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Case Report
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Leukemia Presenting as Cauda Equina Syndrome

Syed Shumon[1], Georgia Ineson[2], Terence Chin[2], Konstantinos Oikonomou[1], Gareth Dobson[1], Priya Bhatnagar[1] and Mohammed Husain[1]*

[1]Department of Neuroscience, The Royal Victoria Infirmary, UK

[2]Department of Neuroscience, The University of Newcastle upon Tyne, UK

Abstract

Spontaneous Spinal Subdural Hematoma (sSDH) is a rare entity with an incidence of 0.1/100,000 patients. Patients commonly present with a sudden onset of back pain and progressive neurological deficit. In the majority of the cases, it is due to a hemorrhage at the thoracic spinal level of unknown aetiology. Management of such cases can present a treatment dilemma. The difficult decision of conservative *vs*. surgical management must be made in view of the presenting neurology and likely outcome. Here we present a case of the first presentation of leukemia presenting as query cauda equina syndrome due to a sSDH.

Keywords: Back pain; Cauda equine syndrome; Sepsis; Acute myeloid leukemia; Blastic plasmacytoid dendritic cell neoplasm

Introduction

Spontaneous Spinal Subdural Hematoma (sSDH) is a rare condition of unknown aetiology and significant morbidity. Patients often present with acute onset of motor, sensory and/or autonomic dysfunction, coherent with the spinal level of the bleed. Management options include conservative management with best supportive treatment, percutaneous drainage or surgical decompression [1]. There are variable outcomes reported with either management option in the literature but it has been suggested that those with mild neurological deficit should be managed conservatively and those with severe neurological deficit should undergo surgical decompression [2].

Here we present a case of a 49 year-old-female that presented as query cauda equina syndrome secondary to a sSDH. She developed a blood picture of disseminated intravascular coagulation with an unidentifiable cause and was subsequently found to have acute myeloid leukemia.

Case Presentation

A 49 year-old female shop assistant with a background of hypertension who was under investigation by her GP for systemic lupus erythematosus, due to a three month history of generalized malaise and possible butterfly rash, presented to A&E with a three day history of severe sharp lower back pain, bilateral sciatica that was worse on the left, perineal and saddle anesthesia and five episodes of urinary incontinence. The patient also reported that both her legs were feeling weak. Patient denied any fever, night sweats or weight loss. There was no history of trauma. The patient was previously independently mobile, did not smoke, drank occasionally and was not on any anticoagulation. On examination the patient was of large habitus weighing 133.2 kg and had bruising to multiple areas of her arm. Cardio, respiratory and abdominal examination was entirely normal. Her GCS was 15; cranial nerves were intact, upper limb neurological examination was normal. Lower limb neurological examination elicited weak hip flexors bilaterally (MRC scale 3/5) and reduced sensation over left L5 dermatome. The patient also had saddle anesthesia and a lax anal tone.

Patient's blood tests on admission showed hemoglobin of 10^9 g/L, platelet of 157 (ref range 150 to 450) and a raised white cell count of 17.99 (ref range 4 to 11) with a neutrophilia. Coagulation screen revealed a PT of 18 (ref range 10 to 13) with a normal APTT and fibrinogen. Biochemistry analysis showed hyponatremia of 132 mmol/L and raised CRP of 142 (ref range 0 to 5).

The patient underwent an urgent MRI that showed a subacute subdural hematoma extending from the cauda equina to the craniocervical junction with compression of the cauda equine (Figure 1). The MRI also showed an abnormal diffuse abnormal T1 low signal throughout the spine with patchy enhancement suggestive of hematological malignancy (Figure 1). A CT chest, abdomen and pelvis revealed no signs of malignancy.

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Correspondence:
Syed Shumon, Department of Neuroscience, The Royal Victoria Infirmary, Queen Victoria Rd, Newcastle Upon Tyne NE1 4LP, UK, E-mail: syed.shumon@hotmail.co.uk

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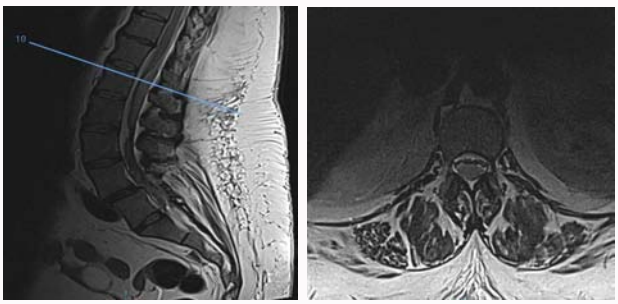

Figure 1: An MRI showing the SSDH.

After consideration for surgery and review of the pathology, it was decided to manage the patient expectantly with a patient-controlled analgesia device for pain management and a short course of steroids. Repeat blood tests two days from admission revealed hemoglobin of 110, a reduced platelet count of 92, white cell counts of 18.84 with a continued neutrophillia. The clotting function test revealed a PT of 19 and a reduced fibrinogen of 0.9 with a clauss fibrinogen and D-dimer of 0.5 and 13,245 respectively. The developing picture of disseminated intravascular coagulation with no clear cause and abnormal signal on MRI was discussed with the hematology team. The patient underwent further investigations under the hematology team, multiple infusions of blood products to correct the clotting deficiency and empirical treatment with broad-spectrum antibiotics. A bone marrow biopsy revealed an unusual case that proved to be difficult to distinguish between Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) and AML. The immunophenotype suggested a picture of BPDCN although it would also be compatible with AML. However genetic analysis showed a tetrasomy of chromosome 8 and a 4 bp insertion within exon 12 of the NPM1 gene. AML with tetrasomy of chromosome 8 is rare and BPDCN with NPM1 mutation has only been reported in one case report before. Furthermore, the significance and correlation of tetrasomy of chromosome 8 in BPDCN is unknown. The patient was transferred to the hematology service where she was treated with daunorubicin, cytarabine and mylotarg chemotherapy. The patient's stay was further complicated by neutropenic colitis, urosepsis, line sepsis and respiratory failure requiring further antibiotic treatment and respiratory support on ITU.

