Case Report تقرير حالة طبية

SPONTANEOUS SPINAL EPIDURAL HEMATOMA: A CASE REPORT AND REVIEW OF THE LITERATURE

الورم الدموي النخاعي العفوي فوق الجافية: تقرير حالة طبية ومراجعة في الأدب الطبي

Fakhr Fakhouri, MD; Anwar Abed, MD; Abdelwahed Ghajar, MD, FRCS

د.فخر فاخوري، د.أنور العابد، د.عبد الواحد غجر

ملخص الحالة

يعتبر الورم الدموي النخاعي العفوي فوق الجافية حالة إسعافية نادرة وذات عقابيل خطيرة. نورد هذه الحالة لتأكيد أهمية الإدراك المبكر والتداخل الجراحي العاجل في هذه الحالة. تظاهر طفل في العاشرة من عمره ببدءٍ تدريجي لألم رقبي وظهري، إقياء وصلابة نقرة تلاه خزل سريع الترقي في الطرفين السفليين، فرط حس في البطن والطرفين السفليين وسوء في وظيفة المصرات. أظهر التصوير بالرنين المغناطيسي MRI وجود ورم دموي فوق الجافية في المستويات الفقرية الرقبية السابعة C7 وحتى الصدرية الثالثة T3. أجريت جراحة إسعافية تم فيها خزع الصفائح الفقرية وإفراغ الورم الدموي وذلك في اليوم الثالث من حدوث شلل الطرفين السفليين. حدث تحسن سريري ملحوظ لكن بطيء وتم تخريج المريض بعد عشرة أيام يمشي بالمساعدة. تحسن المريض بعد شهرين من المتابعة وعادت الوظيفة العصبية طبيعية. نستنتج من هذه الحالة أن الورم الدموي النخاعي العفوي فوق الجافية حالة نادرة لكنها مسببة العجز وقد تكون مهددة للحياة. يمكن الحصول على أفضل النتائج بالتشخيص المبكر والتداخل الجراحي العاجل لهذه الحالة. إن التظاهرات البدئية المضللة التي قد تتظاهر بها هذه الحالة وخاصةً عند الأطفال قد تسبب تأخراً في وضع التشخيص، ولهذا يجب لفت نظر الأطباء ذوي الصلة إلى هذه الحالة النادرة لأخذها بالاعتبار في التشخيص التفريقي لحالات متلازمات النخاع الشوكي سريعة الترقي.

ABSTRACT

Spontaneous spinal epidural hematoma (SSEH) is a rare emergency with potentially serious sequel. We report a case of SSEH to emphasize the importance of early recognition and urgent surgery for this challenging emergency. A 10-year-old boy presented with gradual onset of neck and back pain, vomiting, and nuchal rigidity followed by rapidly progressive paraparesis, abdomen and lower extremities hyperesthesia and sphincter dysfunction. MR imaging demonstrated epidural hematoma of cervico-thoracic spinal segments (C7-T3). Emergent decompressive laminectomy with hematoma evacuation was performed the third day after paraplegia onset. Neurological improvement was obvious but slow, and the patient was discharged walking with aid 10 days after surgery. Two months later he got full functional recovery. We conclude that SSEH is a rare but disabling or even fatal clinical challenge. Early diagnosis and prompt surgery improve the neurological and functional outcome. Initial nonspecific symptoms can lead to a delay in diagnosis, especially in younger children. Relevant physicians should pay attention to the symptoms of this rare entity and SSEH should be one of differential diagnoses of rapidly evolving spinal cord syndrome.

*Fakhr Fakhouri, MD; Neurosurgery Senior Resident, Division of Neurosurgery, Department of Surgery, Aleppo University Hospital, Aleppo, Syria.
E-mail: fakhrf@msn.com

*Anwar Abed, MD, Neurosurgeon, Division of Neurosurgery, Department of Surgery, Aleppo University Hospital, Aleppo, Syria.

*Abdelwahed Ghajar, MD, FRCS, Consultant Neurosurgeon, Division of Neurosurgery, Department of Surgery, Aleppo University Hospital, Aleppo, Syria.

