Case report

Medical Ultrasonography
2010, Vol. 12, no. 3, 245-248

Ileal neuroendocrine tumor – ultrasonographic and capsule endoscopy appearance: a case report

Simona Grad[1], Dan Dumitraşcu[1], Radu Badea[2], Silviu Iobagiu[2], Oliviu Pascu[2]

[1] 2nd Medical Clinic, University of Medicine and Pharmacy Iuliu Haţieganu, Cluj Napoca, Romania
[2] 3rd Medical Clinic, University of Medicine and Pharmacy Iuliu Haţieganu, Cluj Napoca, Romania

Abstract

We present the case of a female patient admitted for an abdominal tumoral mass corresponding to large liver metastases. The abdominal ultrasonography detected a solid intestinal tumor. The capsule endoscopy showed a vegetative tumor on the terminal ileum. After intestinal and partial liver resection, the histologic examination identified a neuroendocrine tumor. The patient responded incompletely to standard chemotherapy. In the presence of large hypoechoic liver metastases, a neuroendocrine tumor should be suspected.

Key words: neuroendocrine tumor, ultrasonography, capsule endoscopy

Rezumat

Prezentăm cazul unei paciente internată în clinica noastră, pentru astenie, diaree, dureri abdominale, scădere ponderală. La examenul obiectiv al abdomenului s-a evidenţiat o masă tumorală în epigastru. Ecografia abdominală a decelat prezenţa metastazelor hepatice multiple, precum şi o formaţiune la nivelul intestinului subţire. Examinarea cu videocapsulă endoscopică a evidenţiat o tumoră vegetativă, stenozantă la nivelul ileonului terminal. După ce s-a practicat hemicolectomie dreaptă şi rezecţie parţială hepatică, examenul histopatologic a relevat aspect de tumoră neuroendocrină. Pacienta a răspuns parţial la chimioterapia efectuată postoperator.

În cazul prezenţei metastazelor hepatice hipoecoice, de dimensiuni mari, trebuie întotdeauna luat în considerare şi diagnosticul de tumoră neuroendocrină.

Cuvinte cheie: tumoră neuroendocrină, ecografie abdominală, videocapsulă endoscopică

Introduction

Neuroendocrine tumours arise from the diffuse neuroendocrine system and secrete several peptides and bioactive amines (serotonin, histamine, dopamine, norepinephrine, corticotropin, calcitonin, bradykinin, kalikrein, gastrin, cholecystokinin, prostaglandins) [1]. The most common occurrence site of neuroendocrine tumors is the ileum [2].

Received 27.04.2010 Accepted 30.06.2010
Med Ultrason
2010, Vol. 12, No 3, 245-248
Address for correspondence: Simona Grad
University of Medicine and Pharmacy
Iuliu Haţieganu
Second Medical Department
Str. Clinicilor , No 2-4
Cluj Napoca , Romania
costinsimona_m@yahoo.com

The symptoms of small bowel carcinoids are represented by intermittent intestinal obstruction and carcinoid syndrome. Presence of the carcinoid syndrome usually indicates hepatic or retroperitoneal metastases. The typical carcinoid syndrome is characterized by flushing, diarrhea, nonspecific abdominal pain and bronchospasm [1].

The diagnosis of small bowel tumors is often difficult due to their rarity, nonspecific and variable nature in signs and symptoms. The most useful diagnostic test for the carcinoid syndrome is the urinary excretion of 5-hydroxyindolacetic acid (5 HIAA) per 24-hour. 5 HIAA is the end product of serotonin metabolism [3]. Capsule endoscopy is a more recent diagnostic tool [4].

Surgery is the radical form of curative therapy for carcinoid tumors. Numerous therapies are available for palliation including surgery, pharmacologic therapy, interventional radiologic therapy, embolization and chemoembolization of hepatic metastases, immunotherapy (Interferon alfa) and chemotherapy [5,6].

