

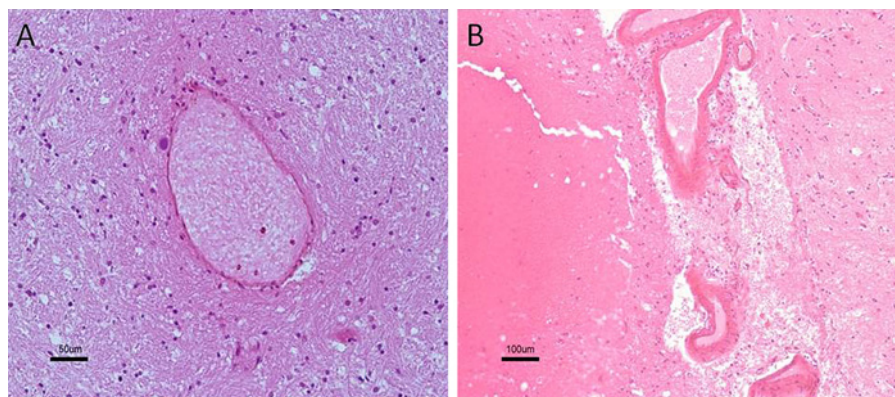


Figure 2 Histology demonstrating (A) bloody effusion around small arteries and (B) diffuse effusion of erythrocytes and haemorrhagic exudate in the periphery of the thalamic haemorrhage. H&E staining.

reports in the 1960s.^{4–8} Post and Desforges reported that ethanol infusion leads to a decrease in the platelet count, resulting in swelling and vacuolation.⁸ The authors postulated that alcohol may cause a metabolic alteration in the platelet membrane, leading to a decreased life span and diffuse early sequestration in the vascular system. Since then, no other theories have been presented in the literature.

In our case, the thrombocytopenia and hypertension on admission may have resulted in the rapid progression of the TH and subdural and subarachnoid haemorrhages (figure 1A). After surgical decompression, the midline shift by brain swelling was relieved. However, a diffuse hypodensity in the right hemisphere and the left subdural, thalamic and midbrain haemorrhages were enlarged (figure 1B). The histological findings showed oedema as the cause of the hypodensity, but neglected ischaemia due to thrombus, traumatic stenosis or occlusion in the right carotid or an artery in the right circle of the Willis ring.

Histology demonstrated bloody effusion around small arteries in the periphery of the

TH (figure 2A,B). The red neurons around the TH suggested ischaemia, which may have been caused by compression of the thalamic arteries or the perforating branches of the posterior cerebral artery by brain swelling or tentorial hernia. We estimated that decompression surgery may relieve the ischaemia, thereby inducing bilateral TH through reperfusion. The TH had a round shape, in contrast to the irregularly demarcated contusions in the frontal cortex and cerebellum, and was accompanied by petechiae. The TH was detected on the right on admission and became enlarged bilaterally post-surgery. Collectively, in our case, the decompression-related reperfusion, hypertension and thrombocytopenia were likely to have caused the bilateral TH.

CONCLUSION

AIT, hypertension and reperfusion after decompression may cause the rapid progression of bilateral THs, and possibly subdural and subarachnoid haemorrhages, induced by violence.

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Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Published Online First 26 May 2011

J Clin Pathol 2011;**64**:1034–1035.
doi:10.1136/jclinpath-2011-200023

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Mast cell sarcoma of the small intestine: a case report

Mast cell sarcoma is an extremely rare mast cell proliferative disorder characterised by localised tumour, destructive growth and poor prognosis. Here we describe a case of mast cell sarcoma arising in the small intestine. To the best of our knowledge, so far only four cases have been reported in literature, presenting in the larynx, ascending colon, cerebrum and tibia.^{1–5}

CASE REPORT

An elderly man presented with a 1.5-month history of abdominal pain. On examination, there was no lymphadenopathy or hepatosplenomegaly. Peripheral blood counts were normal except for mild anaemia. Serum lactate dehydrogenase was normal. CT abdomen showed a circumferential thickening of the wall of the ileum, with dilatation of bowel loops and a few enlarged mesenteric lymph nodes.

