CASE REPORT

Caecal schwannoma: a rare cause of per rectal bleeding in a 72-year-old man

Richard A Dickson-Lowe,¹ Christopher L James,² Charles MH Bailey,³ Yasser Abdulaal⁴

¹Department of General Surgery, Tunbridge Wells Hospital, Tunbridge Wells, UK

²Foundation Year 1, Tunbridge Wells Hospital, Tunbridge Wells, UK

³Department of Colorectal Surgery, Tunbridge Wells Hospital, Tunbridge Wells, UK

⁴Department of Emergency Surgery, Tunbridge Wells Hospital, Tunbridge Wells, UK

Correspondence to

Richard Alexander Dickson-Lowe, richarddickson-low@doctors.org.uk

Accepted 23 September 2014

SUMMARY

The presence of per rectal (PR) bleeding in elderly people usually alerts us to the possibility of colonic malignancy, particularly if a concomitant altered bowel habit is present, without a history of the symptom. We present a case where the cause of PR bleeding turned out to be a caecal schwannoma. Caecal schwannoma is a rare tumour of the gastrointestinal tract of mesenchymal origin and diagnosis prior to radical surgical resection is difficult. Endoscopic biopsy often fails to provide an adequate specimen and immunohistochemistry is essential to the diagnosis.

BACKGROUND

This case is of interest given the rarity of the eventual histopathological diagnosis of caecal schwannoma. Given that a schwannoma is a benign tumour that normally arises in intracranial (acoustic) nerves and spinal nerves, it is of interest to a variety of medical specialties including surgeons, gastroenterologists, neurologists, radiologists and histopathologists. Distinct characteristics are present following histology, but tissue biopsy is often difficult and differentiating these tumours from other gastrointestinal (GI) tumours radiologically is also tricky.

Schwannomas are benign, slow-growing mesenchymal neoplasms that arise from Schwann cells with a low malignant potential.¹ GI schwannoma represents 2–6% of all submucosal tumours of the GI tract, with the stomach being the most affected organ (60–70%).¹ Schwannomas of the colon are equally distributed between men and women and generally tend to appear in the sixth decade of life.²

CASE PRESENTATION

A 72-year-old man, previously fit and well, was admitted to hospital with a 12 h history of 'mild gripping pain' in his lower abdomen and three per rectal (PR) bleeds. There was no history of bleeds, change in bowel habit, upper GI symptoms or weight loss. There was no medical or family history and he was not a smoker or an alcohol drinker.

At first presentation he was clinically dry with generalised, mild abdominal pain with no palpable masses. His blood pressure and pulse were normal and stable. A PR examination showed a small amount of altered blood and clots on the glove, but no masses were palpable and he had a normal prostate. Rigid sigmoidoscopy did not reveal a source of bleeding. Blood tests were normal with a

haemoglobin (Hb) of 140 g/L. On the basis of these findings, an initial diagnosis of diverticulitis was made and a course of coamoxiclav was prescribed, as per trust guidelines, with a planned outpatient colonoscopy. He was in hospital for 24 h on this occasion.

Two weeks later, prior to his colonoscopy he had a second series of bleeds and another presentation to the hospital. Similar clinical findings were present with a normal blood pressure and pulse, but an Hb of 102 g/L, which decreased to 65 g/L over the next 8 h and required a three unit blood transfusion. A CT scan of the abdomen and pelvis showed a 5 cm×5 cm mass in the caecal pole. Given the ongoing PR bleeding without stability of Hb following transfusion, he was booked for an operation on the emergency list. A colonoscopy was performed in the operating theatre which revealed a smooth, solid mass in the caecum with ulceration (figure 1). This ulceration accounted for the PR bleeding and drop in Hb, therefore an open right hemicolectomy via a midline laparotomy followed.

The postoperative recovery was uneventful, a staging CT of the chest was clear and the patient was discharged home after 7 days. Two weeks later he was contacted to reveal the nature of his histology, which showed a caecal schwannoma. He had no further PR bleeding and a normal Hb.