Case presentation

A 41-year-old female patient was admitted for progressive upper abdominal pain, diarrhea (3-4 stools per day), weight loss (10 kg in 6 month), anorexia, asthenia, many episodes of dyspnoea associated with facial flush (lasting approximately 30 minutes), symptoms lasting for 1 year. The physical examination revealed facial flush during the interview, diffuse abdominal sensitivity at palpation, hepatomegaly of a firm consistence, irregular surface and a hard 6-cm palpable mass in the epigastrium.

Abdominal CT scan detected multiple nodules in the liver (segments V-VIII; VI-VII), some of them of pseudocystic pattern, other with low density, suggesting liver metastases.

Transparietal abdominal ultrasonography (US) confirmed the presence of multiple and large liver metastasis with various aspects- cystic, mixt hypoechoic with predominant liquid component, mixt hyperechoic with predominant solid component (fig 1). The largest metastasis (10 cm) corresponded with the palpable epigastric tumoral mass. In the right iliac fossa, a hypoechoic tumor in the small bowel wall, moderate vascularized was detected (fig 2). Regional adenopathy was also observed.

Colonoscopy and gastroscopy, performed in order to identify the primary tumor, were with no abnormal finding.

The videocapsule endoscopy detected a stenosing vegetative tumor of the terminal ileum (fig 3).

Urinary excretion of 5-hydroxyindolacetic acid (5 HIAA) could not be determined for technical reasons.

The patient was referred to surgery. A stenosing 3/3 cm tumor was found in the terminal ileum. Lymph nodes (smaller than 1 cm) were indentified along right colonic artery, ileum, and caecum. Right hemicolectomy, with ileo-transversal termino-lateral anastomosis was performed together with partial hepatic resection (segments V –VI of the liver) and anterograde cholecystectomy.

Histopathologic examination revealed the aspect of a malignant intestinal neuroendocrine tumor, with hepatic and lymphatic metastases – T3N1M1 (fig 4).

Postsurgical recovery and evolution was favorable and the patient continued with chemotherapy (5-Fluorouracil and somatostatine).

At one-month follow-up, the patient had an improved clinical condition and increased in weight. The capsule endoscopy was repeated and no intraluminal lesion was detected. Abdominal ultrasonography showed no modification of remaining hepatic metastases. The patient continued chemotherapy. Six months after the diagnosis, the health status was poorer, the patient presented weight loss and facial flush. The size of liver metastasis increased. A decision was taken on the continuation of chemotherapy.

Discussion

We presented a rare type of small intestine tumor diagnosed by transparietal abdominal ultrasonography and confirmed by videocapsule endoscopy.

Neuroendocrine tumors have a low prevalence and are localized in 3/4 cases in the gastrointestinal tract [7,8,9].