The patient underwent ileal resection and anastomosis. Gross examination revealed a segment of the small intestine measuring 15 cm in length, 4 and 8 cm in circumference at either resection margin. The external surface showed a constriction about 3 cm in length. The lumen showed an annular constrictive tumour measuring 4.5×4×1.5 cm, located 5 cm from wider resection margin, with a light brown homogeneous and firm cut surface. The tumour appeared to involve the muscle coat and was 0.1 cm from the nearest serosal surface (figure 1). Microscopically, sections of small intestinal wall showed focal ulceration of the mucosa and transmural infiltration by the tumour. The tumour was composed of a diffuse infiltrate of large immature cells having round to oval, lobated pale-staining nuclei and moderate amounts of eosinophilic to clear cytoplasm. Several eosinophils were present scattered between the tumour cells (figure 2). Focally, Peyer's patches were observed in the

Take-home messages

- This is the first report on the bilateral thalamic hemorrhage related to trauma and surgery, and the first autopsy report on the death associated with alcohol-induced thrombocytopenia.
- After emergency surgery for the super-acute subdural hematoma, bilateral thalamic hemorrhage developed due to ischemia-reperfusion after the decompression.
- The causality of the intrinsic thrombocytopenia and violence (blow) to the lethal subdural hematoma and the thalamic hemorrhage was important in legal judgement.



Figure 1 Small intestinal resection showing an annular constrictive tumour.

submucosa of the intestinal wall. The tumour cells showed metachromatic staining of the cytoplasm with toluidine blue (figure 3). On immunohistochemistry, the atypical mast cells were positive for tryptase, CD117 (figure 4), CD45 and CD68 (KP1) and were negative for MPO, CD20, CD3, CD30, S100 and VS38. There was no morphological involvement of the bone marrow. Mutation analysis on peripheral blood was done by RT-PCR, which showed c-KIT mutation Asn-822-Lys in exon 17 while no mutation was detected in exon 8. The patient was given prednisolone 40 mg once daily for a month and the dosage was then tapered with imatinib 400 mg once daily; subsequently, he was only on imatinib for another 2 months. His symptoms improved remarkably in a month; however, at the end of 3 months, he developed exudative ascites and was started on interferon α -2b three million units twice a week. More intensive systemic therapy could not be given because of the frail general condition and advanced age. CT of the abdomen done after 6 months of interferon therapy showed resolution of the disease.

DISCUSSION

A diagnosis of mast cell sarcoma was made based on histopathological findings and ancillary studies. This patient did not fit into the criteria of systemic mastocytosis as the lesion was localised to the small intestine only, without morphological involvement of the bone marrow. We believe that the lesion probably arose in an intestinal mast cell

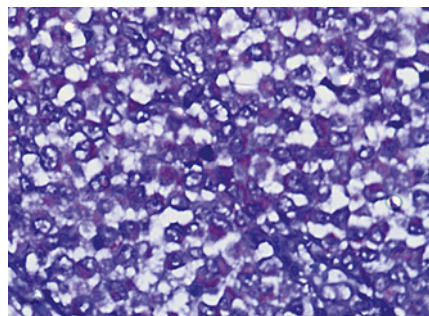


Figure 3 High-power view demonstrating metachromatic staining of neoplastic cells (toluidine blue; original magnification $\times 400$).

probably within a Peyer's patch and then mutant cells spread systemically.

This is the first case of mast cell sarcoma described in the small intestine; the only other case of gastrointestinal mast cell sarcoma is reported by Kojima *et al* in the ascending colon.³

The c-KIT mutation Asp-816-Val is detectable in most patients with systemic mastocytosis and in some cases of mast cell leukaemia.⁶ The study by Chott *et al* showed the lack of this mutation in a case of intracranial mast cell sarcoma.⁵ In our case, the c-KIT mutation detected was Asn-822-Lys. Other case studies on mast cell sarcoma do not report the status of c-KIT mutations. Whether c-KIT mutations other than Asp-816-Val are more common in mast cell sarcomas in contrast to systemic mastocytosis remains to be investigated and it is difficult to reach a conclusion based on the very few cases reported so far.

There is no established treatment protocol for mast cell sarcoma in view of the rarity of the condition. So far, the reported cases have been treated with local resection and radiotherapy with additional systemic therapy if the disease is refractory or in case of recurrence. The prognosis has been dismal in all the reported cases except two where there has been stable response to treatment for some time.²⁻³ An acquired KIT D816V mutation confers full resistance to imatinib, while the N822K mutation retains susceptibility to imatinib.⁷⁻⁸ Since this patient had

Take-home messages

- Mast-cell sarcomas can originate in small intestine.
- c-KIT mutation spectrum in mast-cell lesions needs to be expanded further.
- Optimal therapy for mast cell sarcoma including targeted therapies needs to be defined by further studies.

already undergone complete resection of the tumour and DNA sequencing analysis showed KIT N822K mutation, he was started on imatinib. However, the response was short-lived. The disease appears to be stable 8 months after starting interferon. The role of tyrosine kinase inhibitors and individual chemotherapeutic agents in mast cell sarcoma needs to be defined by use in larger number of patients.

