


Colonic schwannoma: A case of unusual presentation and outcome

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Abstract

Background: Schwannomas are mesenchymal tumors arising from neural sheath cells and whose diagnosis is based on immunohistochemistry. The digestive and especially colonic location of this tumor is rare. Commonly described in elderly patient, their malignancy is unusual. **Case report:** We report the case of a 23-year-old girl, with learning disability, operated in emergency for acute peritonitis. Peroperatively, we discovered a peritonitis secondary to a bulky perforated cecal tumor. We performed a right colectomy and an ileostomy. The posterior margin of the resection was macroscopically involved. The diagnosis of colonic schwannoma was confirmed with immunopathological examination of the surgical specimen. Surgical exploration 4 months later and morphological investigations during 2 years showed stability of the tumoral residue in the right iliac fossa. However, we noticed, on CT scanning control 2 years postoperatively, the appearance of a mesenteric recurrence. Exploratory laparotomy confirmed the unresectability of this mesenteric mass and showed the presence of multiple parietal nodules whose biopsies revealed their neurofibromatous nature. The 4 year follow-up of the patient didn't reveal any complication rather than need to right nephrostomy due to ureter compression by the primary tumoral residue. **Discussion:** Colonic schwannoma is a rare disease, commonly described in uncomplicated stages. The learning disability of our patient had favored the absence of declared symptoms and promoted the evolution of the cecal schwannoma until its perforation. This complication was not described before. Local recurrence has been also rarely reported in the literature. Association with neurofibroma may be hereditary in a context of neurofibromatosis or sporadic. **Conclusion:** Colonic schwannoma may have polymorphic presentation mimicking malignant tumor in such cases. Not treated in time, it can lead to severe complications, such as tumoral perforation. Surgical resection remains the mainstay treatment. The slow evolutionary genius of schwannoma, even when incompletely resected by necessity, confirms its good prognosis.

Keywords

colon, schwannoma, treatment, outcome

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Background

The Schwannoma is a tumor arising from the Schwann cells. This tumor can be found throughout the body along the peripheral nerves, but its colonic location is extremely rare (<5% of all gastrointestinal tract schwannomas).¹ In decreasing frequency, they are located in the caecum, sigmoid, rectosigmoid, transverse colon and the descending colon.²

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Colonic schwannomas are commonly described in elderly patient. The circumstances of diagnosis are those of an uncomplicated colonic tumor. Although considered benign, it may recur locally if incompletely resected.^{1,3,4} Locoregional or liver metastasis are observed in aggressive tumors (2%).¹ Radical surgery is, therefore, the gold standard of treatment.¹

Case Report

This is the case of a 23-year-old girl with a history of learning disability since childhood secondary to meningitis; who presented to a regional hospital emergency with a generalized abdominal pain that had been evolving for 2 days. At initial examination; the patient had a clinicobiological presentation of generalized peritonitis. Abdominal ultrasonography showed significant infiltration of the periappendicular fat with intraperitoneal effusion of low to medium abundance. As the patient had signs of septic shock, we decided to operate her through median laparotomy, peritonitis due to appendicular perforation was highly suspected.

Peroperative exploration showed a purulent effusion of average abundance with food debris secondary to a perforation of a voluminous tumoral mass of the caecum. The tumor was fixed posteriorly. It invades the adjacent structures. The palpation of the liver and the right annexe did not find secondary lesions and there were no nodules of carcinomatosis. The right mesocolon was strewn with adenomegaly, the tumor adhered posteriorly to the second duodenum. A right hemicolectomy R2 was performed with ileostomy (Figure 1). The postoperative course was marked by a state of septic shock requiring her transfer to our university hospital and 12 days stay in Intensive Care Unit.

Pathological examination of the surgical specimen concluded to a mesenchymal tumor of stromal origin and the immunohistochemistry allowed the diagnosis of right colon schwannoma (Figure 2).

A thoracoabdominopelvic CT scan performed 2 months later showed a 3.5 × 2.5 cm tumoral residue at right iliac fossa and no secondary site (Figure 3).

This case was discussed in multidisciplinary meeting and the decision was to make a complementary resection of the tumoral residue when restoring continuity (after 3 months from initial surgery). This complement of resection was not feasible since the tumoral residue was invading right annexe; right iliac vessels and right ureter. We had only restore the digestive continuity.

The patient has been followed regularly at our outpatient clinic. Two years postoperatively CT scanning control showed a tissue mass involving the small bowel measuring 13 x 8 x 6 cm corresponding to mesenteric recurrence, with stability of the old tumoral residue in the right iliac fossa (Figure 4).

