

Epidural Hematoma in an Infant Revealing Hemophilia A

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Abstract

Case Report

Epidural hematoma is a rare complication of hemophilia. **Case report:** We report the case of a 10-month-old child admitted with motor deficits; a spinal MRI revealed an extensive epidural hematoma and the initial diagnosis of hemophilia. The child is responding well to medical treatment. **Conclusion:** Sudden spinal pain and/or motor deficits should raise the suspicion of an epidural hematoma in a child, and hemophilia should be investigated as an underlying cause. Treatment is primarily medical, involving substitution with coagulation factors. This should be initiated before the diagnosis is confirmed by magnetic resonance imaging (MRI). The clinical evolution is most often favorable.

Keywords: Hemophilia, Epidural hematoma, Infant, Spinal cord compression, Magnetic resonance imaging (MRI), Case report.

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INTRODUCTION

The occurrence of an epidural hematoma is rare in hemophiliacs. The bleeding can lead to compression of the spinal cord or cauda equine roots, causing neurosensory symptoms. We report here the case of a 10-month-old child admitted with motor deficits and, on spinal MRI, an extensive epidural hematoma, with hemophilia diagnosed for the first time, and whose evolution is favorable under medical treatment.

CASE PRESENTATION

This is a 10-month-old infant with a history of recurrent bruises and hematomas that have never been investigated, who presents with torticollis; The onset of symptoms dates back to one day before admission, marked by the development of torticollis; on physical examination, the child is conscious, with right-sided hemiplegia graded at 3/5, is breathing normally, afebrile, in good general condition, with skin and mucosal pallor and some bruising on the lower limbs.

An emergency spinal MRI was requested, revealing a large epidural hematoma extending throughout the cervical, thoracic and lumbar spine (from

C1 to L5), associated with adjacent meningeal contrast enhancement. This is responsible for spinal cord compression at this level with signs of spinal cord injury at the C4-C5 level (see Figure 1).

An etiological assessment was initiated, including a coagulation profile: complete blood count, PT, APTT and factor VIII and von Willebrand factor assays, revealing anemia (with Hb at 8 g/dl), a prolonged APTT (at 1.17), normal PT (100%), severely reduced factor VIII (0.9), and normal von Willebrand factor; the overall findings are consistent with hemophilia A.

Treatment was initiated immediately with factor VIII substitution therapy combined with corticosteroid therapy.

The patient's condition improved favorably, marked by the spontaneous onset of movement in the right upper limb on day 5.

A follow-up spinal MRI at one month showed complete resolution of the epidural hematoma and the absence of signs of spinal cord injury (Figure2).

