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Solitary Ganglioneuromatosis of the Descending Colon, Presenting as Giant Retroperitoneal Tumour

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Abstract
Ganglioneuroma (GN) is a benign neoplasia of the autonomous nervous system, colonic GN is uncommon in adults. There are three subgroups: polypoid GN, ganglioneuromatous polyposis and diffuse ganglioneuromatosis. Ganglioneuromatosis is highly-associated to neurofibromatosis type 1 (NF1) and multiple endocrine neoplasia type 2b (MEN2B). A 68-year-old female, with a discrete retarded emission of stools, was admitted for a large tumor in the left flank; CT scan, urography and barium enema demonstrated a large retroperitoneal mass, presumed as sarcoma. Open surgery discovered a 16/10/11 cm solid and encapsulated tumor, attached to the retroperitoneal descending colon, with no macroscopic mucosal involvement; the pathologic diagnosis of the resected specimen (en-bloc tumorectomy with limited colectomy) was intramural colonic ganglio-neuromatosis. Anamnesis, physical examination and complete endoscopic explorations showed no evidence of personal bearing or familial aggregation of genetic syndromes. In adults, association of transmural ganglioneuromatosis to NF1 or MEN2B is not mandatory; presentation often mimics obstructive carcinoma and positive diagnosis is provided by pathological examination of the resected specimen. In this peculiar case, the loose tissue of the retroperitoneal space favoured a slow development of intramural ganglioneuromatosis, presenting as a gigantic retroperitoneal mass with no radiological evidence of its colonic origin.
Key words: colonic ganglioneuroma, intestinal ganglioneuromatosis, retroperitoneal tumor

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