

Solitary retroperitoneal neurofibroma: not as small as it seems

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A 41-year-old man presented to our endoscopy unit for a colonoscopic examination. On admission, he complained of recent mild abdominal pain in the right lower quadrant. He denied altered bowel movements, weight loss, or blood in a stool. Another reason for performing colonoscopy was a recent death of his father due to colon cancer. There was no family history of hereditary diseases (eg, neurofibromatosis). On physical examination, except mild abdominal tenderness in the right iliac fossa, no other abnormalities were detected (in particular, skin changes such as café au lait spots, axillary freckling, or subcutaneous nodules). Routine laboratory test results were within the reference ranges.

The colonoscopy revealed a small subepithelial tumor of 15 mm in diameter in the cecum (FIGURE 1A). The patient was screened with abdominal and pelvic computed tomography (CT), which showed a homogeneous retroperitoneal tumor of 80 mm in diameter between the right psoas and iliacus muscles (FIGURE 1B). A laparotomy revealed a well-defined, soft, and encapsulated tumor measuring 90 mm in the pelvis (FIGURE 1C). The lesion was removed completely (FIGURE 1D), without sequelae. The postoperative histopathological examination of the specimen showed proliferation of spindle cells with wavy nuclei without pleomorphism and wire-like collagen background as well as positive staining for S100 protein (FIGURE 1E). A neurofibroma was diagnosed. The patient remains free from any symptoms in a 3-year follow-up.

Neurofibromas are tumors arising from the neural sheath.¹ Their typical location is the skin of the trunk and head. They are usually multiple and associated with von Recklinghausen disease.^{1,2} However, they can also develop in patients without any hereditary disorder as solitary

tumors of deep organs, in various locations, preferably in the pelvis.² Retroperitoneal neurofibromas are indolent, benign, and slow-growing tumors. However, the malignant transformation is also possible.³ Typically, the tumor is detected when it is large enough to cause symptoms.¹ The tumor dimensions and its vascularization often make surgical removal challenging.¹ The clinical symptoms (eg, pain, motility disorders, or urinary obstruction) are frequently associated with compression of the adjacent organs.²

Neurofibromas are treated surgically, and the diagnosis is based on the combination of histopathological and immunohistochemical analysis of the postoperative specimen (typical features are shown in FIGURE 1E). Preoperative imaging (eg, CT) followed by a differential diagnosis of the colonoscopic findings of submucosal lesions in the cecum are essential to avoid biopsy, rupture, and adverse events at the time of surgery.^{4,5}



FIGURE 1 A – colonoscopy showing a small subepithelial tumor (diameter, 15 mm) in the cecum

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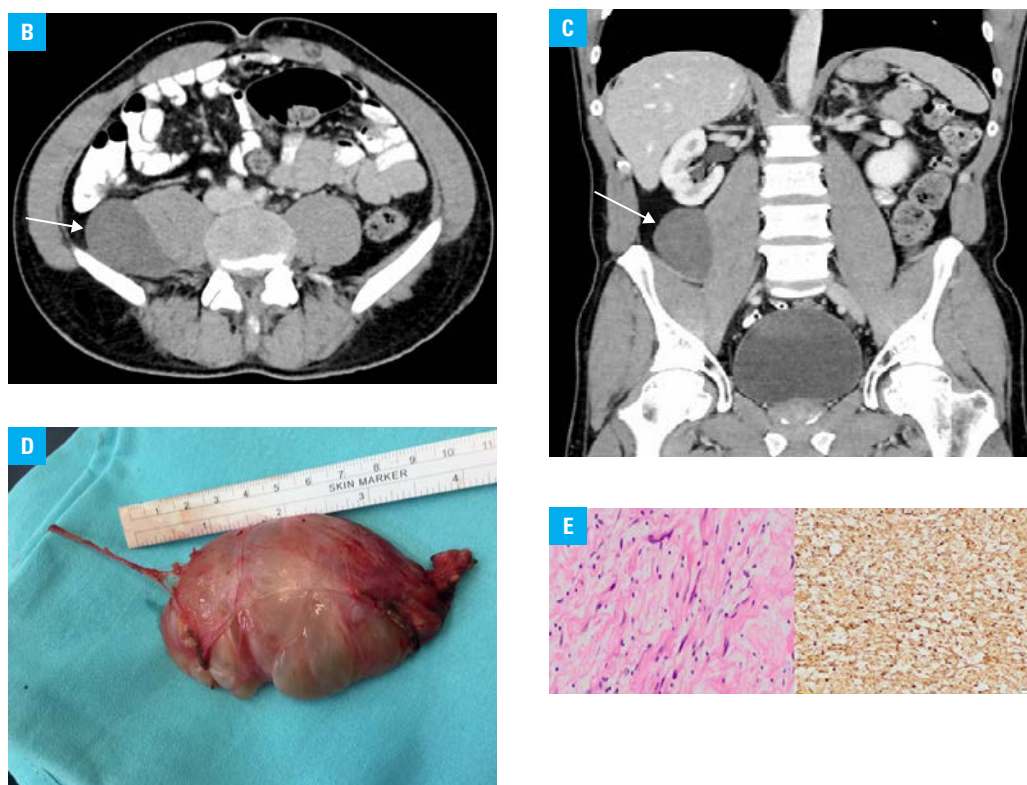
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FIGURE 1 B, C – contrast-enhanced computed tomography scan of the abdomen and pelvis in 4-mm slices: axial (**B**) and coronal (**C**) views show a well-circumscribed and homogeneous lesion 80 × 65 × 43 mm (arrows) between the right psoas and iliacus muscles; **D** – an excised tumor (diameter, 90 mm); **E** – a histopathological specimen of a neurofibroma showing spindle-shaped cells with wire-like collagen and twisting nuclei without pleomorphism (hematoxylin and eosin staining) and positive staining for S100 protein.



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