Exploratory laparotomy was performed showing a voluminous mass invading the root of the mesentery and

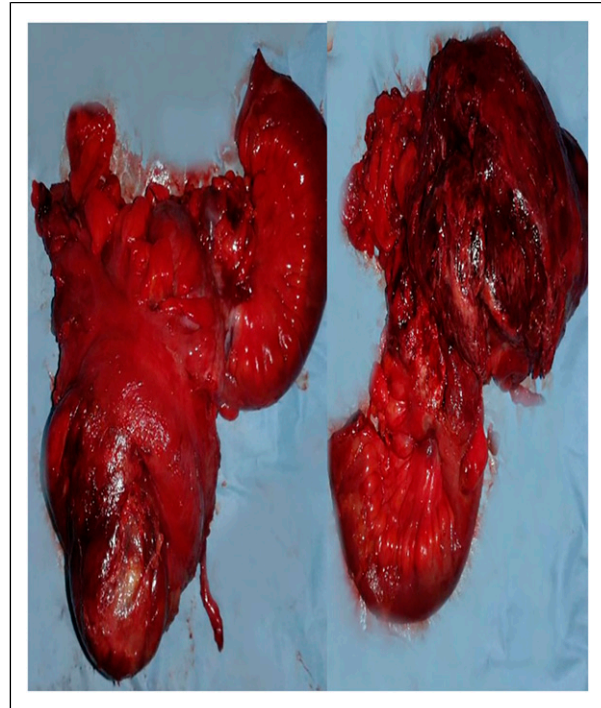


Figure 1. Right colectomy specimen: perforated tumor of the caecum.

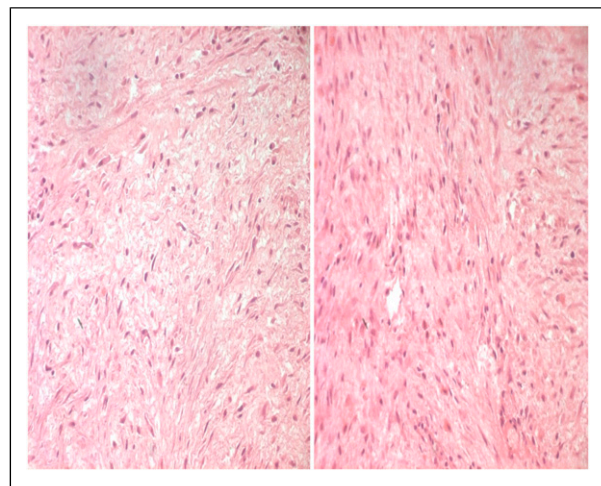


Figure 2. Anatomopathological examination of the specimen H.E. *200: Verocay Nodule: alternation between hypercellular area and hypocellular area (Antoni A and B areas respectively).

multiple parietal nodules (Figure 5). This mass was unresectable given the risk of compromising the residual small bowel's vascularization. Surgical biopsies of the parietal nodules confirmed their neurofibromatous nature.

No adjuvant treatment was indicated on multidisciplinary meeting, since the tumor was unresectable and since there is no based evidence medicine indication of radiotherapy or chemotherapy.

Otherwise, neurofibromatosis was suspected in our patient, but she didn't have oncogenetic investigation since lack of major criteria justifying the realization of such relatively expensive investigations.

The 4 year follow-up of the patient didn't reveal any complications, rather than need to right nephrostomy due to ureter compression by the primary tumoral residue.

Discussion

Schwannomas were first described by Verocay in 1910.^{1,3} They are neurogenic tumors of the gastrointestinal tract arising from the Schwann cells of the nerve sheath.

Because of the small number of cases of schwannomas located in the digestive tract, the epidemiological pattern

and the clinicopathological features of this tumor are not fully established.

Schwannomas are seen in both genders equally and are mostly found after the sixth and seventh decades of life, but can appear at any age.^{1,3,5} The young age of our patient is unusual.

Intraperitoneal schwannomas are mostly located in the stomach.⁵ Schwannomas of the colon and rectum are uncommon and incompletely characterized tumors.^{1,4}

Patients with colorectal schwannoma are usually asymptomatic, but in some cases, tenesmus, rectal bleeding, colonic obstruction and abdominal pain can be experienced.^{1,3,6} The most common location is cecum followed by sigmoid and transverse colon, descending colon, and rectum.⁶ In our literature review we didn't find a case presenting colonic schwannoma perforation. Our patient may not complain of any symptom until the tumor perforation due to the context of the mental disability.

The diagnosis of certainty is mostly obtained postoperatively through the pathologic examination of the specimen.¹ Preoperative diagnosis of schwannoma is difficult due to its tissue density and the tendency for ulceration. Biopsies are oftenly noncontributing. Colonoscopy, abdominal ultrasound (US), abdominal CT and abdominal magnetic resonance imaging (MRI) may aid in evaluating the contours of colorectal schwannomas and their relationship with surrounding organs, as well as tumor multiplicity or metastasis, such as the diagnostic approach in any other colorectal tumor.⁷

Immunohistochemical analysis of the tumor cells remains the most important diagnostic tool.^{5,7}

Many pathological criteria have been studied in relation to tumor behavior, and so far no single criterion alone allows sufficient prediction of the degree of malignancy. A mitotic activity rate of more than 5 mitoses per field at high



Figure 3. CT Scan 2 months postoperatively: stability of the tumoral residue on right iliac fossa.