INTRODUCTION

After Jackson reported the first case of spontaneous spinal epidural hematoma SSEH in 1869,[1] SSEH became a well-known though rare cause of spinal cord compression in the emergency department. The estimated incidence is about 1 patient per million individuals and represents less than 1% of spinal space-occupying lesions.[2] The underlying cause cannot be identified in more than 40% of the cases.[3,4] However, many underlying diseases, such as coagulation disorders, vascular malformations may be the cause.[5-8] Association with spine diseases such as ankylosing spondylitis[9,10] and Paget's disease[11] has also been reported. SSEH usually presents with neck or back pain followed by acute spinal cord compression syndrome. Only about 30 pediatric cases of SSEH have been documented in the medical literature.[8,12] We report a pediatric case of SSEH and review the relevant literatures.

CASE PRESENTATION

A previously healthy 10-year-old boy was referred to the emergency department (ED) with rapid onset of upper back pain, occipital headache and sharp pain radiating to the right upper limb. There was no recent history of trauma or infection. He was so agitated and had misleading manifestations such as abdominal pain, vomiting and noticeable nuchal rigidity. Although he had difficulty with walking but he looked rather lethargic for lower limbs weakness was not as prominent. After admission to the pediatric department, investigations included LP and CSF analysis was normal except for mildly high protein level (82 mg/dl). Laboratory investigation included complete blood count, chemistry panel, and coagulation profile were all within normal limits. Next day he started to experience increasing lower limbs weakness and burning sensation and was unable to walk. Spinal MRI and neurosurgical consult was ordered.

On examination the patient was orientated, but looked irritated and disturbed with pain to a degree that made communication difficult. Abdominal examination revealed soft but dysesthetic abdomen, with distended bladder. He was afebrile and had unremarkable cardiovascular and chest examination.

Neurological examination revealed almost complete paraplegia, severe lower limbs hyperesthesia, and prominent nuchal rigidity. Impaired sensation below T1 level was noted. His reflexes were hypertonic and Babinski sign was positive bilaterally.

MR imaging demonstrated acute spinal epidural hematoma from C7 through T3 causing compression of the spinal cord. The hematoma was relatively isointense on T1-weighted images and hypointense on T2-weighted images (Figure1).

Emergent decompressive laminectomy was performed to remove relatively large epidural hematoma about 4 days after admission and 3 days after the onset of paraplegia. Intraoperatively there was

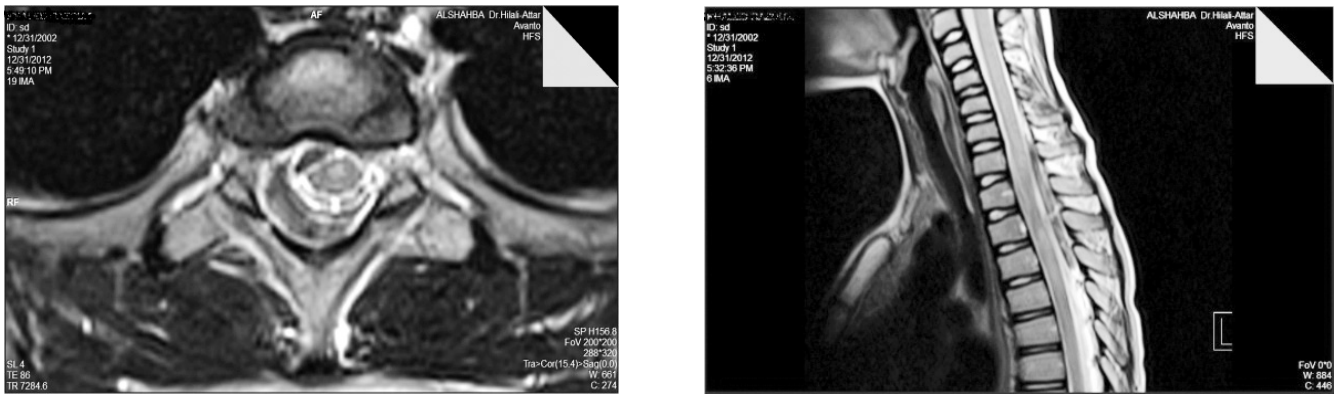

Figure 1. (A) Sagittal T2W MRI shows a spontaneous spinal epidural hematoma extended from C7 to T3 with spinal cord signal change. (B) Axial T2W MRI shows multi-loculated SSEH compressing spinal cord and deviating it to left.

an abnormal artery on the dorsal surface of dural sac that caused problematic intraoperative bleeding which was controlled with isolation and bipolar coagulation. The hematoma was evacuated except for small anterior residue. Two days after operation sensation and urinary continence regained, but muscle strength recovery was very slow. The patient was discharged 10 days later walking with aid. Two months later he got full recovery of neurological function.

