

Schwannoma of the rectum: A case report and literature review

Maddalena Zippi, Roberta Pica, Renzo Scialpi, Claudio Cassieri, Eleonora Veronica Avallone, Giuseppe Occhigrossi

Maddalena Zippi, Roberta Pica, Claudio Cassieri, Eleonora Veronica Avallone, Giuseppe Occhigrossi, Department of Gastroenterology and Digestive Endoscopy, Sandro Pertini Hospital, 00157 Rome, Italy

Renzo Scialpi, Department of Pathology, Sandro Pertini Hospital, 00157 Rome, Italy

Author contributions: Pica R made substantial contribution to study conception and design; Cassieri C and Scialpi R were involved in acquisition, analysis and interpretation of data; Zippi M and Avallone EV were involved in drafting the article and revising it critically for important intellectual content; Occhigrossi G gave final approval of the version to be published.

Correspondence to: Maddalena Zippi, MD, Department of Gastroenterology and Digestive Endoscopy, Sandro Pertini Hospital, Via dei Monti Tiburtini 385, 00157 Rome, Italy. maddyzip@yahoo.it

Telephone: +39-6-41735005 Fax: +39-6-41735005

Received: December 29, 2012 Revised: January 29, 2013

Accepted: February 8, 2013

Published online: April 16, 2013

dimensons. No recurrence of the lesion was observed after 6 mo of follow-up.

© 2013 Baishideng. All rights reserved.

Key words: Rectum; Schwannoma; Immunohistochemistry; S-100 protein; Treatment

Core tip: Schwannomas of the colon and rectum are tumors that are rarely detected. For achieving a definitive histopathological diagnosis, the use of an appropriate immunohistochemical panel is request. Although schwannomas are considered to be benign tumors, their risks of recurrence must be not ignored. The best therapeutic option is complete surgical removal. In the present study, we describe a case of a rectal schwannoma occurred in a 72-year-old man, presented as a small sessile polypoid lesion, which was successfully removed *in toto* by hot-biopsy, during the same procedure. No recurrence of the lesion was observed after 6 mo of follow-up.

Abstract

Schwannoma is a tumor originating from the Schwann cells. Gastrointestinal schwannomas are uncommon stromal tumors of the intestinal tract and, in particular, rectal schwannomas are extremely rare. In fact, it is well established that schwannomas appear more frequently in the stomach and in the small intestine, while location in the colon or in the rectum is uncommon. Reading the literature, only few cases of rectal schwannoma have been reported. Their diagnosis is confirmed by the immunohistochemical panel (S-100 protein). When these tumors are located in the colon and in the rectum, radical excision with wide margins is mandatory, due to their tendency to recur locally or become malignant, if left untreated. In the present study, we describe a case of a rectal schwannoma occurred in a 72-year-old man, presented as a small polypoid lesion, which was successfully removed *in toto* by hot-biopsy, during the same endoscopy, due to the

Zippi M, Pica R, Scialpi R, Cassieri C, Avallone EV, Occhigrossi G. Schwannoma of the rectum: A case report and literature review. *World J Clin Cases* 2013; 1(1): 49-51 Available from: URL: <http://www.wjgnet.com/2307-8960/full/v1/i1/49.htm> DOI: <http://dx.doi.org/10.12998/wjcc.v1.i1.49>

INTRODUCTION

Schwannomas are rare tumors derived from the cells of Schwann that form the neuronal sheath. Generally these are benign tumors, which, if left untreated, have the tendency to recur locally and become malignant^[1,2]. These lesions are classified in gastrointestinal autonomic nerve tumours (GANTs) and were described for the first time by Herrera *et al*^[3] in 1984. There are many synonyms for neurogenic tumors of the gastrointestinal tract, such as

schwannoma, neurinoma, neurofibroma, neurogenic fibroma, neurilemoma and plexiform neurofibromatosis. Usually, these different names are used with the intent of relating them to the structure from which they originate^[4]. The correct name of gastrointestinal schwannoma is based on the evidence showing that this neoplasm originates from Schwann cells in the neurons of the myenteric plexus^[5,6].

