SHORT REPORT

Spontaneous haematomyelia: a necropsy study

Richard W Leech, Jan V Pitha, Roger A Brumback

Abstract

Spontaneous haematomyelia (intramedullary spinal haematoma), is an uncommon event. Predisposing conditions have been reported including syringomyelia, pregnancy and delivery, angioma, spinal artery aneurysm, and haemophilia, but only rarely has a pathological evaluation been performed. Two such cases studied at necropsy are reported. In one case, the haematoma was restricted to the cervical spinal cord, while in the second case it extended from the medulla into the lowest thoracic cord segments. In both cases the haematomyelia was fatal. In the first case the clinical course was subacute, but in the other the course was more acute. Careful neuropathological examination showed no apparent cause for the haemorrhages.

Haematomas of the spinal canal are uncommon events, but those reported are generally associated with some precipitating event such as trauma, vascular malformation, bleeding diathesis, or syringomyelia. Spontaneous, non-traumatic, haemorrhages without an apparent underlying aetiology are rare. We report two examples of such spontaneous spinal cord haemorrhages in which careful necropsy examination could not show an obvious cause other than generalised atherosclerosis.

Case reports*Case 1*

This 72 year old man, who was generally in good health, was transferred to the hospital for evaluation of a recent myocardial infarct and spinal cord infarct secondary to a possible embolus. Clinical evaluation, including laboratory and other appropriate studies, was consistent with a recent anterior myocardial infarct. He entered the referring hospital with chest pain, and three days after admission began to complain of numbness and weakness in his lower extremities. Four days later this progressed to profound weakness in both lower extremities, more on the left side than the right, with bilateral extensor plantar responses (Babinski reflexes). While in the referring hospital he had received a mild jolt to the back, and therapy had included anticoagulation which was continued after

transfer. There was a T6 sensory level on the left, and a T11 sensory level on the right. Myelography was negative.

Two weeks after transfer he experienced abdominal distention and diarrhoea associated with a sensation of a band about his upper abdomen. The following day, he had an exploratory laparotomy and a perforated sigmoid diverticulum was repaired. He subsequently experienced a cardiac arrest associated with electrocardiographic evidence of right bundle branch block and ventricular fibrillation. Although successfully resuscitated, he died two weeks later after repeated episodes of bradycardia.

Necropsy confirmed the successful repair of the ruptured diverticulum and documented the presence of generalised severe atherosclerosis involving essentially all arteries without occlusion. The heart weighed 600 gm with 70% stenosis of the left anterior descending coronary artery, and 40% stenosis of the right coronary and left circumflex coronary arteries. A recent, two to three week, myocardial infarction was demonstrated. The brain was unremarkable. Serial cross sections of the spinal cord demonstrated an intramedullary haematoma which was largest at T4, and extended two segments rostrally and caudally (fig A). Microscopic examination of serial step sections throughout all levels of the haematoma as well as levels above and below revealed no evidence of arterial occlusion, embolus, aneurysm formation, or other possible precipitating aetiological factors.

Case 2

This 59 year old man presented with the acute onset of quadriplegia, dysphagia, and respiratory distress. CT scans of the head and cervical spine were unremarkable, as were plain radiographs of the spine. The patient remained alert and awake, but required artificial ventilation. His hospital course was complicated by thermoregulatory hyperthermia (which resolved with a cooling blanket) and progressive hypotension. The patient became lethargic but the cause for this was never determined. Blood cultures were negative. He developed renal failure, became increasingly hypotensive, and died on day 17.

At necropsy there was pulmonary oedema, severe anasarca, and slight interstitial pancreatitis. There was cardiomegaly, the heart weighing 595 gm with coronary arteries demonstrating slight to moderate atherosclerosis with 40% stenosis of the left anterior

Department of Pathology, University of Oklahoma College of Medicine, Oklahoma City
R W Leech
J V Pitha*
R A Brumback*
Laboratory Services,* Veterans Affairs Medical Center, Oklahoma City, Oklahoma

Correspondence to:
Dr Leech,
Department of Pathology,
University of Oklahoma,
College of Medicine,
BSMB 451, PO Box 26901,
Oklahoma City, OK 73190,
USA

Received 23 October 1989
and in revised form
28 July 1990.
Accepted 23 April 1990

Figure A Lower level of the cervical spinal cord in case 1 demonstrating the central haemorrhage with surrounding degeneration of the peripheral white matter.

Figure B Case 2. Multiple cross sections of the spinal cord showing the extensive intramedullary haemorrhage destroying much of the cord substance.

descending coronary artery. External examination of the spinal cord showed that the upper cervical cord was expanded, with brownish-red blood oozing from the cephalad cut surface. Serial cross sections revealed an intramedullary haematoma extending from the caudal floor of the fourth ventricle through the distal most thoracic segment (fig B). Serial step sections revealed no origin of the haemorrhage, other than severe arteriosclerosis. Aneurysm formation was not detected.

Discussion

In neither case was a precipitating cause of the haemorrhage found at necropsy other than the known presence of arteriosclerosis. The negative CT scan in case 2 can only be explained by the haemorrhage beginning at a level lower than inspected, expanding throughout the length of the spinal cord as demonstrated. In case 1, contribution of anticoagulation therapy to the origin of the haemorrhage is unlikely as the patient was treated conservatively. In both cases, bleeding diatheses, arteritis, berry aneurysm formation, and other well known causes of haemorrhage can be excluded on the basis of the history, laboratory results, and the pathological examination.