DISCUSSION

Spontaneous Spinal Epidural Hematoma (SSEH) is an accumulation of blood in the vertebral epidural space in the absence of trauma or iatrogenic procedure such as lumbar puncture. The location and onset age of SSEH have a bimodal distribution with the location peaks at C6 and T12 and onset age peaks at 15-20 and 65-70 years respectively.[13,14] The male/female ratio is 1.4:1.[3,15] Pediatric SSEH is extremely rare, with only about 30 cases of SSEH documented in the medical literature.[8,12]

Idiopathic cases account for at least 40% of all cases.[6] However, certain precipitating factors are suggested to be correlated with SSEH. This could be divided into two categories. First; medications or disorders that alter normal coagulation such as anticoagulant therapy for prosthetic cardiac valve,[5,7] thrombolysis therapy for acute myocardial infarction,[7] uncontrolled hypertension,[16] aortic coarctation,[17] end-stage renal disease receiving hemodialysis,[18] long-term antiplatelet usage[19] and congenital diseases with factor XI deficiency or hemophilia B.[20-23] Second; spinal diseases such as ankylosing spondylitis[9,10] and Paget's disease.[11]

The origin of these hematomas is still debatable. Many authors accept the venous etiology hypothesis due to lacks of venous valves in epidural venous plexus.[24] However, several authors have proposed the spinal epidural arteries as a source of hemorrhage,[25] due to the fact that pressure from arterial bleeding is more likely to compresses the spinal cord, as intra-thecal pressure is higher than the venous pressure.[26,27] Only rarely, SSEH associated with an identifiable underlying vascular lesion[28] such as hemangioma[29] or vascular malformation.[30-32] In our case an abnormally tortuous dural artery was evident at surgery. Anyway, further studies are needed to clarify the precise pathogenesis of SSEH.

The usual clinical presentation of SSEH is sudden onset of severe neck and/or back pain. Motor and/or sensory deficits caused by compression of nerve roots and spinal cord follow and progress rapidly with sphincter incontinence. Despite the characteristic syndrome of SSEH, initial nonspecific symptoms can lead to a delay in diagnosis, especially in younger children. Unfortunately, children often present with nonspecific presentation and may suffer from additional symptoms of irritability, nuchal rigidity, and occasionally urinary retention.[12] Furthermore, a recent case of SSEH in a 10-year-old girl is interesting in that she presented with what initially appeared to be a pure brachial plexopathy.[33] In fact, in our case the abdominal pain, right shoulder pain, and vomiting were so deceiving so that hepatitis, cholecystitis and meningitis were all on top differential diagnosis.

Currently the introduction of MR imaging help making the diagnosis early and accurate, and define the location and size of hematoma. According to the age of hematoma the MR images of acute SEH reveal variable findings from isointense to hypointense to hyperintense on T1-weighted images and from hyperintense to hypointense on T2-weighted images. Beside specific signal changes, contrast enhancement pattern and morphological findings on MR images can differentiate acute SSEH from spinal epidural neoplastic mass or abscess.[34] In addition, MRI may rarely demonstrate the underlying lesions, such as spinal vascular malformations.[31,35]

The differential diagnosis of spontaneous spinal epidural hematoma includes an acute herniated intervertebral disc, acute ischemia of the spinal cord, epidural tumor or abscess, spondylitis, transverse myelitis, or even a dissecting aortic aneurysm and acute myocardial infarction.