Although rare, schwannomas must be included in the differential diagnosis from other intestinal mesenchymal neoplasms, such as smooth muscle tumors, neurofibromas, and gastrointestinal stromal tumors (GISTs)^[7]. Primary schwannoma of the colon and rectum, not associated with systemic neurofibromatosis (von Recklinghausen disease), are extremely rare^[8].

CASE REPORT

A 72-year-old man was referred to our unit for performing a colonoscopy due to abdominal pain. The medical history of the patient included a type 2 diabetes mellitus and arterial hypertension. The therapy was assumed at the moment for these diseases. There was no family history of inflammatory bowel disease or cancer and no previous abdominal surgery. Physical examination of the patient revealed good general condition. Laboratory tests were normal. A colonoscopy disclosed evidence of a small sessile polypoid lesion, of about 1.5 cm in diameter, in the left lateral wall of the rectum, approximately at 1 cm from the superior anal margin. No other lesions were found in the other colon segments, including the cecum, except for a diverticular disease of the sigmoid colon. This lesion was removed *in toto* by hot-biopsy (Figure 1). The related histopathological findings showed typical features of schwannoma, with Antoni A/verocay bodies (cells forming a typical palisade arrangement in a well-organized pattern) and Antoni B (small lacunar foci with loss of palisade architecture; Figure 2A) and strong positivity for S-100 protein at immunohistochemical assay (Figure 2B). After 6 mo, a follow-up colonoscopy, performed with methylene blue staining, disclosed no evidence of recurrence of the lesion.

DISCUSSION

GANTs are uncommon stromal tumors accounting for 0.1% of benign tumors of the gastrointestinal tract^[5]. The most frequent site for GANTs include the stomach followed by duodenum, jejunum, ileum^[9], rarely they are located in the colon, and in the literature we have found only few reported cases of rectal schwannomas^[2,6,9-16]. Moreover, Gibson *et al.*^[17] have studied 26 colorectal sessile polyps containing S-100 positive neural proliferations in the lamina propria, not associated with type 1 neurofibromatosis (NF1). The authors proposed to define them as “mucosal Schwann cell hamartoma”, to avoid confusion with the neural lesions that have significant associations with inherited syndromes. In particular, schwannoma has the same incidence in men and women with a mean

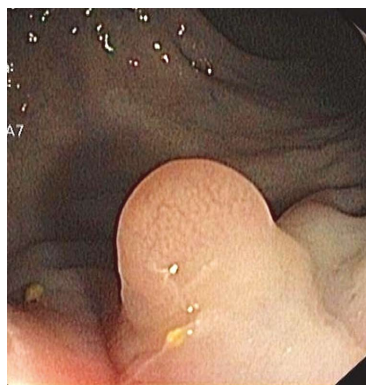


Figure 1 In case of a rectal schwannoma, the colonoscopy disclosed evidence of a little polypoid lesion in the left lateral wall of the rectum.

