irregular pattern of cancerous colonization that may represent the first manifestation of an occult small ileal carcinoid tumor.

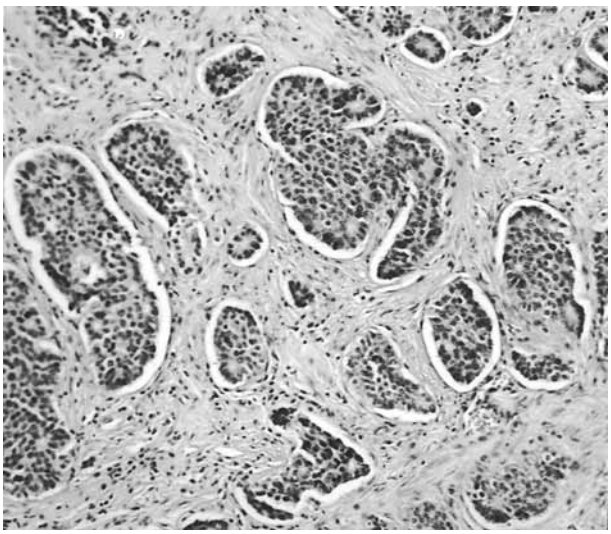

Figure 6. Histology of the ileal tumor with pattern of classic carcinoid.

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