SSEH is generally surgical emergency with rapid hematoma evacuation being the most effective treatment.[36,37] Conservative treatment is still an important option of treatment in some selected patients

with mild and rapid spontaneous recovery or in cases that is highly risky for surgery.[38] It is interesting in this regard to note that some non-surgical SSEH patients have been noted to improve following lumbar puncture; that may be due to reduction of intraspinal pressure via thecal sac puncture and cerebrospinal fluid egress which leads to partial cord decompression.[39]

The prognosis of SSEH correlates with the size and level of hematoma, severity of preoperative neurological deficits and time interval between symptoms onset and surgery. Of these, the interval from symptoms onset to surgery appears to be the single most important factor.[40] Recurrence is extremely rare and there is a case that documents recurrence of a cervical epidural haematoma 3 years after the first incidence.[41,42]

CONCLUSIONS

Still remain a clinical challenge, SSEH is a rare and serious condition that can be managed with good outcome. Early diagnosis with MRI and urgent surgical decompression is the key. Relevant physicians should keep in mind SSEH as one of differential diagnoses of rapidly evolving spinal cord syndrome.

REFERENCES

1. *Jackson R. A case of spinal apoplexy. Lancet 1869;2:5-6.*
2. *Holtas S, Heiling M, Lonntoft M. Spontaneous spinal epidural hematoma: findings at MR imaging and clinical correlation. Radiology 1996;199:409-13.*
3. *Liu Z, Jiao Q, Xu J, et al. Spontaneous spinal epidural hematoma: analysis of 23 cases. Surg Neurol 2008;69:253-60.*
4. *Penar PL, Fischer DK, Goodrich I, et al. Spontaneous spinal epidural hematoma. Int Surg 1987;72:218-21.*
5. *Vaya A, Resureccion M, Ricart JM, et al. Spontaneous cervical epidural hematoma associated with oral anticoagulant therapy. Clin Appl Throm Hemost 2001;7:166-8.*
6. *Betty R, Winston K. Spontaneous cervical epidural hematoma. A consideration of etiology. J Neurosurg 1984;61:143-8.*
7. *Van Schaeybroeck P, Van Calenberg F, Van De Werf F, et al. Spontaneous spinal epidural hematoma associated with thrombolysis and anticoagulation therapy: report of three cases. Clin Neurol Neurosurg 1998 Dec;100(4):283-7.*
8. *Tailor J, Dunn IF, Smith E. Conservative treatment of spontaneous spinal epidural hematoma associated with oral anticoagulant therapy in a child. Childs Nerv Syst 2006;22:1643-5.*
9. *Wu CT, Lee ST. Spinal epidural haematoma and ankylosing spondylitis: case report and review of the literature. J Trauma 1998;44:558-61.*
10. *van de Straete S, Demaerel P, Stockx L, et al. Spinal epidural haematoma and ankylosing spondylitis. J Belge Radiol 1997;80:109-10.*
11. *Hanna JW, Ball MR, Lee KS, et al. Spontaneous spinal epidural hematoma complicating Paget's disease of the spine. (case report). Spine 1989;14(8):900-2.*
12. *Poonai N, Rieder MJ, Ranger A. Spontaneous spinal epidural hematoma in an 11-month-old girl. Pediatr Neurosurg 2007;43:121-4.*
13. *Kreppel D, Antoniadis G, Seeling W. Spinal hematoma: a literature survey with meta-analysis of 613 patients. Neurosurg Rev 2003;26:1-49.*
14. *Groen RJM. Non-operative treatment of spontaneous spinal epidural hematomas: a review of the literature and a comparison with operative cases. Acta Neurochir (Wien) 2004;146:103-10.*
15. *Lonjon MM, Paquis P, Chanalet S, et al. Nontraumatic spinal epidural hematoma: report of four cases and review of the literature. Neurosurgery 1997;41:483-7.*
16. *Spengos K, Tsivgoulis G, Zakopoulos N. Could high blood pressure be the cause of acute spontaneous spinal epidural hematoma? Eur J Emerg Med 2007;14:59.*
17. *Zizka J, Elias P, Michl A, et al. Extensive spinal epidural haematoma: a rare complication of aortic coarctation. Eur Radiol 2001;11:1254-8.*
18. *Deger SM, Emmez H, Bahadirli K, et al. A spontaneous spinal epidural hematoma in a hemodialysis patient: a rare entity. Intern Med 2009;48:2115-8.*
19. *Weber J, Hoch A, Kilisek L, et al. Spontaneous intraspinal epidural hematoma secondary to use of platelet aggregation inhibitors. Dtsch Med Wochenschr 2001;126:876-8.*
20. *Mustafa MH, Bernstain RA. Spontaneous spinal epidural hematoma, Brown-sequard syndrome, and factor XI deficiency. Ann Intern Med 1987;106:477-8.*