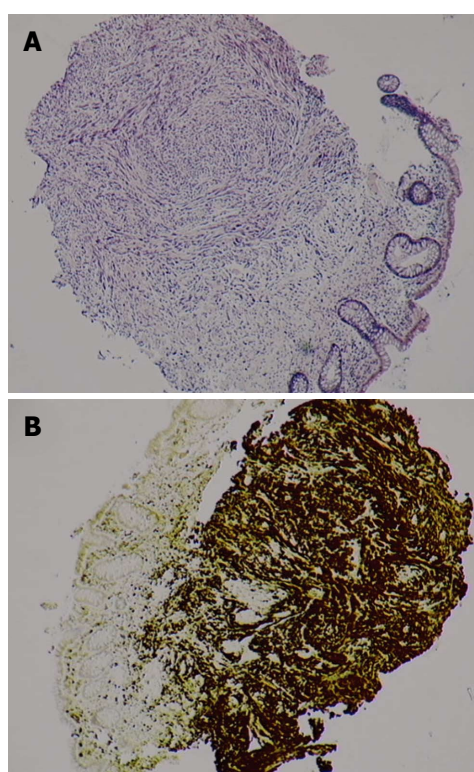


Figure 2 In case of rectal schwannoma, the tumor, located in the submucosa, has a nodular well-defined structure (haematoxylin and eosin, $\times 20$). The structure that consists in proliferation of Schwann-like cells, showing a palisading arrangement (A), being marked by S-100 protein (B).

age of 60-65 years^[18]. Their size may vary and a case of a rectal schwannoma of about 12 cm has been reported^[11]. Schwannomas are mostly asymptomatic or can present non-specific symptoms such as pain, fatigue and fever. Sometimes, rectal bleeding and signs of colonic obstruction may occur^[18].

Schwannomas should be differentiated from other intestinal mesenchymal neoplasms. In fact, the most accurate diagnosis is based on immunohistochemical test^[6]. Especially, it is difficult to distinguish schwannoma from leiomyoma and GIST. Generally, gastrointestinal schwannomas are not encapsulated, and this finding may

help to distinguishes them from schwannomas in peripheral nervous system. On histopathological examination these kind of tumors have a lymphoid cuff with germinal centre. They may resemble GISTs but the presence of lymphoid cuff helped in diagnosing of schwannomas. The tumor cells of schwannoma are positive for S-100 protein, as in our case, and negative for smooth muscle markers, such as actin and desmin, which are positive in myogenic tumors. Although these are considered as benign tumors, the risks of recurrence of local and of distant metastases are 30% and 2%, respectively^[19,20]. The mainstay management of these tumors is based prevalent on surgery with radical excision.

In conclusion, rectal schwannoma is a rare tumor with a benign behaviour and a good prognosis.

