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Nonoperative management of spontaneous epidural hematoma in hemophilia A

Ankit Thora[1], Nishit Bhatnagar[2]*, Anurag Tiwari[3], Purushotham Lingaiah[2], Nitesh Rustagi[4]

[1]Department of Orthopaedics, Shalby Hospital, Ahmedabad, Gujurat, India
[2]Department Orthopaedics, Maulana Azad Medical College and Lok Nayak Hospital, New Delhi, India
[3]Department of Orthopaedics, All India Institute of Medical Sciences, Bhopal, Madhya Pradesh, India
[4]Department of Orthopaedics, Guru Gobind Singh Government Hospital, New Delhi, India

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***Correspondence:**
Dr. Nishit Bhatnagar,
E-mail: nishitbhatnagar@yahoo.co.in

ABSTRACT

Background: Spontaneous epidural haematoma is a rare clinical presentation and may be associated with coagulation disorders. These hematomas present usually with rapidly developing paraparesis and any delay in diagnosis may be disastrous.
Methods: We included five patients of spontaneous epidural hematomas in hemophilia A with neurological deficit. They were treated non-operatively with factor VIII replacement therapy with oral tranexamic acid and intravenous dexamethasone as per protocol. All patients were followed up for 6 months and assessed for signs of neurological recovery and radiological resolution of hematoma.
Results: All patients had complete motor recovery within 3 months of initiating treatment and showed resolution of hematoma on MRI.
Conclusions: Prognosis following early initiation of correction of factor deficiency is good.

Keywords: Hemophilia, Spontaneous epidural hematoma, Nonoperative

INTRODUCTION

Spontaneous epidural haematoma (SEH) is a rare clinical presentation and may be associated with coagulation disorders, blood dyscrasias and following lumbar punctures.[1-4] Around 2-8% of hemophilic patients develop hematomas of the CNS, of which around 10% are in the spine.[5] These hematomas present usually with rapidly developing paraparesis and any delay in diagnosis may be disastrous.

METHODS

The study was conducted at a tertiary level referral centre which has a regional hemophilia day care centre, between May 2011 and August 2016. All patients of spontaneous epidural hematomas with hemophilia A with neurological deficit were considered for inclusion in the study. Patients with history of trauma, surgically intervened cases were excluded from the study. Patients unwilling to participate in the study were also excluded. Written informed consent was obtained from each of the patients authorizing treatment, photographic documentation and radiographic examination. An Institutional Review Board clearance was obtained.

The patients age, severity of hemophilia, neurological status prior to initiation of treatment, imaging investigations, treatment received and pattern of recovery was recorded. All patients underwent a magnetic resonance imaging of the spine to localize the site and extent of hematoma. Factor VIII levels and coagulation studies were carried out.

The patients were managed by non-operative means, by giving factor VIII correction. As per the severity of hemophilia and weight of the patient the dose of factor VIII replacement was calculated. For the first 2 weeks, 100% factor VIII correction was given, which was subsequently tapered and finally stopped after 4 weeks as given in Figure 1. MRI was repeated at 4 weeks, just prior to discontinuation of factor VII replacement to look for any residual hematoma.

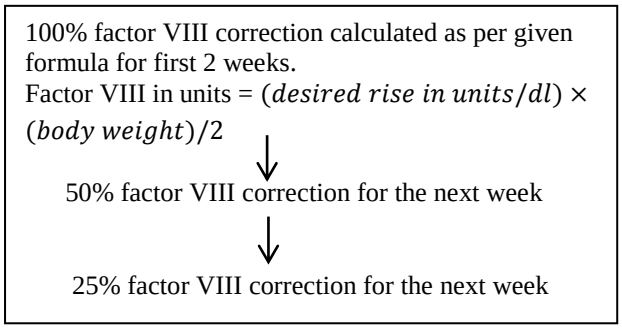

Figure 1: Flowchart of factor VIII replacement for spontaneous epidural hematoma in hemophilia A.

Patients were also given oral tranexamic acid (20 mg/kg/day in three divided doses) till resolution of hematoma on follow up MRI. In addition, patients with cord edema on MRI were given intravenous dexamethasone (0.1 mg/kg/day) for 5 days followed by gradual tapering over the next 8 days. The patients were admitted in the hospital for a period of 4 weeks, during which daily monitoring of neurological status was done. After disharge the patients were followed up for neurological assessment at 2 weekly intervals for 1 month and then at monthly intervals till 6 months.

A literature review using PubMed with several combinations of the following key words, "hemophilia, nonoperative, conservative, epidural hematoma and spontaneous" was performed to further identify similar reported cases. Thirteen cases of spontaneous epidural hematoma in hemophiliacs were identified in the English language.

RESULTS

Five patients of hemophilia A that developed spontaneous epidural hematoma were included in the study. A summary of the 5 cases is shown in Table 1.

The patients were between the age of 10 to 29 years. The patients presented to us between 3 days to 2 weeks after onset of symptoms. Two out of the five were not known cases of hemophilia. Three patients had severe hemophilia (<1% factor VIII levels) and two had moderate hemophilia (between 1-5% factor VIII levels). All three patients with severe degree of hemophilia had complete spastic paraplegia with paralysis of bladder and bowel functions. The other two patients with moderate degree of hemophilia had spastic paraparesis without involvement of bladder or bowel functions. Three patients had hematomas in the lower thoracic and thoracolumbar junction. The others were localised to the upper thoracic and mid-thoracic region. Paralysis was sudden onset in all the patients, and appeared without any history of trauma, back pain, or any constitutional symptoms. None of the patients had any spinal deformity or tenderness.