21. *Bisson EF, Dumont T, Tranmer B. Spontaneous spinal epidural hematoma in a child with hemophilia B. Can J Neurol Sci 2007;34:488-90.*
22. *Caldemeyer KS, Mocharla R, Moran CC, et al. Gadolinium enhancement in the center of a spinal epidural haematoma in a hemophiliac. J Comput Assist Tomogr 1993;17:321-3.*
23. *Meena AK, Jayalakshmi S, Prasad VS, et al. Spinal epidural haematoma in a patient with haemophilia-B. Spinal Cord 1998;36(9):658-60.*
24. *Groen RJM, Ponssen H. Vascular anatomy of the spinal epidural pace: Considerations on the etiology of the spontaneous spinal epidural hematoma. Clin Anat 1991;4(6):413-20.*
25. *Solheim O, Jorgensen JV, Nygaard OP. Lumbar epidural hematoma after chiropractic manipulation for lower back pain: Case report. Neurosurgery 2007 Jul;61(1):E170-1; discussion E17.*
26. *Guzel A, Simsek O, Karasalihoglu S, et al. Spontaneous spinal epidural hematoma after seizure: a case report. Clin Pediatr 2007;46:263-5.*
27. *Park J, Lee JB, Park JY, et al. Spinal cord infarction after decompressive laminectomy for spontaneous spinal epidural hematoma. Neurol Med Chir (Tokyo) 2007;47:325-7.*
28. *Ghanem Q, Ivan LP. Spontaneous spinal epidural hematoma in an 8-year-old boy. Neurol 1978;28:829-32.*
29. *Ter Spill HW, Tijssen CC. Spinal epidural haematoma due to a vertebroepidural hemangioma. Clin Neurol Neurosurg 1989;91:91-3.*
30. *Solero CL, Fornari M, Savoiardo M. Spontaneous spinal epidural haematoma arising from ruptured vascular malformation: case report. Acta Neurochir (Wien) 1980; 53:169-74.*
31. *Muhonen MG, Piper JG, Moore SA, et al. Cervical epidural haematoma secondary to an extradural vascular malformation in an infant: case report. Neurosurg 1995;36:585-8.*
32. *Muller H, Schramm J, Roggendorf W, et al. Vascular malformations as a cause of spontaneous spinal epidural haematoma. Acta Neurochir (Wien) 1982;62:297-305.*
33. *Ravid S, Schneider S, Maytal J. Spontaneous spinal epidural hematoma: an uncommon presentation of a rare disease. Childs Nerv Syst 2002;18:345-7.*
34. *Chang FC, Lirng JF, Chen SS, et al. Contrast enhancement patterns of acute spinal epidural hematomas: A report of two cases. AJNR 2003;24:366-9.*
35. *Miyagi Y, Miyazono M, Kamikaseda K. Spinal epidural vascular malformation presenting in association with a spontaneously resolved acute epidural haematoma. Case report. J Neurosurg 1998;88:909-11.*
36. *Matsumura A, Namikawa T, Hashimoto R, et al. Clinical management for spontaneous spinal epidural hematoma: diagnosis and treatment. Spine J 2008;8:534-7.*
37. *Liao CC, Hsieh PC, Lin TK, et al. Surgical treatment of spontaneous spinal epidural hematoma: a 5-year experience. J Neurosurg Spine 2009;11:480-6.*
38. *Hentschel SJ, Woolfenden AR, Fairholm DJ. Resolution of spontaneous spinal epidural hematoma without surgery. Spine 2001;26:E525-7.*
39. *Robertson WC Jr, Lee YE, Edmonson MB. Spontaneous spinal epidural haematoma in the young. Neurol 1979; 29:120-2.*
40. *Lawton MT, Porter RW, Heiserman JE, et al. Surgical management of spinal epidural hematoma: relationship between surgical timing and neurological outcome. J Neurosurg 1995;83:1-7.*
41. *Demierre B, Unger PF, Bongioanni F. Sudden cervical pain: spontaneous cervical epidural haematoma. Am J Emerg Med 1991;9:54-6.*
42. *Hans P, Delleuze PP, Born JD, et al. Epidural hematoma after cervical spine surgery. J Neurosurg Anesthesiol 2003;15:282-5.*