

Spontaneous Hematomyelia in a Patient with Hemophilia A: A Case Report

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Abstract : Intraspinal hemorrhage is very rare and intramedullary hemorrhage, also called “hematomyelia” is the rarest form of intraspinal hemorrhage. There are only few reports in medical literature. We report a case of 34 year old male having haemophilia A presented with numbness and weakness of both lower extremities and acute retention of urine resulting from thoracic hematomyelia. The patient showed gradual improvement on medical management with cryoprecipitate infusions. High degree of suspicion is required for early diagnosis and management of this rarely reported spinal hematomyelia in patients with hemophilia.

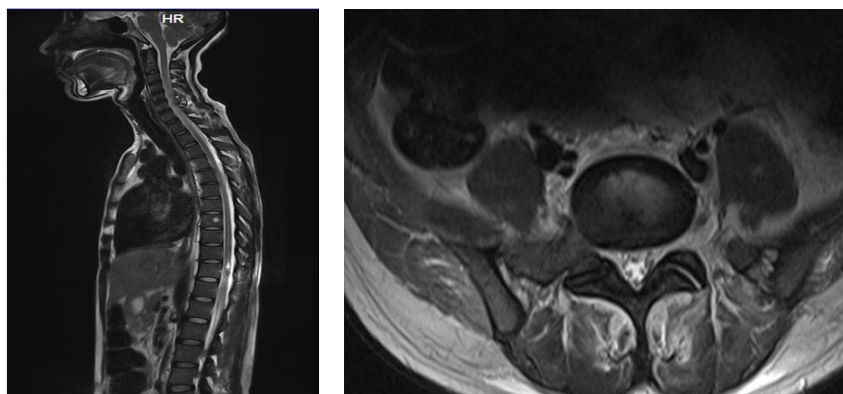
Keywords: Intramedullary spinal cord hemorrhage, Hematomyelia, Spinal cord, Spontaneous.

I. Introduction

Intramedullary spinal cord hemorrhage (hematomyelia) is an uncommon cause of myelopathy and can present in an acute, subacute, or chronic fashion¹. There are very few reports of hematomyelia in the medical literature. Anticoagulation therapy, trauma, arteriovenous malformations, tumors, and hemorrhagic diathesis are the possible predisposing factors². From the first report of “spontaneous hematomyelia,” published by Richardson⁷ in 1938, some cases have been reported but its incidence has not been established due to paucity of this disorder. Hematomyelia in patients with hemophilia have been rarely reported in literature. We are presenting a case of 34-year-old male having haemophilia A presented with lower limb weakness due to thoracic hematomyelia. Medical management showed gradual improvement.

II. Case Report

A 34-year-old man having haemophilia A visited our emergency department with complaints of weakness and sensory disturbances in both lower extremities, voiding difficulties which had started three days prior to his visit. An initial neurological examination revealed grade I motor power for the right and grade II motor power for left lower extremity, and he was unable to walk. Sensation was decreased below the T10 dermatome. Sensation to touch, pin prick and vibration were impaired up to T10 level on both sides. Her anal tone was decreased, and voiding was not possible. A trauma history was denied. His CBC and serum electrolytes were normal. Blood sugar was 124 mg/dl. Urine analysis was normal. Factor VIII level was <1%, PT was normal and aPTT was prolonged. MRI of dorsolumbar spine showed relatively well defined Intramedullary lesion seen in spinal cord from T10-T11 up to T12-L1 vertebral level, which was shown as hyperintense signal on T2W, STIR images and Hypointense on T1W images which likely represents sub-acute cord hematoma. Patient was managed conservatively with replacement therapy with cryoprecipitate infusions. Patient improved gradually and got grade III power in both lower limbs and discharged on 20th day.



(MRI Dorsolumbar Spine T1W and T2W images: showing thoracic hematomyelia)

III. Discussion

The intramedullary spinal cord haemorrhage, also called hematomyelia, is the rarest form of intraspinal haemorrhage³⁻⁶. It is usually related to trauma. Spinal vascular malformations such intradural arteriovenous malformations are the most common cause of atraumatic hematomyelia. Other considerations include warfarin or heparin anticoagulation therapy, hereditary or acquired bleeding disorders, primary spinal cord tumors, spinal cord tumors⁸⁻¹⁰. The most frequent localization in children is C5-T1, while in adults, it is more frequently found at a low cervical and thoracolumbar level^{11,12}. MRI is considered the most valuable diagnostic tool^{13,14}. The stages and extent of hematoma can be seen clearly, and sometimes the underlying pathology can be identified. The role of angiography has been debated. Kaaravelis *et al.* indicated that spinal angiography was not necessary, if MRI was diagnostic and did not show any abnormal vessels¹⁷.