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Competing interests None.

Contributors All the authors listed in the paper have contributed significantly to the design and contents of this article.

Provenance and peer review Not commissioned; externally peer reviewed.

Published Online First 21 July 2011

J Clin Pathol 2011;**64**:1035–1037.
doi:10.1136/jclinpath-2011-200154

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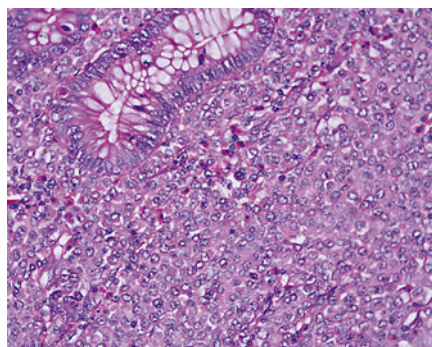


Figure 2 High-power view showing diffuse infiltration of intestinal wall by atypical mast cells (—H&E; original magnification $\times 400$).

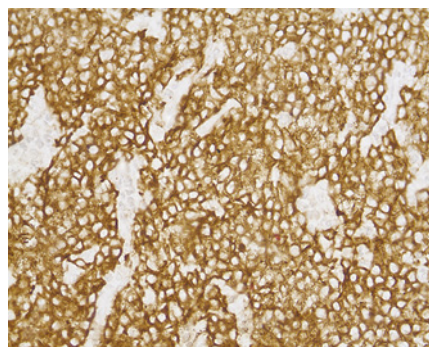


Figure 4 Membrane positivity of neoplastic cells by CD117 (original magnification $\times 400$).

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LETTER TO THE EDITOR

The attitudes of patients to their own autopsy: a misconception

We read with interest the report by Tsitsikas and colleagues about the attitudes of relatives to autopsy in a division of lymphoid malignancies.¹ Their report shows that the attitude of the general public is positive overall and translates into high autopsy rates when the value of the examination is presented honestly and the details of the procedure are adequately explained.

We would like to add to this interesting report our previous experience from a prospective opinion survey about the consent of patients for their own autopsy.² Interestingly, Tsitsikas and colleagues had an 89% consent rate to autopsy when asking relatives; close to their findings, when we used a questionnaire, 86% of the individuals and 94.6% of health professionals were not opposed to their own autopsy.²

The reasons for opposition encountered by Tsitsikas and colleagues were: one relative found autopsies to be undignifying and another family refused to encounter them.¹ When asking patients, religious belief, body integrity, dignity and respect were the main reasons for refusal.²

Our work also showed that only 42.5% patients had ever received information about non-coronial autopsies. Surprisingly, only 33.9% of healthcare professional had previously received specific information about medical autopsies.² Furthermore, the refusal rate is lower in our study (8.9% vs 6.9%) where a physician explains the autopsy than by giving only a questionnaire to patients. This highlights the efficiency of personalised information.

The rate of autopsy refusal by relatives depends on the country, but is similar in these two studies compared with previously published works.³

A decline in medical autopsy rates has been encountered in most countries.⁴ Based on the data mentioned above, we suggest that if information about autopsies is provided to the public and specific information is provided to healthcare professionals, the autopsy rates would be higher.

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Competing interests None.

Patient consent Obtained.

Contributors FF: wrote the letter, managed data collection and performed data analysis; SD: monitored data collection, revised the draft paper and initiated the collaborative project; MP: monitored data collection, revised the draft paper and initiated the collaborative project.

Provenance and peer review Not commissioned; internally peer reviewed.

Accepted 8 June 2011

Published Online First 29 June 2011

J Clin Pathol 2011;**64**:1037.

doi:10.1136/jclinpath-2011-200236

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WITHDRAWN

doi:10.1136/jclinpath-2011-200046

Yamaguchi R, Mitsuyama S, Tanaka M, *et al.* Practical considerations in the detection of HER2 status: the pathological perspective. *J Clin Pathol* 2011. Published Online First 21 September 2011. doi:10.1136/jclinpath-2011-200046. This article has been withdrawn.



Mast cell sarcoma of the small intestine: a case report

Amit Bugalia, Aby Abraham, Poonkuzhali Balasubramanian, et al.

J Clin Pathol 2011 64: 1035-1037 originally published online July 21, 2011

doi: 10.1136/jclinpath-2011-200154

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