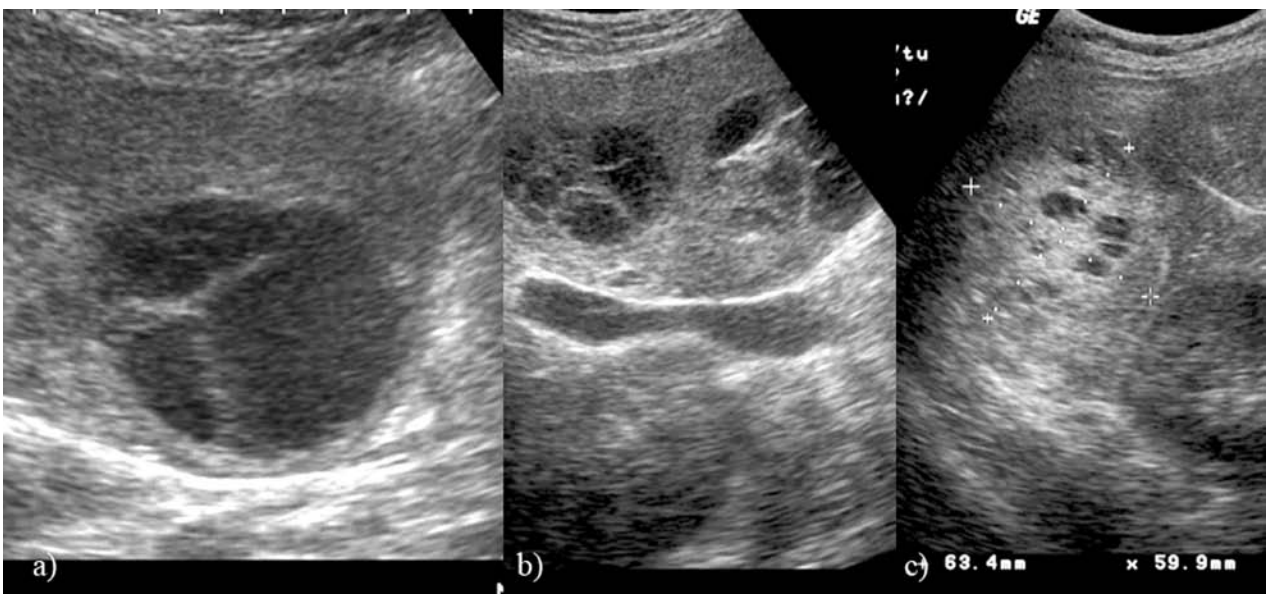


Fig 1. Liver metastases: a) cystic b) mixed hypoechoic with predominant liquid component; c) mixed hyperechoic with predominant solid component

