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Review article

Rare Tumors of the Rectum. Narrative Review☆

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ABSTRACT

Most rectal neoplasms are adenocarcinomas, but there is a small percentage of tumors which are of other histological cell lines such as neuroendocrine tumors, sarcomas, lymphomas, and squamous cell carcinomas, which have special characteristics and different treatments. We have reviewed these rare tumors of the rectum from a clinical and surgical point of view.

Neoplasias de recto poco frecuentes. Revisión de conjunto

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RESUMEN

La mayoría de los cánceres de recto son adenocarcinomas, pero existe un pequeño porcentaje de tumores de otras estirpes histológicas, como neoplasias neuroendocrinas, sarcomas, linfomas y carcinomas de células escamosas, que tienen unas características y tratamientos diferentes. Hemos efectuado una revisión de estos raros tumores del recto desde un punto de vista clínico y quirúrgico.

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Introduction

Rectal cancer is typically adenocarcinoma. Nonetheless, there are other types of tumors that are much less common, such as neuroendocrine neoplasms, lymphomas, sarcomas, and squamous-cell carcinomas, which can also be located in the rectum.[1–4] The incidence of each of these tumors is difficult to calculate. According to the data from 2005 to 2009 from the National Cancer Institute's Surveillance Epidemiology and End Results (SEER),[5] out of 183 000 colorectal cancers (not including lymphomas), 94.3% were adenocarcinomas, 1.7% other carcinomas, 3.3% carcinoid tumors, 0.5% epidermoid carcinomas, 0.1% sarcomas, and 0.1% other types.

All these tumors present very different characteristics from adenocarcinomas, which also makes their treatment and prognosis very different (Table 1). In addition, there have been recent modifications in the diagnosis and treatment of some of the types. This all causes doubt and controversy in their clinical management, and it is recommended that they be treated by a multidisciplinary team including surgeons, oncologists, pathologists, and radiologists.[6–8]

The objective of this article is to review the clinical and surgical management of these uncommon rectal neoplasms. We used PubMed to review the literature from 1997 to 2012 using the key words related to a colorectal location stated at the beginning of this article.

Neuroendocrine Neoplasms

Neuroendocrine neoplasms are epithelial and present neuroendocrine differentiation.[9] They can be located in different organs.

They are classified[10,11] by their degree of differentiation (well or poorly differentiated) and their histologic grade (G1, G2 and G3), based on the number of mitoses and the Ki67 index.[9,12–14] Three different groups are defined: neuroendocrine tumors (NET), neuroendocrine carcinoma (NEC), and mixed adenoneuroendocrine carcinoma (MANEC).[7,12]

Neuroendocrine Tumors

NET are well-differentiated neuroendocrine neoplasms made up of tumor cells that express neuroendocrine markers (chromogranin A, synaptophysin) (Fig. 1) and hormones.[12] Cellular atypia and proliferative activity are low. By definition, they are grade G1 or G2 tumors. This category includes lesions that were previously called ''carcinoid tumors'', a denomination that is now criticized and is no longer included in the gastrointestinal NET classifications, but is still widely used. The rectal location represents 18% of all NET and 27% of all digestive tract tumors.[12,15]

The annual incidence of rectal NET, according to the SEER,[5] is 0.86 per 100 000, which has experienced a large increase in recent decades.[7] The incidence is higher in Asians,[12] and mean age is 56.[16]

NET usually present as small polypoid lesions or submucosal nodules.[4] 45% measure 10 mm or less, while only 17% measure more than 20 mm.[17] These tumors are frequently asymptomatic[12,18] or accompanied by mild symptoms such as bleeding, tenesmus or discomfort.[17,18]

49% of the NET only affect the mucosa and submucosa, 24% infiltrate the muscularis propria, and 15% extend to the perirectal fat.[17] 75%–85% are located in the rectal wall.[12] Tumor size and lymphovascular invasions are risk factors for lymph node involvement.[16,17,19,20]

Liver metastases become more frequent as tumor size increases.[16] These are present in 1.7% of the NET$\leq$1 cm, in 15% of those between 1 and 2 cm and in 50% of those $>$2 cm.[20]

The majority of NET are diagnosed endoscopically.[12] Endorectal ultrasound seems to be the best method for

Table 1 – Particularities of Each Type of Tumor.

Tumor type	Cells of origin	Risk factors	Prognostic factors	Usual treatment	Adjuvant treatment	5-year survival %
NET	Kultschitzky cells	Unknown	– No. of mitoses – Ki67	<1 cm: local resection >2 cm: oncologic resection	–	90
NEC, MANEC				Oncologic resection	CTx	15
GIST	Interstitial cells of Cajal	Unknown	– Size – No. of mitoses – Tumor rupture	Resection with free margins	Imatinib	70
Other sarcomas	Rectal connective tissue	– Previous RT	– Differentiation – Tumor necrosis – No. of mitoses	Resection with free margins	RT	50
Lymphomas	Rectal lymphoid tissue	– HIV – IBD – Immunosuppression	– Histologic type – Histologic grade	Oncologic resection	CTx	30–60
Squamous carcinoma	Rectal epithelium	– HPV – Chronic rectal inflammatory processes	–	CRT Oncologic resection	–	30

IBD: inflammatory bowel disease; GIST: gastrointestinal stromal tumor; HIV: human immunodeficiency virus; HPV: human papilloma virus; MANEC: mixed adenoneuroendocrine carcinoma; NEC: neuroendocrine carcinoma; NET: neuroendocrine tumor; CRT: chemoradiotherapy; CTx: chemotherapy; RT: radiotherapy.