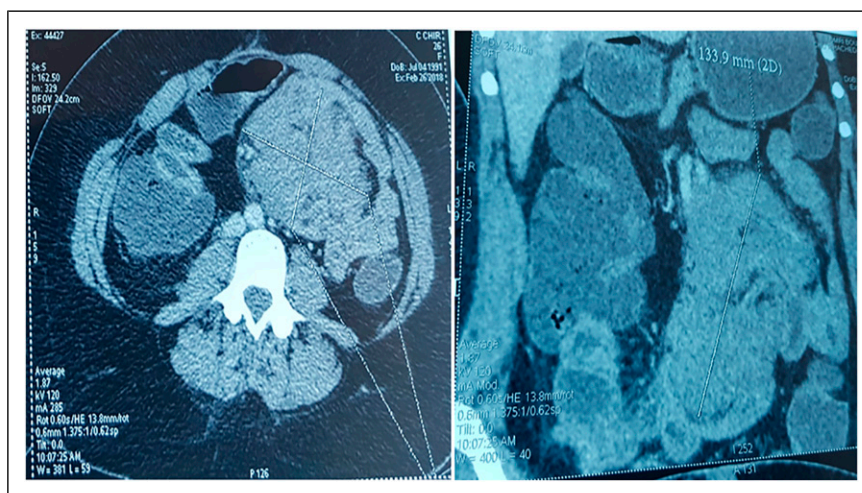


Figure 4. CT scanning control 2 years postoperatively: bulky mesenteric mass.

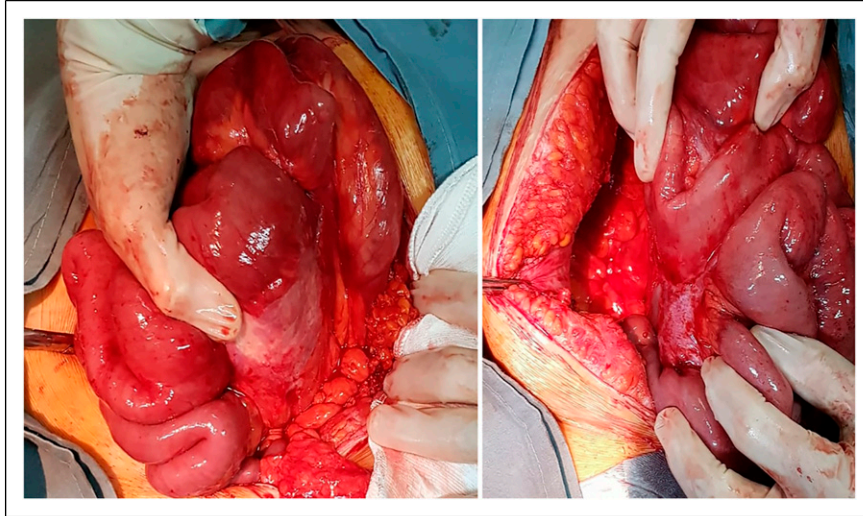


Figure 5. Exploratory laparotomy: bulky unresectable mass invading the root of the mesentery.

magnification and a tumor size greater than 5 cm tend to be associated with a high risk of metastasis or recurrence.^{5,8,9} Malignant profile was judged based on long-term local and distal recurrence.¹

Colonic schwannomas are known as usually slow-growing tumors. Although they are considered as benign tumors in 98% of cases, local recurrence has been reported rarely in the literature after complete surgical resection and was considered as a clinical criterion of malignancy.¹ Moreover, they may recur locally if excision is incomplete.^{4,10} In the case of our patient, we can't retain malignancy because the primary tumor was incompletely resected and was perforated initially. Not only that, but also our patient survived 4 years after the initial surgery with the tumoral residue and 2 years with the mesenteric recurrence. All these facts confirmed the slow evolutionary genius of schwannoma.

The best therapeutic option is complete surgical removal with wide resection margins.^{1,11} Lymph nodes resection is not recommended when the diagnosis is certain preoperatively, because the risk of malignant transformation is low.^{11,12} For more aggressive tumors, the same surgical criteria should be used, but greater rigor in postoperative follow-up is recommended. The surgical approach depends on the size and location of the tumor and on its histopathologic pattern.^{4,5} Our patient had an R2 right hemicolectomy because of the involvement of the right annexes, ureter and iliac vessels. Our conservative attitude for the residue was justified by the histological benignity of the tumor and the stability of its aspect in the different CT scanning control.

Furthermore, association with neurofibroma may be hereditary in a context of neurofibromatosis or sporadic.¹³

The role of radiotherapy or chemotherapy to date remains unclear and there is no consensus for the indication of adjuvant chemoradiotherapy.^{3,14,15}

Conclusion

Colonic schwannoma may have multiple presentation mimicking malignant tumor in such cases. Not treated in time, it can lead to severe complications that can compromise the vital prognosis, such as perforation and peritonitis. The mainstay treatment is complete surgical resection. The slow evolutionary genius of schwannoma, even when incompletely resected by necessity, confirms its good prognosis.

Declaration of Conflicting Interests

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Informed Consent

Written informed consent to publish this case was obtained

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