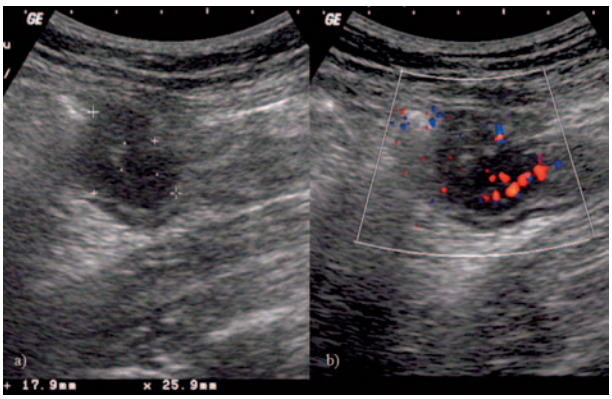


Fig 2. Small bowel tumor: a) gray scale US; b) color Doppler US

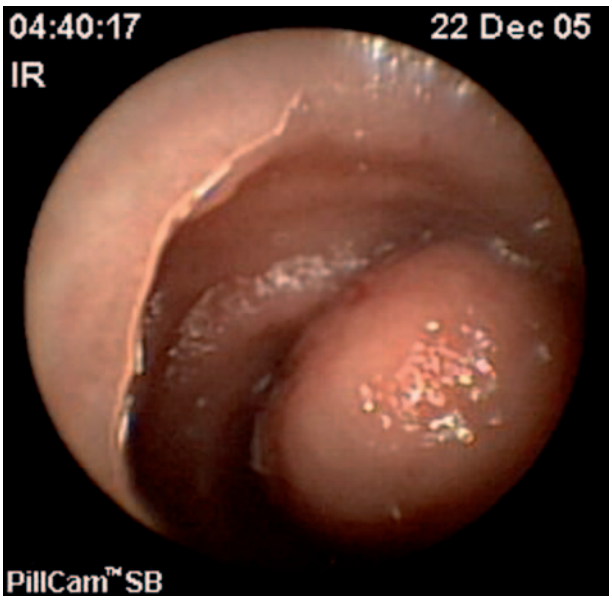


Fig 3. Capsule endoscopy – stenosing vegetative tumor of the terminal ileum

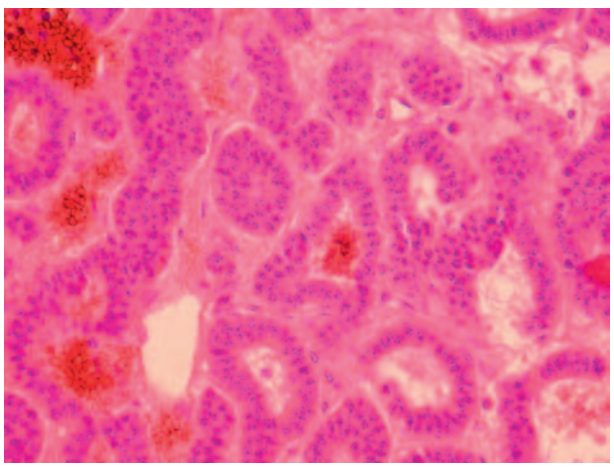


Fig 4. Microscopic pathology – tumor islands are formed by atypical tumor cells, with ovoid or round nucleus and finely granular chromatin

The carcinoid syndrome is present in approximately 5 to 7% of these patients. Presence of the carcinoid syndrome usually indicates hepatic or retroperitoneal metastases and is an indicator of unfavorable outcome [10]. This association was encountered also in our case.

The prevalence of distant metastases increases with the size of the primary tumor. The rate of metastases from tumors smaller than 1 cm is 2%, between 1 to 2 cm – 50%, and from tumors over 2 cm – 80% [11]. The most common symptoms of the carcinoid syndrome are flushing, diarrhea, nonspecific abdominal pain, bronchospasm, pellagra-like skin reactions, and progressive right-sided congestive heart failure due to valvular cardiac disease [11,12]. Therefore the surgical therapy should be addressed not only to the intestinal tumor but also to the liver metastases [13,14].

The diagnosis of small bowel tumors is often difficult due to their rarity and nonspecific and variable nature of the presenting signs and symptoms. Physicians rarely suspect this diagnosis and it arrives frequently as a surprise. Thus, delay in diagnosis is common, which may result in the discovery of disease at a late stage and a poor treatment outcome [15].

Liver metastases are easy to find using abdominal CT or US, but the challenge is to find out the primary tumor which sometimes can have 3 or less cm in diameter [16]. US does not have usually a high accuracy in the diagnosis of intestinal tumors, unless they are not very large. But an experienced investigator can overcome this limit of the methodology , even in cases when the CT scan has a negative result. US can detect a hypoechoic tumoral mass on the intestinal parietal wall and assess the blood supply with the Doppler mode.

A good control of the US diagnostic accuracy is increasingly represented by capsule endoscopy, which allows the diagnosis of tumors from the intestinal lumen, as it was in our case. The diagnostic yield of this method for intestinal tumors is high [17,18]. In a large series it proved to be superior to enteroclysis [19]. A proximal localization of the neuroendocrine tumors, i.e. in the duodenum, can be better diagnosed by endoscopic ultrasonography

Suggestive for small bowel tumors at US examination are identification of a hypoechoic mass with intraluminal presentation, attached to the wall by a large base, and the thickening of the wall mainly due to the muscular layer [20]. Signs of ileus can sometimes be found in cases of obstructive tumors. Doppler shows the vascularization of the tumor. When comparing US with CT in a Chinese series of jejunoileal tumours, CT found the tumor in 32% cases and US in only 22% cases [21]. US has a better performance in the diagnosis of ileal intususception, an

acute condition where the intestinal wall is also altered [22].

We presented this case due to the rarity of malignant intestinal neuroendocrine tumors. The contribution of US for identifying the primary lesion and the confirmation of US finding by videocapsule endoscopy is important. Another particularity of the case is the varied aspect of the liver metastasis, especially the pseudocystic type.