Hematomyelia is manifested by bilateral or unilateral paralysis of the extremities, reduction in sensitivity to pain and temperature in one or both halves of the body, and sometimes by retention of urine and stool. Muscular atrophy and autonomic disorders (such as increase or decrease in perspiration) are often observed. Hematomyelia usually presents with an acute onset and rapid deterioration in neurologic status and usually leads to an acute spinal cord syndrome. Prompt diagnosis of hematomyelia first requires recognition of a myelopathy syndrome (transverse, central, anterior, posterior, or hemi-cord) often accompanied by sudden, severe back or neck pain and sometimes redicular pain. Besides the common type of hematomyelia, there is documentation of stepwise or chronic progressive patterns of hematomyelia¹⁵.

Intramedullary spinal cord haemorrhage in patients with hemophilia has been occasionally reported. Treatment is based on prompt replacement therapy as the occurrence and development of neurologic dysfunction are related to the length of time between the onset of symptoms and the factor replacement.

There are no clinical trials to guide the management of acute intramedullary spinal cord hemorrhage, and subsequent treatment is usually directed toward the underlying cause. Most of the literature recommends early diagnosis and surgery for cases of intramedullary haemorrhage with progressive neurologic deterioration^{13,14,16}. Our case was managed conservatively with replacement therapy with cryoprecipitate and patient improved gradually. More Number of cases needs to be studied to know this rare condition better.

IV. Conclusion

In conclusion, hematomyelia is an infrequently encountered condition in patient with haemophilia. The other causes of spontaneous, nontraumatic hematomyelia include vascular malformations of the spinal cord (the most common), clotting disorders and spinal cord tumors. High index of clinical suspicion, a timely diagnosis using MRI, and early intervention are essential in attaining a better neurological prognosis

References

- [1]. Hunderfund AN Leep, Wijidicks EF. Intramedullary spinal cord hemorrhage (hematomyelia). *Rev Neurol Dis* 2009;6:E54-61.
- [2]. Pullakart VA, Kalapura T, Pincus M, Baskharoun R. Intraspinal hemorrhage complicating oral anticoagulant therapy. *Arch Intern Med* 2000;160:237-40.
- [3]. Hylek EM, Singer DE. Risk factors for intracranial hemorrhage in outpatients taking warfarin. *Ann Intern Med*. 1994;120:891-902.
- [4]. Constantini S, Ashkenazi E, Shoshan Y. Thoracic hematomyelia secondary to coumadin anticoagulant therapy: a case report. *Eur Neurol*. 1992;32:109-11.
- [5]. Pisani R, Carta F, Guiducci G. Hematomyelia during anticoagulant therapy. *Surg Neurol*. 1985;24:578-80.
- [6]. Pullakart VA, Kalapura T, Pincus M, Baskharoun R. Intraspinal hemorrhage complicating oral anticoagulant therapy. *Arch Intern Med*. 2000;160:237-40.
- [7]. Richardson JC. Spontaneous hematomyelia: a short review and a case report illustrating intramedullary angioma and syphilis of the spinal cord as possible causes. *Brain* 1938;61:17-36.
- [8]. Pullakart VA, Kalapura T, Pincus M, Baskharoun R. Intraspinal hemorrhage complicating oral anticoagulant therapy. *Arch Intern Med*. 2000;160:237-40.#
- [9]. Koss WT, Bock F. Spontaneous multiple intramedullary hemorrhages. *J Neurosurg*. 1970;32:581-4.
- [10]. Schenk VWD. Hemorrhages in spinal cord with syringomyelia in a patient with hemophilia. *Acta Neuropathol*. 1963;2:306-8.
- [11]. Chen CJ, Hsu WC. Experience in the surgical management of spontaneous spinal epidural hematoma. *J Formos Med Assoc*. 1997;96:283-7
- [12]. Patel H, Boaz JC, Philips JP. Spontaneous spinal epidural hematoma in children. *Pediatr Neurol*. 1998;19:302-7.
- [13]. Chao CH, Tsai TH, Huang TY, Lee KS, Hwang SL: Idiopathic spontaneous intraspinal intramedullary hemorrhage: a report of two cases and literature review. *Clin Neurol Neurosurg* 115: 1134-1136, 2013
- [14]. Hwang JH, Sung JK, Hwang SK, Hamm IS, Park YM, Kim SL: Spontaneous hematomyelia: Case report. *J Korean Neurosurg Soc* 29:411-419, 2000
- [15]. Barnwell SL, Dowd CF, Davis RL, et al. Cryptic vascular malformations of the spinal cord: diagnosis by magnetic resonance imaging and outcome of surgery. *J Neurosurg* 1990;72:403-7.
- [16]. Licata C, Zoppetti MC, Perini SS, Bazzan A, Gerosa M, Da Pian R: Spontaneous spinal haematomas. *Acta Neurochir (Wien)* 95:126-130, 1988
- [17]. Purroy F, Arenillas JF, Rovira A, et al. [Hematomyelia as late side effect of radiation]. *Med Clin (Barc)* 2002;119:799.