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Extra Dural Hematoma of the Dorso-Lumbar Region in a Hemophiliac: A Rare Entity

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Abstract

Introduction and objective: Hemophilia is a genetic bleeding disorder inherited as a recessive train linked to the male gender. Bleeding into the central nervous system in patients with hemophilia is an extremely dangerous condition that can be directly life-threatening, if left untreated. Extradural hematoma of the dorso-lumbar region is rare but potentially deadly disease in children. This condition can result in severe neurological deficits. We aim to discuss the clinical, radiological and progressive clinical aspects of this illness. *Case report*: We report the case of a 5-year-old child with severe hemophilia A treated for extradural hematoma of the dorso-lumbar region resulting from trauma. A spinal magnetic resonance imaging revealed an extradural hematoma. The patient was successfully treated with intensive replacement therapy and did not require surgical intervention. *Conclusion*: Extradural hematoma is a rare complication of hemophilia, that needs to be looked for in children who have bleeding disorders. For the best neurological outcome, early diagnosis is crucial.

Keywords

Extradural Hematoma, Children, Hemophilia

1. Introduction

Hemophilia is the most common hereditary bleeding disorder, with an incidence of 0.7 - 0.8/10,000 [1]. A child's central nervous system damage with bleeding issues can be exceedingly serious, even fatal, regardless of what kind of hemophilia a child can have. These conditions mostly involve bleeding into the central nervous system and affect 2% - 8% of children with hemophilia [2]. There have only been a few reported cases of hemophilia-related bleeding in and around the

spinal cord [3]. Severe back or radicular pain is typically the first clinical symptom, followed by motor weakness and sensory disturbance that varies according to the severity of cord compression [3].

As soon as a spinal cord injury is identified or as soon as symptoms suggestive of central nervous system bleeding manifest, treatment with supplements should begin.

We report a case of 5-year-old male with hemophilia A who had extradural hematoma in dorso-lumbar region. This case highlights the features of this rare condition and discusses the role of factor replacement therapy.

2. Clinical Observation

A 5-year-old male child with severe hemophilia A (factor VIII levels < 1% of normal) diagnosed from the age of 14 months, indicated by post-circumcisional hemorrhage, suffered slight spinal column trauma after falling on his back from a sitting position. He was admitted to the pediatric emergency department for acute pain over the lumbosacral region.

Clinical examination revealed a child in severe pain (visual analog scale (VAS) rated at 8/10), a sensitivity in the lumbar fossae. He was afebrile with normal vital signs. Thus, we noted the presence of a few ecchymosis on the lower limbs and back. There was no evidence of vertebral fracture. His abdomen was soft, nondistended, and nontender. Neurological examination revealed no focal deficits, moreover the rest of the clinical examination showed no abnormalities. Supplementation of recombinant factor VIII was started from the very beginning, as per applicable recommendations, at a dose of 50 U/kg.

Laboratory tests indicated a normal complete blood cell counts, electrolytes, liver function. A Computed tomography (CT) of the abdomen and pelvis scan shows a spontaneously hyperdense masse 34 × 8 mm at the L2-L3 Level. Imaging using magnetic resonance MRI of the spine was conducted in order to determine whether surgical decompression was necessary and to assess the full extension and mass effect of the hematoma. MRI (**Figure 1**) demonstrated a posterolateral extradural hematoma of the dorso-lumbar level.

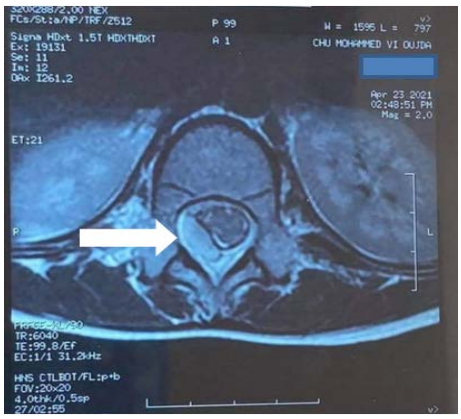

Figure 1. Magnetic resonance imaging showing an extra dural medullary hematoma in a child with major hemophilia A.

Surgical abstention was indicated by the neurosurgeons, treatment with factor VIII supplementation was initiated at a rate of 50 IU/kg every 8 hours for 10 days, then 50 IU/kg/12h for 4 days then 50 IU/kg/day.