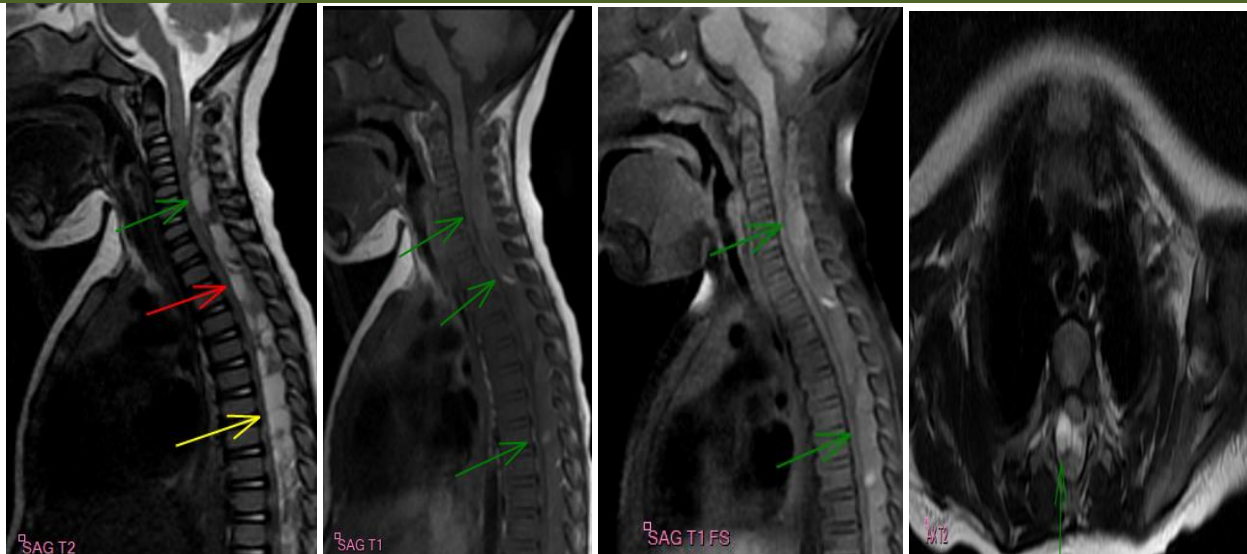


Figure 1: Spinal MRI with sagittal T1, T2 and T1FS images, and axial T2 images showing a large epidural hematoma extending throughout the cervical, thoracic and lumbar spine (from C1 to L5), appearing isointense on T1 and hyperintense on T2, containing areas of marked T2 hypointense signal and other areas of T1 hyperintense signal; following gadolinium injection, adjacent meningeal enhancement is noted; the whole complex is responsible for spinal cord compression at this level with signs of spinal cord injury at the C4-C5 level

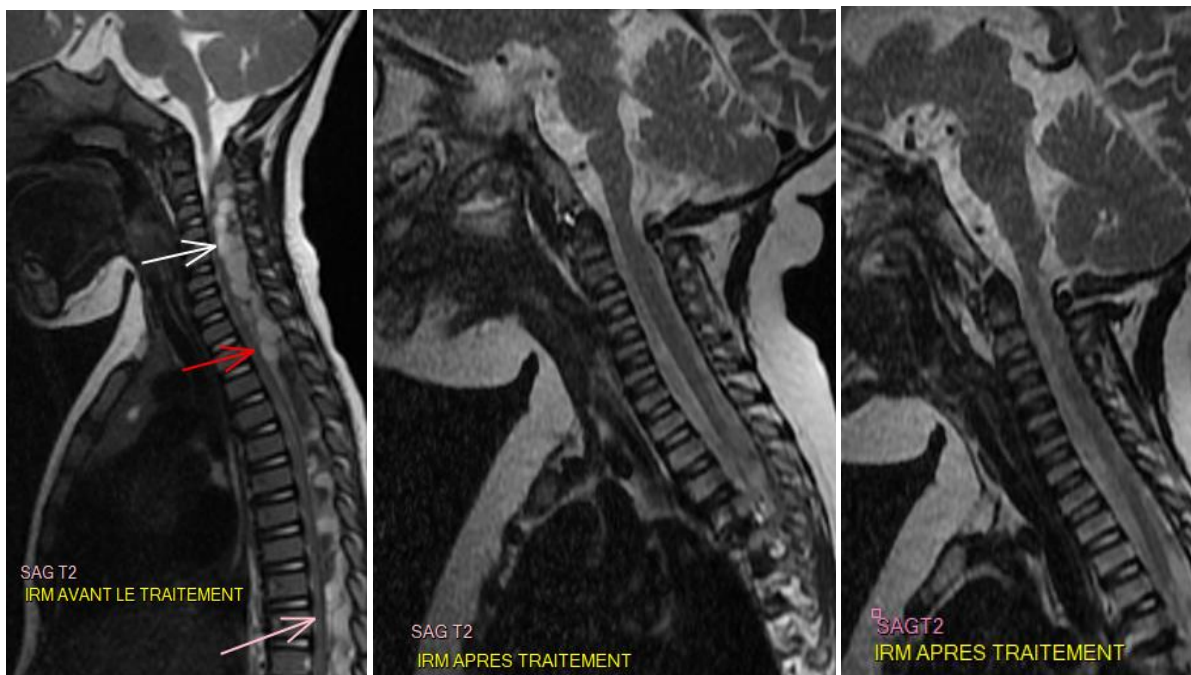


Figure 2: Sagittal T2-weighted spinal MRI scans taken before and after treatment, showing complete resolution of the hematoma after one month of treatment

The clinical follow-up two months later revealed no neurological sequelae.

