

This case illustrates a severe and debilitating consequence of acute intracerebral haemorrhage that has not previously been documented. It highlights that a successful resolution of OH can occur using a conservative approach supported by physiotherapy with the emphasis on graduated postural methods of improving cardiovascular conditioning.

Key points

- OH following acute intracerebral haemorrhage is rarely documented.
 - Screen for OH in all stroke patients before starting physiotherapy in the upright position.
 - Exclude remedial causes of OH.
 - Conservative management and physical methods can effect resolution of OH in this setting.
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References

1. Korpelainen JT, Sotaniemi KA, Suominen K, Tolonen U, Myllylä VV. Cardiovascular autonomic reflexes in brain infarction. *Stroke* 1994; 25: 787–92.
2. Panayiotou B, Reid J, Fotherby M, Crome P. Orthostatic haemodynamic responses in acute stroke. *Postgrad Med J* 1999; 75: 213–18.
3. Carlsson A, Britton M. Blood pressure after acute stroke—a one-year follow-up study. *Stroke* 1996; 27: 247–51.
4. Kong KH, Chuo AM. Incidence and outcome of orthostatic hypotension in stroke patients undergoing rehabilitation. *Arch Phys Med Rehabil* 2003; 84: 559–62.
5. Robbins AS, Rubenstein LZ. Postural hypotension in the elderly. *J Am Geriatr Soc* 1984; 32: 769–74.

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Spindle cell sarcoma: a rare cause of a large abdominal mass

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Abstract

A 73-year-old man presented with a huge intra-abdominal mass. Initially a gastrointestinal stromal tumour (GIST) was diagnosed, but his tumour was subsequently classified as a spindle cell sarcoma. Difficulties in the classification of rare intra-abdominal tumours are discussed.

Keywords: spindle cell sarcoma, gastrointestinal stromal tumours (GIST), elderly

Case Report

A previously fit 73-year-old man was referred complaining of backache, abdominal discomfort and possible weight loss. A huge, solid, non-tender and non-pulsatile intra-abdominal mass, the size of a 28-week uterus, was found. Ultrasound examination showed an inhomogeneous midline mass containing small cysts. Abdominal CT confirmed the mass, with solid and cystic spaces within it (see Figure 1). No origin was identified but the liver, spleen, pancreas and

adrenals appeared normal. Ultrasound-guided biopsy revealed necrotic tumour tissue, which stained positively with antibodies to CD34, vimentin and smooth muscle actin, but was negative for melanoma, mesothelioma, epithelial markers, and with antibodies to CD117 (also called KIT). A provisional diagnosis of gastrointestinal stromal tumour (GIST) was made and it was thought likely to be malignant owing to its large size and central necrosis. The tumour was judged to be surgically resectable. At laparotomy, the tumour was found to arise from within two leaves of the small bowel mesentery and involved a segment of the



Figure 1. CT of the tumour showing central necrosis and cystic spaces.

ileum. The tumour was resected and the patient was discharged home after 9 days. He remains well with no evidence of recurrence 12 months post-surgery.

The tumour weighed in excess of 5 kg and measured 28×25×16 cm. The spindle-shaped tumour cells were strongly positive with CD34 and S100, weakly positive with smooth muscle actin, but negative for CD117 (KIT). A second histology opinion was sought. The tumour was diagnosed as an unclassifiable spindle cell sarcoma.

Discussion

Tumours arising in the small bowel may present with haemorrhage or obstruction, the latter due to mass effect or intussusception. However, many are asymptomatic and, extramural lesions especially, may reach enormous sizes.

This tumour proved difficult to classify but was finally labelled as a spindle cell sarcoma, an extremely rare tumour, which is an important differential diagnosis of GIST, GI schwannoma and true smooth muscle tumour [1].

Diagnosing very rare tumours can be difficult, and this case highlights the confusion regarding the exact definition of GISTs. Previously, GISTs were classified as visceral leiomyomas or leiomyosarcomas, but are now distinguished from true smooth muscle tumours by the presence of KIT staining. One recent view is that GISTs are an immunohistochemical diagnosis, i.e. GI mesenchymal tumours always expressing the CD117 antigen, a membrane receptor with a tyrosine kinase component [2]. This definition would exclude this tumour from being a GIST as it was CD117 negative. However, it did express CD34, expressed in 70–80% of GISTs [3].

A more traditional definition of GIST is a GI mesenchymal tumour arising in the muscularis propria of the intestinal wall, subgrouped into spindle cell GISTs (70%) and epithelioid GISTs (30%). This tumour was made up of closely packed spindle cells; however, it was arising from the mesentery of the small bowel, not the submucosa [4]. Mutation of the *CD117* gene leads to activation of the CD117

protein, which promotes anti-apoptotic signalling and cell proliferation. GISTs have been found in extra-GI locations in the mesentery, retroperitoneum and in the gallbladder and urinary bladder. Demonstration of KIT expression has helped to validate their existence [5, 6]. More recently, KIT-negative GISTs have been described, most of which show platelet-derived growth factor mutations as an alternative oncogenic mechanism [7]. Approximately 4% of GISTs belong to this subset. Whether these tumours are indeed GISTs and whether they may respond to KIT inhibitor therapy (imatinib) is controversial. KIT-negative tumours exhibit classic histological features of GIST, being composed of cellular sheets or nests that lack significant nuclear pleomorphism, unlike our patient's tumour, which was therefore labelled as an unclassifiable spindle cell sarcoma.

Treatment for these tumours is principally surgical resection, if feasible, as they are usually resistant to radiotherapy and chemotherapy. Long-term survival is not seen with unresectable lesions. Lifelong follow-up with serial CT scanning is suggested for such large tumours. This case demonstrates the late presentation of small bowel tumours and highlights the lack of consensus on the definition of GISTs.

Key points

- Extramural small bowel tumours may reach large sizes prior to presentation.
- Classification of tumours can be difficult and this can have implications for the use of adjuvant therapy.

References

1. Miettinen M, Lasota J. Gastrointestinal stromal tumours (GISTs): definition, occurrence, pathology, differential diagnosis and molecular genetics. *Pol J Pathol* 2003; 54: 3–24.
2. Miettinen M, Sobin LH, Sarlomo-Rikala M. Immunohistochemical spectrum of GISTs at different sites and their differential diagnosis with a reference to CD117 (KIT). *Mod Pathol* 2000; 13: 1134–42.
3. Pithorecky I, Cheney RT, Kraybill WG, Gibbs JF. Gastrointestinal stromal tumours: current diagnosis, biological behaviour and management. *Ann Surg Oncol* 2000; 7: 705–12.
4. Miettinen M, Lasota J. Gastrointestinal stromal tumours – definition, clinical, histological, immunohistochemical and molecular genetic features and differential diagnosis. *Virchows Arch* 2001; 428: 1–12.
5. Reith JD, Goldblum JR, Lytes RH *et al.* Extra gastrointestinal (soft tissue) stromal tumours: an analysis of 48 cases with emphasis on histologic predictors of outcome. *Mod Pathol* 2000; 13: 577–85.
6. Lasota J, Carlson JA, Miettinen M. Spindle cell tumour of urinary bladder serosa with phenotypic and genotypic features of gastrointestinal stromal tumour. *Arch Pathol Lab Med* 2000; 124: 894–7.
7. Medeiros F, Corless CL, Duensing A *et al.* KIT-negative gastrointestinal stromal tumors. Proof of concept and therapeutic implications. *Am J Surg Pathol* 2004; 28: 889–94.

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