The patient showed clinical improvement: VAS pain 0/10th on day 10 of treatment. A follow-up magnetic resonance imaging (MRI) revealed resolution of spinal extradural hematoma carried out on day 15 of treatment. Subsequently, the patient was put on a prophylactic dose factor VIII substitution at a rate of 20 IU/Kg three times a week.

At the last follow-up, one year after starting treatment, the patient was doing well.

3. Discussion

Hemophilia A and B are X chromosome-linked bleeding disorders included among the rare diseases and caused by mutations in the factor VIII (FVIII) and factor IX (FIX) genes. The intrinsic pathway of blood coagulation is influenced by both factors, and those who are affected by the diseases can have mild, moderate, or severe symptoms, defined by factor plasma levels of 1% or less, 2% to 5% and 6% to 40%, respectively [4]. Hemophilia A is an X-linked recessive disorder caused by factor VIII deficiency and accounts for 85% of hemophiliac cases [5]. The diagnosis of hemophilia A should be suspected whenever unusual bleeding is encountered in a male patient, and this suspicion is confirmed when the results of screening laboratory tests show a normal platelet count and prothrombin time but a prolonged activated partial-thromboplastin time. Specific factor assays are then required to determine the specific deficiency, since this is the only method to distinguish between hemophilia A and hemophilia B (factor IX deficiency). Hemophilia A's clinical characteristics include joint and muscle hemorrhages, bruising, and deep hemorrhages in Closed-space bleeding is especially dangerous, since it can lead to nerve paralysis or to vascular which can compromise the functional and vital prognosis of the affected child [6]. Spinal epidural hematoma (SEH) is a rare complication from hemophilia and represents 2% - 8% of all central nervous system hemorrhages. [7]. Extradural hematomas are a dangerous medical condition with potentially high rates of morbidity and death. Both traumatic and spontaneous spinal epidural hematoma patients typically present with a uniform constellation of symptoms, such as neck and/or back pain and paresis, followed by sensory deficits and bladder or bowel dysfunction [8].

Our case illustrates the unusual clinical presentation possible with epidural hematoma in an infant with hemophilia. In our presented patient, FVIII activity was <1%. The most likely reason for the bleeding was a minor trauma occurring after falling on his back from a sitting position. The investigation of choice remains noncontrast MR imaging, which can detect even small amounts of blood in the spinal canal. Noncontrast computed tomography is useful as an adjunct to rule out bony lesions. In our case, MRI demonstrated a posterolateral extradural hematoma of the dorso-lumbar level. The most common site for spontaneous

SEH in children is the cervical segment because of its greater mobility [9].

Early and prompt treatment is the most important factor influencing the outcome of the patient [10]. It is advised to administer prolonged replacement therapy for 7 to 14 days in order to keep factor VIII levels above 30% to 50% in cases of intracranial hemorrhage. The same guidelines should be followed in cases of intraspinal hemorrhage [7].

In the case of CNS trauma or suspected trauma in a hemophilia patient, supplementation treatment should be started immediately before performing imaging diagnostics. In the case of hemophilia A, the dose is selected so that the first seven days enable achievement of an activity level of 80 - 100 IU/dL as the starting dose, followed by a maintenance dose of 50 IU/dL over the next 14 days [11]. The mainstay treatment of children with spinal cord hematoma in the course of hemophilia consists in the substitution of the deficient factor. According to Nirupam *et al.*, the decision to proceed with surgery should be made after a patient's condition has been evaluated, neurological signs and symptoms and intensification of bleeding identified in MRI or CT. Additionally, Kubota and Miyajima consider that in patients with bleeding disorders, high doses of recombinant factor supplementation may enable treatment and result in the resolution of the neurological signs of an epidural hematoma without the need for surgical intervention [7].

Physicians treating patients with hemophilia should consider early use of commercially available factor VIII concentrate since bleeding into or around the central nervous system can have severe repercussions, even if the signs and symptoms are mild and the trauma appears insignificant [7].

4. Conclusion

In hemophiliacs, neurological hemorrhagic complications are unusual. Extradural hematoma is an exception, particularly at the dorso-lumbar level. Even in the absence of neurological clinical symptoms, a hemophiliac's acute backache leads to the diagnosis of hematoma. The primary method of treatment is by substituting coagulation factors. It must begin before confirmation of the diagnosis by magnetic resonance imaging. The clinical outcome is most likely favourable.

Consent

Written informed consent was obtained from the patient parents for publication of this case report and accompanying images.

Author Contributions

All authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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