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The exceptionally rare phenomenon of well-differentiated colon neuroendocrine tumors

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BACKGROUND

Colonic neuroendocrine tumors (NETs), excluding rectal NETs, are often described as relatively common and aggressive, with inferior median survival compared to other gastrointestinal (GI) primary sites. However, epidemiological databases may conflate well-differentiated NETs with poorly differentiated NECs, leading to a lack of precise data on the prevalence, clinical behavior, and prognosis of well-differentiated colonic NETs.

METHODS

We analyzed a large institutional database to identify patients with well-differentiated NETs originating in the colon, excluding rectal NETs. Cecal NETs were included, however Ileocecal NETs (overlapping the ileocecal valve) were not. We assessed their prevalence compared to other primary sites, grade, stage, and prognosis.

RESULTS

Among 3639 patients with gastroenteropancreatic (GEP) NETs, only 19 (0.5%) had well-differentiated colonic NETs. This included 11 cecal and 8 sigmoid colon primaries (2 of them described as 'rectosigmoid'). No tumors originated in the ascending, transverse, or descending colon. Sigmoid NETs were typically early-stage polyps, discovered incidentally during colonoscopy. In contrast, 8 of the 11 cecal NETs metastasized (p=0.04), with 6 of these patients (55%) exhibiting carcinoid syndrome and none in the sigmoid cases (p=0.01).

CONCLUSIONS

Well-differentiated colon NETs are exceptionally rare, comprising approximately 0.5% of GEP-NETs. These tumors fall into two distinct categories: cecal NETs, which resemble ileal NETs in behavior, and sigmoid NETs, which appear to be quite similar to rectal NETs. The broad categorization of colonic 'NETs' in epidemiologic databases likely includes NECs, obscuring the true clinical picture.

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