The patient made a good recovery and a partial neurological recovery. On transfer to the hematology services the patient had power 5/5 in all muscle groups of the lower limb but required intermittent catheterization for bladder management. On discharge from hospital the patient was mobilizing independently but required a wheelchair for long distances.

Discussion

Less than 250 cases of SSDH have been reported in literature with up to 50% reported to be sSDH. Spontaneous SSDH remain a rare entity with an incidence of 0.1/100,000 patient [3]. Those presenting due to hematological malignancy remain a rarer subset with less than 5 reported cases in literature.

sSDH are most commonly reported to occur in the thoracic spine and can span between one vertebral level up to 23 vertebral levels [4]. They have no gender predominance and are more common between the fourth to the sixth decade of life. The main presenting complaint is sharp stabbing back pain at the site of the hematoma with radiculopathy and concurrent progressive neurological deficit.

Traumatic SSDH (tSSDH) often arise due to spinal surgery, epidural catheterization, lumbar puncture, and chiropractic manipulation. The aetiology of sSSDH is unclear but hypertension, pregnancy, arteriovenous malformations; coagulopathy, therapeutic anticoagulation or underlying neoplasms are all thought to be risk factors.

Various theories have been suggested as to the pathogenesis of SSDH including bleeding originating in the more vascular subarachnoid space and then extending into the subdural space due to rupture of vascular structures following a sudden raise in intra-abdominal/thoracic pressure such as when coughing, sneezing, voiding or lifting heavy objects. Others have suggested that the bleeding originates directly in the epidural space from rupture of epidural veins, arteries, cryptic angiomas, vascular malformations, hemangiomas or spinal angiomas, which can all be further exacerbated by anticoagulation therapy.

Hematological disorder leading to coagulopathy or anticoagulation therapy are thought to account for 46% of sSSDH [4]. Other causes include ankylosing spondylitis, systemic lupus erythematosus, fibromuscular dysplasia, cystic fibrosis, polycystic kidney disease, chronic renal failure, rhabdomyolysis, rheumatoid arthritis, pregnancy and eclampsia. To date only 3 cases of leukemia have been associated with sSSDH with only one other case presenting as an index presentation of leukemia with sSSDH. The first case was following anticoagulation of a patient with known vertebral body osteolytic lesion from a plasma cell myeloma who developed a thoracic sSSDH and subsequent paraplegia [5]. The second was a 26-year-old male also known to have leukemia who presented with acute back pain, lower limb weakness and urinary difficulty who was found to have T9-T11 hematoma [6]. And finally, the third case was that of 55-year-old male who presented with acute back pain who developed paraplegia and a sensory level at T1 found to have a C5-T3 hematoma who was subsequent found to have BCR-ABL gene positive Chronic Myeloid Leukemia (CML) [3]. It is thought that spinal malignant lesions can induce epidural inflammation, cause epidural venous plexus to become more friable and induce vertebral microfractures which can predispose to sSDH [5].

The unique features of our cases are the chronicity of the presentation, the extent of the hematoma from the cauda equina to the cranial-cervical junction, the remarkable sparring of motor function and the unusual histopathology/genetic makeup of the bone marrow neoplasm. The three reported cases all reported acute onset back pain with progressive motor function deterioration within 6 h to 10 h of pain onset. Our patient had a 3-day history of severe back pain with only a mid-hip flexion weakness and bladder dysfunction despite the extensive nature of the hematoma on imaging. This would lead us to hypothesis that the bleed must have occurred from the lumbar epidural venous plexus which would allow for the relatively low-pressure venous bleed clot to slowly expand over time. This would account for chronicity of presentation, the extensive bleed in the epidural space on imaging and localize the pathology to the lumbar-sacral region accounting for the presenting symptoms of bladder dysfunction with lower back pain. AML is an acquired myeloproliferative disorder of growth arrest and failure of apoptosis in myeloid lineage cells. This is thought to occur due to chromosomal translocation and genetic abnormalities. The defective myeloid cells lead to drop in red blood cells, white blood cells and platelets predisposing to bleeding and infection. AML has an incidence of 5-8/100,000 with a median age of

onset of 67 years of age and AML being more common in the older population [7]. Presentation of AML can vary from an acute onset over days in children to a chronic fatigue over months in older people but a recognized presentation at onset is DIC, which was seen in our patient. Prognosis in AML varies depending on the age of the patient a disease burden with those having the highest disease burden and advanced age fairing the worst.

The strongest marker of outcome in SSDH is reported to be the presence and degree of neurological deficit at presentation. Leigh et al. reported in a systematic review that patients presenting with neurosurgical deficit at presentation had a 34% chance of good recovery compared to 83% of those presenting without neurological deficit [1,4,6]. This is supported by our case as the relative spared motor function on presentation and correlated with good ambulation on discharge. Others have also reported that delay in surgical intervention in patients presenting with neurological deficit can worsen outcome [2,8]. This puts a huge onus on the treating clinician to decide whether a conservative or surgical approach is best for the patient to preserve and improve neurological function.

Conclusion

Spontaneous spinal subdural hematomas, although very rare, should be considered in any patient presenting with sudden onset sharp back pain. The level of suspicion should be further heightened if there is neurological deficit and/or the patient has the aforementioned risk factors. Clinicians should understand that delay in diagnosis and treatment can lead to worsened neurological outcome. CML presenting as SSDH is very rare but now a recognized cause and thus clinicians should be aware of the presenting symptoms.

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