According to Jellinger,¹ intramedullary spinal cord haemorrhage was first reported in

1814, with the term haematomyelia being introduced in 1827. Haematomyelia or haemorrhage into the spinal cord is a rare event when compared with intracranial haemorrhages, and even less commonly demonstrated or studied. In our two cases the precipitating cause appeared to be arteriosclerosis only, and possibly complicated by anticoagulation in case 1.

Intramedullary spinal cord haemorrhages show a predilection for the grey matter and tend to extend longitudinally, as demonstrated in both of our cases. Depending on the level of the lesion, the patient may present with paraplegia or quadriplegia, bowel or bladder dysfunction, and sensory disturbance involving all modalities. Clinically there is usually a rapidly evolving myelopathy, although Licata *et al*² have reported a gradual and lengthy clinical course in three cases of intramedullary haemorrhages. Reported aetiologies for cases of intramedullary haemorrhages has varied widely. Trauma may be the most common cause, although undoubtedly overemphasised.¹⁻³

Haemorrhages originating within the spinal column may be localised within the cord substance or subarachnoid, subdural, or epidural spaces. Spinal subarachnoid haemorrhage has been well-documented, but is clearly less often recognised than its intracranial counterpart.¹⁴ Causes include intradural arteriovenous malformations, and tumours. Rarely, haemorrhages are of unknown aetiology.⁴ Spinal epidural haemorrhages are also uncommon but well documented, with aetiologies including trauma and bleeding diatheses. Those spinal epidural haemorrhages without identifiable aetiology are even more uncommon.^{5,6} Spinal subdural haematomas may be exceedingly rare,¹ but have been reported in patients with bleeding diathesis.⁷

Primary non-traumatic intramedullary haemorrhages in the cord are very uncommon in the absence of predisposing conditions such as syringomyelia,¹⁸ pregnancy and delivery,⁹ angioma,¹⁰ aneurysm,¹¹ and haemophilia.¹² Most cases have been clinically identified. However, an angioma was demonstrated at necropsy in the case of Clark,¹⁰ and an aneurysm of the dorsal spinal artery in the case of Bräutigam.¹¹ A review of current neurology and neuropathology texts has not revealed necropsy confirmed cases.^{13,14} In our cases multiple sections showed no evidence of underlying vascular malformations or tumours, and clinically there were no precipitating factors.

Both of our patients were older men. In the first patient there was strong evidence of generalised atherosclerosis, without occlusion involving all major arteries. Atherosclerosis was much less evident in case 2. In the brain, rupture of an atherosclerotic miliary aneurysm can obliterate the aneurysm precluding its histological identification at necropsy and we assume that a similar event could occur in the spinal cord and this may explain our cases.

Our cases represent examples of spontaneous haematomyelia without proven cause, one with

a subacute course and the other with a rapidly evolving myelopathy. These two cases demonstrate what has characterised both clinically and pathologically those few previously reported cases of spontaneous haematomyelia. If suspected clinically, spinal canal haemorrhages now can be readily demonstrated during life by MRI and CT scans, making therapeutic intervention a possibility.^{2 6 7}

- 1 Jellinger K. Traumatic vascular disease of the spinal cord. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology, Vol 12. Vascular diseases of the nervous system. Part II*. Amsterdam: American Elsevier Publishing, 1975:556-630.
- 2 Licata C, Zoppetti MC, Perini SS, Bazzan A, Gerosa M, Da Pian R. Spontaneous spinal haematomas. *Acta Neurochir* 1988;95:126-30.
- 3 Koos WT, Böck F. Spontaneous multiple intramedullary hemorrhages. *J Neurosurg* 1970;32:581-4.
- 4 Arias MJ. Acute spinal cord compression by spontaneous subarachnoid haematoma. *Neurochirurgia* 1986;29:201-2.
- 5 Douglas AS, McAlpine SG. Neurological complications of haemophilia and Christmas disease. *Scot Med* 1956;1:270-3.
- 6 Mustafa MH, Gallino R. Spontaneous spinal epidural hematoma causing cord compression after streptokinase and heparin therapy for acute coronary artery occlusion. *South Med J* 1988;81:1202-3.
- 7 Gabl M, Kostron H. Acute spinal subdural haematoma. *Neurochirurgia* 1988;31:99-100.
- 8 Perot P, Feindel W, Lloyd-Smith D. Hematomyelia as a complication of syringomyelia: Gower's syringal hemorrhage. *J Neurosurg* 1966;25:447-51.
- 9 King A. Neurologic conditions occurring as complications of pregnancy. *Arch Neurol Psychiatry* 1950;63:471-99.
- 10 Clark JMP. Traumatic haematomyelia from a rupture of intramedullary angioma. *J Bone Joint Surg* 1954;36:418-22.
- 11 Bräutigam W. Über eine spontane Hämatomyelie durch ruptur eines durch angeborene gefäßwandschwäche entstandenen Aneurysmas der Arteria Spinalis Dorsalis. *Deutsche Zeitschrift f. Nervenheilkunde* 1960;181:119-29.
- 12 Aggeler PM, Lucia SP. The neurologic complications of hemophilia. *J Nerv Ment Dis* 1944;99:475-500.
- 13 Adams JH, Corsellis JAN, Duchen LW, eds. *Greenfield's neuropathology, 4th ed*. New York: John Wiley, 1984.
- 14 Adams RD, Victor M. *Principles of neurology, 3rd ed*. New York: McGraw-Hill, 1985.