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Carcinoid tumor of the small intestine: A case report

İnce bağırsakta karsinoid tümör: Olgu sunumu

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Abstract
Carcinoid tumors are the second most common tumors of the small intestine after adenocarcinoma. These tumors are discovered either incidentally in the event of investigating non-specific digestive disorders notably in patients with Konig's syndrome. Diagnosis is confirmed through histopathology complemented with immunohistochemistry. Surgery is the only curative treatment and it is indicated upon decision by a multidisciplinary treatment team. We report a case of a 65-year-old woman with carcinoid tumor of the ileum.
Keywords: Carcinoid tumor, Ileum, Retractile mesenteritis, Surgery

Öz
Karsinoid tümörler adenokarsinom sonrası ince bağırsağın ikinci en yaygın tümörleridir. Bu tümörler, özellikle Konig sendromlu hastalarda spesifik olmayan sindirim bozukluklarını araştırırken tesadüfen keşfedilmiştir. Tanı immünohistokimya ile tamamlanan histopatoloji ile doğrulanır. Cerrahi tek küratif tedavidir ve çok disiplinli bir tedavi ekibinin kararıyla belirtilir. Biz ileumun karsinoid tümörü olan 65 yaşında bir kadın hastayı sunuyoruz.
Anahtar kelimeler: Karsinoid tümör, Ileum, Retraktil mezenterit, Cerrahi

Introduction

Carcinoid tumors are well differentiated neuroendocrine tumors. They are mostly derived from digestive tract, of non-pancreatic origin, developed at the expense of the enterochromaffin cells of the digestive tract. They are located in the gastrointestinal tract in 67% of cases and bronchial tubes in 25% of cases. They are the second most common tumor in the small intestine after adenocarcinoma. We report a case of a 65-year-old woman with carcinoid tumor of the ileum.

Case presentation

A 65 year old female patient with a history of cholecystectomy performed a year ago was admitted for periumbilical pain and hematochezia associated with konig's syndrome. Clinical examination found a patient with a satisfactory general condition, sensitivity in the right iliac fossa and a hard, painful and mobile mass (35mm large). Proctologic exams revealed an anterior large anal fissure which appeared recent in nature. Digital rectal exam was normal. Ileo-colonoscopy was normal. Abdominal computed tomography (CT) detected the presence of an endoluminal ileal mass in the right iliac fossa. This mass enhanced heterogeneously after contrast injection. It measured 30 x 26mm in diameter associated with multiple mesenteric lymph nodes (the largest of which was 18mm) containing calcifications, calcifications with retractile Mesenteritis (Figure 1).

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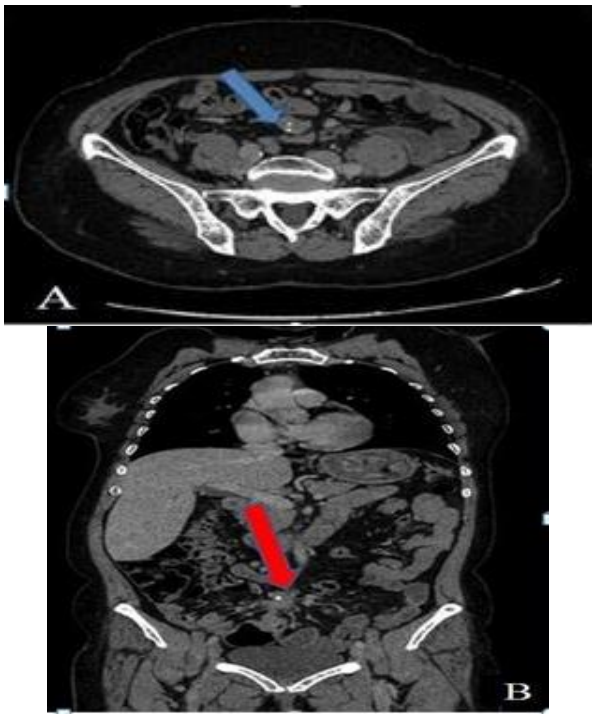

Figure 1: A: Axial abdominal computed tomography showing endoluminal ileal mass in the right iliac fossa, B: Axial image showing retractile mesenteritis

The patient's case was discussed by our multidisciplinary team of specialists who decided on surgery.

A 3 cm mass of the ileum located at 80 cm from the ileocecal valve was found during surgery. The mesentery of this portion of the ileum was retracted with the presence of multiple reactive lymph nodes. We preceded by performing resection the portion of the diseased ileum followed by termino-terminal anastomosis of healthy portions of the ileum (figure 2).

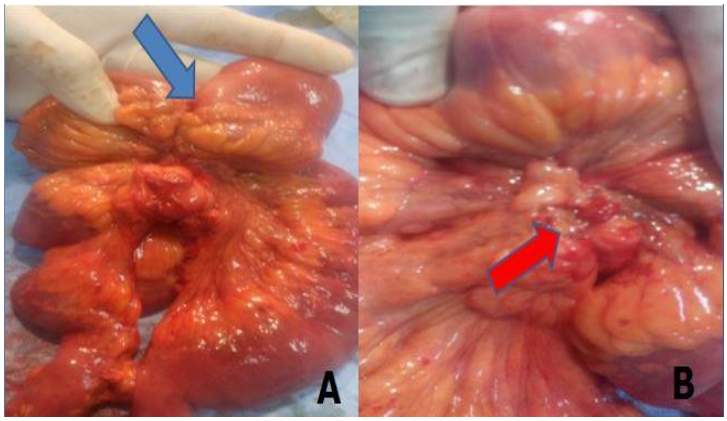

Figure 2: A: The surgical specimen showing the tumor, B: The surgical specimen showing the retractile mesenteritis

The postoperative course was unremarkable. Histopathological examination of the surgical specimen revealed a well differentiated pT4N1, Grade 2 neuroendocrine carcinoma (ENETS), infiltrating the ileal fat. This exam was complemented by Immunohistochemistry which revealed tumor cells expressing synaptophysin and chromogranin A; The Ki67 was estimated at 15%.

The patient's case was discussed again by the multidisciplinary team of specialists who decided on adjuvant chemotherapy. The patient was referred to the department of oncology for adjuvant chemotherapy.

Discussion

Carcinoid tumors are well differentiated neuroendocrine tumors. They develop mostly from enterochromaffin cells located in the gastrointestinal (GI) tract. They are located in The GI tract in 67% of cases and in the bronchial tubes in 25% of cases. Among its localizations in the GI tract, the small intestine represents 42%. It is the second most common tumor of the small intestine after adenocarcinoma [1], Carcinoid tumors are located in the rectum (27%), the stomach (8.7%), and the appendix (5%) [2]. Non carcinoid tumors can be associated with carcinoid tumors of the small intestine in 29% of cases. At the time of diagnosis, 12.9% of patients already have metastases [2].

These tumors of the small intestine are discovered either fortuitously in the event of investigating non-specific digestive disorders [3], or when symptoms such as abdominal mass, GI hemorrhage, mesenteric infarction or Konig's syndrome occur which was the case in our patient.

The confirmation of the carcinoid origin of the tumor is based on:

- Blood immunity test (Serotonin, chromogranins A, B or C, synaptophysin, Neuron Specific Enolase), 5-hydroxyindoleacetic acid urine test (5- HIAA) [4].
- Radiological imaging (ultrasound, CT, octreotide scan) [5]
- Histopathological and immunolabeling findings.

New less radiating radiological modalities such as hydro-MRI or PET-scan are being developed and evaluated [6]. All these examinations are not always necessary. In practice, at least one urine 5-HIAA should be performed during three consecutive days and an additional imaging should be performed by an octreotide scan. An upper GI fibroscopy should be performed if there is gastroduodenal involvement. A cardiac ultrasound should also be performed when there is a carcinoid syndrome. In the presence of hepatic metastases, hepatic MRI, chest CT scan should be performed. Biopsies should be discussed in order to assess the degree of tumor differentiation.

The treatment of carcinoid tumors has a dual purpose: firstly, to perform complete tumor excision or, if possible, to minimize tumor volume (maximum tumor cytoreduction), and secondly to undertake a symptomatic treatment based on somatostatin and its analogues (which has no proven anti-tumor effect but reduces the symptoms of carcinoid syndrome). Many forms of treatment can be undertaken (surgery, immunotherapy, arterial embolization of hepatic metastases). Hence, it is recommended to discuss the case by a multidisciplinary team of specialists (doctors, surgeons, oncologists and interventional radiologists) [7]. Treatment of non-metastatic forms is based on complete surgical excision in order to obtain negative margins (R0).This may be the only way to significantly improve the survival of patients at five years [2,8,9] .

In practice, intestinal resection combined with systematic mesenteric lymph node dissection should be performed, taking into account the incidence of lymph node invasion (40%) even in the case small primary tumors(<1 cm). The excision of mesenteric metastases may justify a colectomy in the case where e mesenteric vessels are affected.

If the tumor is unresectable, non-progressive and non-symptomatic, simple monitoring with regular multidisciplinary re-evaluation will be performed [10].

In the case of an unresectable and evolutive or symptomatic tumor, several attitudes are possible [10]: In case of isolated liver metastases, chemoembolization will be performed

(contraindicated if biliary prosthesis, cholestasis or biliodigestive anastomosis). If it proves to be effective, the possibility of surgical excision will be discussed again in a multidisciplinary setting.

If metastasis is hepatic and extrahepatic, chemotherapy, transient hepatic ischemia of one hour combined with alternate chemotherapy (carcinoid tumors being 90% vascularized by the hepatic artery), or interferon alpha can be discussed.

The combination of these different therapeutic modalities improves overall survival and in most cases the quality of life [11]. In all cases, the management of these patients must be "aggressive": Current chemotherapies make it possible to regress certain lesions considered previously unresectable. Therefore, regular discussions may be necessary to decide on the possibility of an excision which remains the only curative treatment.

The prognosis of carcinoid tumors of the small intestine is better than that of adenocarcinomas (given the low scalability of these lesions), with 67-75% survival at 5 years for resectable tumors and 50% survival at 5 years for unresectable tumors [10,12]. However, it is worse than rectal tumors (88% survival), tumors of the appendix (71%), and bronchopulmonary tumors (73.5%).

Conclusion

Carcinoid tumors of the small intestine are often small and relatively evolve slowly. Thus, these tumors are lately diagnosed. In a majority of cases, the diagnosis is suspected when there is a retractile mesenteritis or a rare carcinoid syndrome. Diagnosis is confirmed by the pathological examination. Surgery is the only curative treatment to date.

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