DISCUSSION

The incidence of hemorrhagic complications affecting the central nervous system varies between 2.2% and 7.8% depending on the study [1, 2]. Among these complications, epidural hematoma remains relatively rare, making it difficult to establish its exact incidence. A study conducted in South America, involving 1,410 hemophiliac patients with central nervous system

hemorrhages, found that only two cases of epidural hematoma had occurred [1]. In approximately 10% of cases, a history of prior trauma was documented [3]. The cause of the bleeding is often attributed to the anatomical characteristics of the epidural venous plexuses, which are weak and lack anti-reflux valves. Consequently, maneuvers that abruptly increase abdominal or thoracic pressure could significantly increase the risk of hemorrhage [4, 5].

The most commonly observed sites of bleeding are in the cervical and thoracolumbar regions, which is consistent with our clinical case. These hematomas can lead to complications such as spinal cord compression or nerve root damage. Initial clinical signs generally manifest as severe spinal pain, accompanied by motor or sensory deficits. In the most severe cases, paraplegia or tetraplegia, as well as sphincter dysfunction, may occur, with symptoms sometimes appearing later than the initial event.

MRI remains the examination of choice for diagnosing an epidural hematoma and assessing the extent of the injury. What makes an epidural hematoma distinctive is the variable signal intensity on T1-weighted images, where isointense signals may be observed. The study by Sklar *et al.*, in 1999 revealed that, within a group of 17 patients who presented with an epidural hematoma, ten of them showed an isosignal on T1-weighted imaging, whilst the other seven showed a T1 hyperintensity [7].

Treatment is urgent in order to limit the progression of the hematoma and reduce the risk of residual sequelae. It generally involves factor VIII or IX substitution therapy, with the aim of maintaining factor levels above 50%. The doses of factors administered and the duration of treatment vary considerably, but on average, treatment is continued for around fifteen days [6, 8–11]. Regarding corticosteroid therapy, although it is commonly recommended, its efficacy remains difficult to establish, as only a few teams, such as that of Travis *et al.*, [11], have used it. The need for surgical interventions, such as laminectomy, is often debated due to the high rate of postoperative morbidity. Surgical indications are generally limited to cases where severe neurological symptoms persist despite appropriate medical treatment.

Overall, the clinical evolution in children treated with an appropriate substitution therapy is generally favourable, with a resolution of both clinical and radiological symptoms within a few days to weeks [11]. Although no data are available on the risk of recurrence of epidural hematomas in hemophilia patients, some studies have documented the recurrence of intracranial hemorrhages. Consequently, several teams have considered initiating prophylactic treatment following an acute episode. Thus, preventive supplementation could be considered for episodes of epidural hemorrhage.

CONCLUSION

Epidural hematoma is a rare occurrence in hemophiliacs. It should be considered in any case of spinal pain and/or neurological deficit. Spinal magnetic resonance imaging remains the diagnostic test of choice, both for confirming the diagnosis and for assessing the

extent of the lesions and their complications, as well as for post-treatment follow-up.

With replacement therapy, the outcome is favourable in the majority of cases, preventing spinal cord compression and limiting the need for surgery.

This observation, despite its rarity, encourages us to remain vigilant and to consider the diagnosis of hemophilia in a child presenting with motor deficits due to compression from an epidural hematoma.

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12. MRI images courtesy of the Radiology Department at Mohammed VI University Hospital in Oujda.