INVESTIGATIONS

CT scan: The caecum was abnormal with a 5 cm×5 cm mass at the caecal pole with mildly enlarged nodes in the mesentery medially (figure 2). Of note, no small bowel dilation was

Figure 1 Colonoscopy photo showing an ulcerated 'mass' in the caecum.

CrossMark

To cite: Dickson-Lowe RA, James CL, Bailey CMH, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-205643

Figure 2 CT scan showing a tumour in the caecum.

seen and no free gas. There was no other lymphadenopathy and no metastasis.

Histopathology: Macroscopically much of the caecum is occupied by the polypoid mass as shown by the CT scan (figure 2). Microscopically, a large well-circumscribed spindle cell tumour is seen in the submucosal space (figure 3). Low power magnification shows spindle cell morphology and Verocay bodies (figure 4) and often Antoni A and Antoni B areas. High power magnification shows these Verocay bodies (nuclear palisading) with vascular hyalinisation (figure 5). The tumour cells are diffusely positive for S-100 (figure 6) and negative for GI stromal tumour (GIST) stains including CD117 and DOG1.

DIFFERENTIAL DIAGNOSIS

A man in his 70s with new onset altered blood PR and mild lower abdominal pain may have diverticulitis, colonic adenocarcinoma, colonic polyps, angiodysplasia or anal disease, for example, haemorrhoids. Other rarer differential diagnoses to be considered, which may also give the CT appearances shown, are GIST, GI autonomic neural tumour, leiomyoma, leiomyosarcoma, lymphoma and metastatic melanoma.

Figure 3 Microscopic histology with low power magnification, showing the spindle cell tumour.

Figure 4 Microscopic histology with low power magnification showing spindle cells and Verocay bodies.

TREATMENT

A right hemicolectomy was performed with a complete resection of the tumour. Staging revealed no evidence of metastasis and the resultant histopathology did not negate any further treatment.

OUTCOME AND FOLLOW-UP

Postoperative histology showed complete resection and benign pathology, therefore no further treatment was required. These tumours are noted to have a good prognosis.

DISCUSSION

Schwannomas, when present in the GI tract, are common in the stomach and are rarely found in the large intestine.³ In a review of 20 cases by Miettinen *et al*⁴ of the files of the Armed Forces Institute of Pathology, colorectal schwannomas were more frequently present in the caecum compared to the rectosigmoid junction. In comparison, a review of the Japanese literature of 46 cases by Inagawa *et al* reported the rectum as the commonest location for these tumours.² Clinical features common to many right-sided colonic/caecal tumours are GI bleeding from ulceration (which our patient had), colonic obstruction or abdominal pain.²

Diagnosis of schwannoma can be difficult as scans, for example, CT scan and MRI, only reveal well-defined, homogeneously attenuated masses arising from the colonic mucosa. There is often no evidence of a tumour capsule, cystic change,

Figure 5 Microscopic histology with high power magnification showing Verocay bodies.

Figure 6 Microscopic histology showing S-100+immunoreaction.

cavity formation, necrosis or calcification, which are well described and conversely common to GISTs.⁵

Schwannomas share the gross morphological features with other submucosal tumours and can often be difficult to biopsy due to their hardness. In a review of 41 cases of schwannomas of the large intestine in Japan performed by Tomozawa *et al*,⁶ only 10% were identified preoperatively. Endoscopically, GI schwannomas appear similar to GISTs and may often be misdiagnosed. Both types of tumours are located in the wall of the GI tract and exhibit similar growth patterns.⁵

Immunohistochemistry is the source of diagnosis in schwannoma. Histologically, GI schwannomas are different neoplasms from those that arise in the soft tissue or central nervous system and show distinctive patterns. They are strongly positive for vimentin, glial fibrillary acidic protein and S-100 protein and are negative for muscle cell markers CD34 and CD117 (KIT).⁷ There is a typical, prominent lymphoid cuff with or without germinal centre formation. The reason for the infiltration is not clear, but it has been suggested that it is a feature of a long-standing lesion or a peculiar host response. Most schwannomas consist of spindle cells but epithelioid and plexiform variants have been seen. They have Verocay bodies, vascular hyalinisation, Antoni A (cells forming a typical palisade arrangement in a well-organised pattern) and Antoni B (small lacuna foci with loss of palisade architecture) areas and a palisading structure, which are common features in a peripheral schwannoma.⁷

