Synchronous association of gastrointestinal stromal tumors of the jejunum and sigmoideal adenocarcinoma: A case report

Jejunum and sigmoideal adenokarsinominun gastrointestinal stromal tumörlerinin senkron ilişkisi: Olgu sunumu

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Abstract

The association of a gastrointestinal stromal tumor (GIST) with an epithelial tumor of the digestive tract is very rare and frequently discovered by chance during the surgical treatment of digestive carcinoma. These tumors developed from the mesenchyme are known to have a good prognosis given their low malignancy potential. The possibility of a genetic mutation or an oncogenic agent capable of inducing tumors of different histotypes seems less plausible but it is not to be discarded. We report the case of a 74-year-old patient, operated for a sigmoid adenocarcinoma, with incidental discovery of a small bowel tumor whose histology confirmed the diagnosis of a GIST.

Keywords: Synchronous tumors, Gastrointestinal stromal tumor, Adenocarcinoma, Digestive tract

Özet

Sindirim kanalının epitelial tümörü ile gastrointestinal stromal tümörün (GIST) birlikteliği çok nadirdir ve sindirim karsinominun cerrahi tedavisi sırasında sıklıkla tesadüfen keşfedilmiştir. Mezenşimden gelişen bu tümörlerin, düşük malignite potansiyelleri göz önüne alınında iyi bir prognoza sahip oldukları bilinmektedir. Bir genetik mutasyonun veya farklı histotiplerin tümörlerini indükleyebilen onkojenik bir maddenin olası olduğu daha az akla yakın görülmektedir, ancak yedsiz. Biz histolojisi bir GIST tanısı doğrulan ince bağırsak tümörünün tesadüfen keşfesi, sigmoid adenokarsinominın belirginlik edilen 74 yaşında bir hastayı sunuyoruz.

Anahtar kelimeler: Senkron tümörler, Gastrointestinal stromal tümör, Adenokarsinom, Sindirim sistemi

Introduction

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract. Their metachronous or synchronous association with digestive carcinoma has been rarely reported in the literature [1-3]. We report the synchronous association of a jejunum’s GIST with a sigmoid adenocarcinoma.

Case presentation

A 74-year-old patient, diabetic and hypertensive on treatment, who presented two months before her admission defecatory rectorrhagia of low abundance. An abdominal CT scan revealed a tumor process of the partially stenotic sigmoid loop (figure 1), without metastases found during the extension assessment.

A colonoscopy showed a lesion 25 cm from the anal margin, stenosing and impassable. A biopsy was made and revealed well differentiated and infiltrating lieberkhnhal adenocarcinoma. The patient has undergone a sigmoid resection, the surgical exploration discovered a jejunal tumor, and the surgical procedure was completed by a cuneiform jejunal resection removing the tumor. Anatomiopathological study confirmed the diagnosis of lieberkhnnal adenocarcinoma with clean margins classified stage pT2 N1h M0. The histological analysis of the jejunal tumor concluded to the diagnosis of GIST (figure 2, 3) (CD117 and the strongly positive DOG1) with low risk of recurrence. The postoperative thoracoabdominopelvic CT scan and tumor markers were normal. The patient received adjuvant chemotherapy: oxaliplatin and capecitabine (Xelox).
Synchronous malignancy of gastrointestinal stromal tumors and adenocarcinoma

Discussion

GISTs are the most common mesenchymal tumors of the digestive tract. Often isolated, they can rarely come into association. They can be observed in the context of family syndromes. The Carney Triad associates multiple gastric stromal tumors, a pulmonary chondroma, and an extra-adrenal paraganglioma [1].

Of the 1765 gastric GISTs studied by Miettinen and Lasota [2], only four cases of Carney’s triad were found. GISTs can also be seen in 5 to 25% of cases in type 1 neurofibromatosis. There are familial forms of GIST associated with hyperpigmentation, urticaria pigmentosa and / or systemic mastocytosis [1]. An oncogenetic consultation is recommended in these cases after information and agreement of the patient. Apart from these associations, the synchronous or metachronous development of GISTs and digestive epithelial tumors is very rare. These GISTs are often of low malignancy potential.

About 30 cases of mesenchymal tumors including synchronous GIST with other gastrointestinal tumors have been reported [3], including adenocarcinomas, lymphomas and carcinoid tumors [3,4] but also hemopathies, melanomas, breast, kidney, prostate and female genital cancers [5,6]. In a study involving 783 patients, Pandurengan et al. [7] showed that about 20% of cases with GIST developed other types of digestive cancers. The association of GIST with colorectal adenocarcinoma is the most frequently reported [8]. In most cases, the discovery of GISTs is fortuitous during laparotomy for primary adenocarcinoma [6].

Maiorana et al. [3], by examining 2035 carcinomas, showed six cases of stromal and epithelial synchronous tumors including five adenocarcinomas and one carcinoid tumor (a frequency of 0.29%). Chacon et al. [9] evaluated the incidence of tumors associated with GIST and reported two cases of colonic adenocarcinoma in a series of 86 patients (a frequency of 2.32%). Thirty isolated cases of GIST associated with gastrointestinal adenocarcinoma have been reported mainly in Japanese literature. GISTs can also be associated with other digestive mesenchymal tumors such as lipomas. Exceptionally, triple associations have been reported.

Two etiopathogenic hypotheses have been advanced to explain the association of GIST with digestive carcinoma. Maiorana et al. [3], noting that most of the cases reported come from Japan, have suggested the hypothesis of a simple accidental association, especially in countries with a high incidence of digestive cancer especially gastric cancer. As for Cohen et al. [10], they evoke the possibility of a genetic mutation or an oncogenic agent capable of inducing tumors which are histologically different.

Agaimy and Wunsch [5] analyzed 97 GIST cases, of which 18 (18.6%) were associated with other tumors. This study involved 12 women and six men aged 43 to 87 years. Twelve GISTs were present in the stomach, four in the small intestine, one in the duodenum and one in the appendix. The tumors associated with these GISTs were carcinomas of gastrointestinal and pancreatic origin in nine cases, gynecological in three cases, mammary in two cases, pulmonary in two cases, prostatic in one case, and renal in one case and a lymphomatous proliferation in two cases.

The rather limited number of cases of this association does not allow defining its exact pathogenesis which requires further studies. Given the low risk of recurrence of GISTs in association with digestive carcinomas, the treatment should aim the epithelial lesion, since it is the surveillance that is adopted for low-risk GISTs.

Conclusion

The reported case is about a synchronous development of a sigmoid adenocarcinoma and a jejunal GIST with a low risk of recurrence. Their association is very rare and the hypothesis of a pathogenic relationship between these different histological
types resulting from a common genetic disorder is not yet confirmed but remains plausible for better management.

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