References

1. Feldman M, Friedman LS, Brandt LJ. Gastrointestinal carcinoid tumors and the carcinoid syndrome, Sleisenger & Fordtran's Gastrointestinal and Liver Disease, 8th ed, 2006; 1191–1220.
2. Kehagias D, Moulopoulos L, Smirniotis V, Pafiti A, Ispanopoulos S, Vlahos L. Imaging findings in primary carcinoid tumour of the liver with gastrin production. Br J Radiol 1999; 72:207–209.
3. Onaitis MW, Kirshbom PM, Hayward TZ, et al. Gastrointestinal carcinoids: characterization by site of origin and hormone production. Ann Surg 2000; 232:549–556.
4. D'Halluin PN, Delvaux M, Lapalus MG, et al. Does the «Suspected Blood Indicator» improve the detection of bleeding lesions by capsule endoscopy? Gastrointest Endosc 2005; 61:243–249.
5. Soga J. Early-stage carcinoids of the gastrointestinal tract. Cancer 2005; 103: 1587-1595.
6. Arnold R. Medical treatment of metastasizing carcinoid tumors. World J Surg 1996; 20:203–207.
7. Kloppel G, Anlauf M. Epidemiology, tumour biology and histopathological classification of neuroendocrine tumours of the gastrointestinal tract. Best Pract Res Clin Gastroenterol 2005; 19:507–517.
8. Caplin ME, Buscombe JR, Hilson AJ, Jones AL, Watkinson AF, Burroughs AK. Carcinoid tumour. Lancet 1998; 352:799–805.
9. Burke AP, Thomas RM, Elsayed AM, Sobin LH. Carcinoids of the jejunum and ileum: an immunohistochemical and clinicopathologic study of 167 cases. Cancer 1997; 79:1086–1093.
10. Moertel CG, Sauer WG, Dockerty MB, Baggenstoss AH. Life history of the carcinoid tumor of the small intestine. Cancer 1961; 14:901–912.
11. Gonzalez Cao M, Bastarrika G, Garcia-Foncillas J, Aristu J, Chopitea A, Martin Algarra S. Ileal carcinoid tumor with liver metastases and cardiac involvement treated with intraarterial liposomal doxorubicin and valve replacement. Clin Transl Oncol 2006; 8:369–371.
12. Stella M, Decian F, Mithieux F, Meeus P, Rivoire M. Ileal carcinoid with liver metastasis presenting after ten years with abdominal mass and right heart failure: report of a case. Tumori 2006; 92:83–85.
13. Sarmiento JM, Que FG. Hepatic surgery for metastases from neuroendocrine tumors. Surg Oncol Clin N Am 2003; 12: 231-242.
14. Kerstrom G, Hellman P, Hessman O. Midgut carcinoid tumours: surgical treatment and prognosis. Best Pract Res Clin Gastroenterol 2005; 19:717–28.
15. Costin S, Dumitrascu D.L. The neuroendocrine tumors of the ileum. CEJMed 2008; 3:135–140.
16. Pais R, Dadoun F, Zota MB, et al. Diagnostic and therapeutic approach to small bowel carcinoid tumor. Acta Endocrinol 2008; 4:447–453.
17. Cobrin GM, Pittman RH, Lewis BS. Increased diagnostic yield of small bowel tumors with capsule endoscopy. Cancer 2006; 107:22–27.
18. Schwartz GD, Barkin JS. Small bowel tumors. Gastrointest Endosc Clin N Am 2006; 16:267–275.
19. Marmo R, Rotondano G, Piscopo R, et al. Capsule endoscopy versus enteroclysis in the detection of small-bowel involvement in Crohn's disease: a prospective trial. Clin Gastroenterol Hepatol 2005; 3:772–776.
20. Rioux M, Langis P, Naud F. Sonographic appearance of primary small bowel carcinoid tumour. Abdom Imaging 1995; 20:37–43.
21. Yang YS, Huang QY, Wang WF, Sun G, Peng LH. Primary jejunoileal neoplasmas: a review of 60 cases.World J Gastroenterol 2003; 9:862–864.
22. Sofia S, Casali A, Bolondi L. Sonographic diagnosis of adult intussusception. Abdom imaging 2001; 26:483–486.