While immunohistochemistry provides the best information for diagnosis, ultrastructural studies may be extremely helpful. The ultrastructural criteria that suggest origin from the myenteric plexus are neuron-like cells with long cytoplasmic processes containing microtubules, bulbous synapse-like structures with dense core neurosecretory type granules and empty vesicles.⁸

It is important to correctly diagnose a GI schwannoma, given the malignant potential of other differential diagnoses, for example, GISTs. GISTs pursue an aggressive course in more than 50% cases and can often recur or metastasise; prognosis is influenced by size, mitotic rate, tumour site and completeness of resection.⁹ Schwannomas on the other hand are benign although they can recur. Rare cases of malignant gastric schwannoma have been reported and have poor prognosis, rapid clinical decline and a poor response to chemotherapy, with an average lifespan of 2 years.¹⁰

Management of these stromal tumours is by surgical resection. All published pathology series indicate that GI

schwannomas are benign and that patients have an excellent prognosis after successful resection.

Patient's perspective

The patient was spoken to at the time of 'breaking bad news' initially. At that time he was obviously worried about cancer. He described the wait for histology as difficult but overall was very happy that he had a successful operation without further bleeding and that the tumour was benign.

Learning points

- ▶ Per rectal bleeding is an important finding, particularly in certain (elderly) age groups and should alert any clinician to the possibility of colorectal malignancy.
- ▶ Caecal schwannomas are rare, benign, mesenchymal tumours. Schwannomas represent 2–6% of all submucosal tumours of the GI tract, with the stomach being the most affected organ (60–70%).
- ▶ Preoperative definitive diagnosis via imaging and endoscopic biopsy of caecal schwannoma is difficult.
- ▶ Surgical resection is the treatment of choice with diagnosis often only possible after this.
- ▶ Immunohistochemistry is paramount to diagnosis.

Acknowledgements The authors thank Dr Lamios Munthali (Consultant Histopathologist) and Dr Charles Wetton (Consultant Radiologist).

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Neto J, Leal R, Valente de Brito E, *et al*. Solitary schwannoma of the cecum: case report and review of the literature. *Case Rep Oncol* 2013;6:62–5.
- 2 Kanneganti K, Patel H, Niazi M, *et al*. Cecal schwannoma: a rare cause of gastrointestinal bleeding in a young woman with a review of literature. *Gastroenterol Res Pract* 2011;2011:142781.
- 3 Daimaru Y, Kido H, Hashimoto H, *et al*. Benign schwannoma of the gastrointestinal tract: a clinicopathologic and immunohistochemical study. *Hum Pathol* 1988;19:257–64.
- 4 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;25:846–55.
- 5 Levy AD, Quiles AM, Miettinen M, *et al*. Gastrointestinal schwannomas: CT features with clinicopathological correlation. *AJR Am J Roentgenol* 2005;184:797–802.
- 6 Tomozawa S, Masaki T, Matsuda K, *et al*. A schwannoma of the caecum: case report and review of Japanese schwannomas in the large intestine. *J Gastroenterol* 1998;33:872–5.
- 7 Hou YY, Tan YS, Xu JF, *et al*. Schwannoma of the GI tract: a clinicopathological, immunohistochemical and ultrastructural study of 33 cases. *Histopathology* 2006;48:536–45.
- 8 Mulchandani M, Chattopadhyay D, Obafunwa J, *et al*. Gastrointestinal autonomic nerve tumours—report of a case and review of literature. *World J Surg Oncol* 2005;3:46.
- 9 Miettinen M, Lasota J. Gastrointestinal stromal tumours: review on morphology, molecular pathology, prognosis and differential diagnosis. *Arch Pathol Lab Med* 2006;130:1466–78.
- 10 Wilde BK, Senger JL, Kanthan R. Gastrointestinal schwannoma: an unusual colonic lesion mimicking adenocarcinoma. *Can J Gastroenterol* 2010;24:233–6.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow