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SCIENTIFIC LETTER

Leiomyosarcoma of the colon. A very uncommon condition with poor prognosis☆,☆☆

Leiomiosarcoma de colon. Una entidad muy infrecuente con mal pronóstico

Gastrointestinal mesenchymal tumours are uncommon. The most common types are gastrointestinal stroma tumours (GISTs)[1,2]; the rarest types are smooth muscle tumours (leiomyomas and leiomyosarcomas) and schwannomas. The most common types in the colon are leiomyomas and submucosal lipomas. A diagnosis of a GIST is rare in this anatomical location, and a diagnosis of a leiomyosarcoma is rarer still (<1% of cases[2]). Its incidence is 1 per 100,000 inhabitants, and it accounts for 0.12% of colon neoplasms. Leiomyosarcoma of the colon is an aggressive tumour associated with a poor prognosis.[3]

We present the case of a 71-year-old male with a history of hypertension, COPD, dyslipidaemia and appendectomy. For the purpose of testing due to constipation with a one year period of progression, he underwent a complete colonoscopy which revealed a polypoid lesion in the transverse colon, 80 cm from the anal margin. Pathology diagnosed the lesion as a leiomyosarcoma. Testing was extended to include a CT scan of the chest, abdomen and pelvis; the results were negative for distant lesions. A small locoregional lymphadenopathy measuring 11 mm was discovered. Given these findings, a laparoscopic sub-total colectomy with ileo-sigmoid anastomosis was performed.

The definitive pathology report described a polypoid and ulcerated lesion with a stenosing lumen measuring $4.5 \times 4 \times 4$ cm, with no necrosis and a mitotic rate of 12 per 10 high-power fields, corresponding to a pT1N0M0 tumour with a histological grade of G2.

Leiomyosarcomas of the colon are aggressive tumours with a high rate of local recurrence and fundamentally haematogenous spread that are associated with a poor prognosis.

Unlike GISTs, leiomyosarcomas preferentially occur in the small bowel (45%) or the large bowel (38%); they are rarely found in the stomach or oesophagus.[3] They may also present in the rectum. They most commonly affect males in their fifties and sixties.[4,5]

Morphologically, they may be indistinguishable from GISTs; immunohistochemistry is required for their diagnosis. This explains why many mesenchymal tumours in the pre-immunohistochemistry era were diagnosed as leiomyosarcomas.[4]

They usually present as sub-mucosal polyps or intramural nodules arising from the *muscularis mucosae* or the muscularis propria; ulceration and necrosis are common.[3] Characteristically, they test positive for smooth muscle markers such as desmin and actin and often calponin and h-Caldesmon, and negative for GIST markers (c-kit, CD34 and Dog-1).[2] A proper differential diagnosis between the two diseases is essential, as they have radically different prognoses.

Treatment is surgery with complete removal and suitable margins; even with this treatment, recurrence is seen in many cases.[3,5] The role of lymphadenectomy is debated; however, the guidelines recommend performing conventional oncology procedures with complete excision of the mesocolon as in intestinal adenocarcinoma in this location.[3,5]

As for adjuvant treatment, these tumours are usually insensitive to chemotherapy and do not respond to imatinib. sunitinib has very limited efficacy.[3] Adjuvant therapy with anthracyclines is considered the first-line choice in patients with large tumours and high-grade tumours.[4,5] The efficacy of radiotherapy is also a matter of debate, although it has been suggested that, as an adjuvant therapy, it may reduce local recurrence in sarcomas of the rectum. Given this low sensitivity to adjuvant therapy, in cases of local recurrence and/or metastasis, the recommended treatment is surgical resection.[2]

With regard to follow-up, for tumours with an intermediate to high histological grade, it is recommended that a CT scan of the chest and abdomen be performed every 3–4 months in the first 2 years, then 2 times per year up to 5 years and then once per year after the first 5 years have elapsed. For low-grade tumours, it is recommended that a CT scan of the chest and abdomen be performed every 4–6 months in the first 5 years, then once per year.[5]

The most common sites of metastasis are the liver and the peritoneum, followed by the lungs.[3] Given that spread to the lymph nodes is rare, the most important prognostic indicator is a size $\geq$5 cm; mitotic rate has no prognostic value.

Overall five-year survival following radical surgery is 51.6%.[3,4] The primary cause of death is secondary to liver and lung metastases. Age over 40 years, tumour necrosis,

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distant disease and large tumour size have been identified as poor prognostic factors.

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Black oesophagus (acute oesophageal necrosis)☆

Esófago negro (necrosis aguda esofágica)

We present the case of an 85-year-old female patient with a history of hypertension, diabetes mellitus, dyslipidaemia and pulmonary embolism. She had been experiencing epigastric pain for the past 3 days associated with nausea and vomiting. Laboratory testing found high acute-phase reactants. A plain X-ray and an abdominal CT scan were also performed and showed gastric dilatation and signs suggestive of chronic pancreatitis. It was decided to extend the study by performing a gastroscopy. This revealed an oesophageal surface with blackish discolouration from the proximal oesophagus (Fig. 1A) that did not detach with lavage, suggestive of a necrotic process, associated with longitudinal ulcerations and an abrupt change in the oesophageal–gastric transition (Fig. 1B and C), with no evidence of lesions in the stomach. Multiple ulcers alternating with areas of necrosis were also present in the second duodenal segment (Fig. 1D). Biopsies taken confirmed necrotic ischaemic changes in the duodenal mucosa.

Given the endoscopic findings, another CT scan was performed, this time a thoraco-abdominal CT scan, which ruled out complications. Treatment was started with total parenteral nutrition and intravenous omeprazole as well as empirical antifungal and antibiotic coverage with meropenem and fluconazole. The patient's subsequent clinical course was satisfactory; therefore, following conservative management for 15 days, another gastroscopy was performed. It showed an oesophageal mucosa with a whitish appearance, covered in fibrin, with a decrease in its calibre towards the cardia, which exhibited obstructive stenosis (Fig. 1E). Several sessions of endoscopic dilatation were performed successfully (Fig. 1F).

Acute oesophageal necrosis is an uncommon entity reported for the first time in 1990 by Goldenberg et al.[1] It primarily affects males in their fifties,[2] and its prevalence and incidence are low, though its exact incidence and prevalence cannot be known, as it is an under-diagnosed condition.[3] Its aetiology is multifactorial, and primarily secondary to ischaemic damage in patients with cardiovascular risk factors and in a context of haemodynamic compromise or low output, including sepsis, heart failure, acute haemorrhage and systemic inflammatory response. Circulation from branches of the coeliac trunk indicates that there may be concomitant lesions in the distal oesophagus and duodenum. Other factors that influence the development of lesions are abnormalities in oesophageal defensive mechanisms and massive passage of gastric contents towards the oesophagus.[3] In the case reported, a potential flare-up of chronic pancreatitis in a patient of advanced age with cardiovascular risk factors is put forward as a possible trigger of oesophageal and duodenal lesions.

In up to 90% of reported cases the initial sign is upper gastrointestinal bleeding,[3] though other symptoms such as abdominal pain and vomiting may appear or the patient may be asymptomatic. The condition is diagnosed by gastroscopy. It is not strictly necessary to perform biopsies,

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