REFERENCES

- 1 **Catania G**, Puleo C, Cardì F, Catalano F, Iuppa A, Buffone A. Malignant schwannoma of the rectum: a clinical and pathological contribution. *Chir Ital* 2001; **53**: 873-877 [PMID: 11824066]
- 2 **Reinbold WD**, Hillemanns A, Seesko H, Jehn E. [Malignant schwannoma of the rectum]. *Radiologe* 1996; **36**: 663-666 [PMID: 8975284 DOI: 10.1007/s001170050125]
- 3 **Herrera GA**, Pinto de Moraes H, Grizzle WE, Han SG. Malignant small bowel neoplasm of enteric plexus derivation (plexosarcoma). Light and electron microscopic study confirming the origin of the neoplasm. *Dig Dis Sci* 1984; **29**: 275-284 [PMID: 6321118 DOI: 10.1007/BF01296263]
- 4 **Nonose R**, Lahan AY, Santos Valenciano J, Martinez CA. Schwannoma of the Colon. *Case Rep Gastroenterol* 2009; **3**: 293-299 [PMID: 21103244 DOI: 10.1159/000237736]
- 5 **Maciejewski A**, Lange D, Włoch J. Case report of schwannoma of the rectum—clinical and pathological contribution. *Med Sci Monit* 2000; **6**: 779-782 [PMID: 11208409]
- 6 **Miettinen M**, Shekitka KM, Sobin LH. Schwannomas in the colon and rectum: a clinicopathologic and immunohistochemical study of 20 cases. *Am J Surg Pathol* 2001; **25**: 846-855 [PMID: 11420455 DOI: 10.1097/0000478-200107000-00002]
- 7 **Vasilakaki T**, Skafida E, Arkoumani E, Grammatoglou X, Tsavari KK, Myoteri D, Mavromati E, Manoloudaki K, Zisis D. Synchronous primary adenocarcinoma and ancient schwannoma in the colon: an unusual case report. *Case Rep Oncol* 2012; **5**: 164-168 [PMID: 22666207 DOI: 10.1159/000337689]
- 8 **Genna M**, Leopardi F, Fambri P, Postorino A. [Neurogenic tumors of the ano-rectal region]. *Ann Ital Chir* 1997; **68**: 351-353; discussion 353-354 [PMID: 9454548]
- 9 **Stift A**, Friedl J, Gnant M, Herbst F, Jakesz R, Wenzl E. Gastrointestinal autonomic nerve tumors: a surgical point of view. *World J Gastroenterol* 2004; **10**: 2447-2451 [PMID: 15285041]
- 10 **Bhardwaj K**, Bal MS, Kumar P. Rectal schwannoma. *Indian J Gastroenterol* 2002; **21**: 116-117 [PMID: 12118926]
- 11 **Mulchandani MH**, Chattopadhyay D, Obafunwa JO, Joy-paul VB. Gastrointestinal autonomic nerve tumours—report of a case and review of literature. *World J Surg Oncol* 2005; **3**: 46 [PMID: 16026628 DOI: 10.1186/1477-7819-3-46]
- 12 **Aaronson SA**, Korner A, Munro AJ. Inhibition by soluble ribonucleic acid of stimulatory effect of liver template ribonucleic acid. *Biochem J* 1966; **101**: 448-453 [PMID: 5338167]
- 13 **Lee SH**, Kim TO, Hwang SY, Ryu DY, Lee DH, Park WI, Kim GH, Heo J, Kang DH, Song GA, Cho M. [A case of rectal schwannoma presenting with hematochezia]. *Korean J Gastroenterol* 2006; **48**: 195-199 [PMID: 17047435]
- 14 **Hsu KF**, Lin CT, Wu CC, Hsiao CW, Lee TY, Mai CM, Jin JS, Jao SW. Schwannoma of the rectum: report of a case and review of the literature. *Rev Esp Enferm Dig* 2010; **102**: 289-291 [PMID: 20486757]
- 15 **Tedeschi M**, Cuccia F, Angarano E, Piscitelli D, Gigante G, Altomare DF. Solitary schwannoma of the rectum mimicking rectal cancer. Report of a case and review of the literature. *Ann Ital Chir* 2011; **82**: 309-312 [PMID: 21834483]
- 16 **Yang X**, Zeng Y, Wang J. Hybrid schwannoma/perineurioma: report of 10 Chinese cases supporting a distinctive entity. *Int J Surg Pathol* 2013; **21**: 22-28 [PMID: 22832113]
- 17 **Gibson JA**, Hornick JL. Mucosal Schwann cell “hamartoma”: clinicopathologic study of 26 neural colorectal polyps distinct from neurofibromas and mucosal neuromas. *Am J Surg Pathol* 2009; **33**: 781-787 [PMID: 19065103 DOI: 10.1097/PAS.0b013e31818dd6ca]
- 18 **Miettinen M**, Sarlomo-Rikala M, Lasota J. Gastrointestinal stromal tumours. *Ann Chir Gynaecol* 1998; **87**: 278-281 [PMID: 9891765]
- 19 **Lauwers GY**, Erlandson RA, Casper ES, Brennan MF, Woodruff JM. Gastrointestinal autonomic nerve tumors. A clinicopathological, immunohistochemical, and ultrastructural study of 12 cases. *Am J Surg Pathol* 1993; **17**: 887-897 [PMID: 8394653 DOI: 10.1097/0000478-199309000-00004]
- 20 **Das Gupta TK**, Brasfield RD, Strong EW, Hajdu SI. Benign solitary Schwannomas (neurilemmomas). *Cancer* 1969; **24**: 355-366 [PMID: 5796779]

P- Reviewer Yan YT S- Editor Zhai HH
L- Editor A E- Editor Zheng XM