Plain radiographs of the involved region of the spine did not reveal any abnormality in any of the patients. An MRI was done in all patients prior to starting to treat. It revealed well defined lentiform shaped collection in the epidural space, showing heterogenous hyperintensity on T2w images as shown in Figure 2. MRI of three of the patients also revealed hyperintense foci in the spinal cord on T2w images, suggestive of cord edema. An MRI was repeated 4 weeks after initiation of treatment, which revealed resolution of hematoma in all cases, and mild persistent cord edema in 2 patients as given in Figure 3.

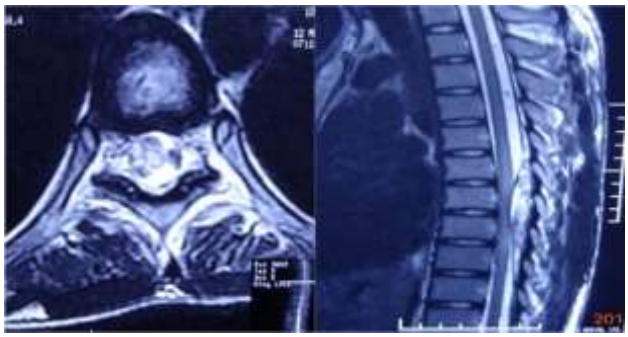

Figure 2: Axial and sagittal T2w images showing a well-defined lentiform shaped collection in the posterior epidural space with heterogenous hyperintense signal and a few hypointense foci within the collection. The spinal cord also showed hyperintense foci at the level of maximal cord compression.

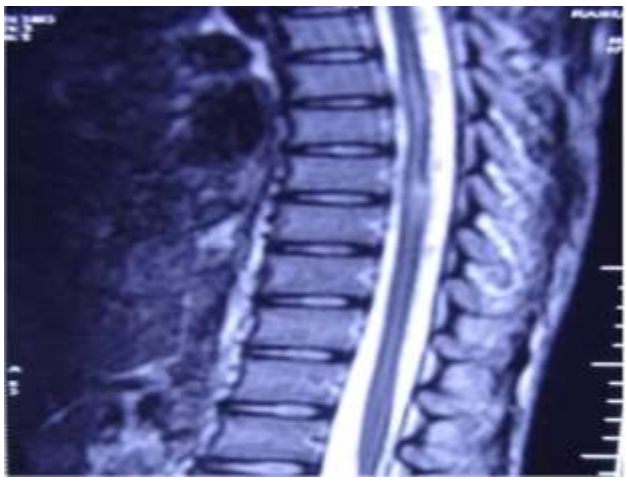

Figure 3: Sagittal T2w image showing resolution of the epidural hematoma with residual slight focal expansion and ill-defined hyperintensity of the cord.

First signs of neurological recovery appeared between 1-4 weeks after initiation of factor VIII replacement therapy. All patients had complete recovery of motor power by 3 months. At 6 months follow up all patients were able to walk unaided, inspite of some residual hypertonicity in 3 patients.

Table 1: Summary of cases in the series.

Age (years)	Hemophilia status	Hemophilia severity	Level	Neuro logical deficit	Management	Time to beginning of recovery	Follow up MRI	Recovery
16	Not previously known	Factor levels <1%	T7-T11	Complete paraplegia	Factor VIII, tranexamic acid and dexamethasone	4 weeks	Complete resolution of hematoma	Complete with residual spasticity at 3 months
12	Previously known	Factor levels <1%	T5-T6	Complete paraplegia	Factor VIII, tranexamic acid and dexamethasone	10 days	Complete resolution of hematoma, some cord edema	Complete , with residual spasticity at 2 months
22	Previously known	Factor levels <3%	T11-L1	Paraparesis, bladder control preserved	Factor VIII and tranexamic acid	2 weeks	Complete resolution of hema-toma	Complete at 3 months
10	Not previously known	Factor levels <3%	C7-T3	Paraparesis, bladder control preserved	Factor VIII and tranexamic acid	2 weeks	Complete resolution of hema-toma	Complete at 3 months
29	Previously known	Factor levels <1%	T8-L1	Complete paraplegia	Factor VIII, tranexamic acid and dexamethasone	1 week	Complete resolution of hematoma, some cord edema	Complete motor recovery, with residual spasticity and partial numbness at 3 months

DISCUSSION

Hemophilia is the most common hereditary bleeding disorder, with an incidence of 0.7-0.8/10,000.[6] Hemophilia A is an X-linked bleeding disorder characterised by the deficiency of clotting factor VIII. Hemophilia is classified as severe if factor level is <1%, moderate if level is between 1% and 5% and mild when it is >5%.[7]

CNS bleeding is one of the most dreaded life-threatening complications of hemophilia. Though, some authors have also reported a chronic course of spinal epidural hematoma in hemophilia A, they usually have a rapid course.[8] CT scan may be done to look for any bony changes, but the investigation of choice remains non-contrast MRI of the spine which can detect even small amounts of epidural collection.

There are reports of medical management and combined medical and surgical management for such scenarios in literature as presented in Table 2. The perioperative risks of operating a hemophilic patient vis-à-vis risk of permanent neurological deficit in conservative management makes treatment complicated and controversial.

Rathi and Rathi described a case of spontaneous epidural hematoma compressing the cervical spinal cord in a 3-month old infant who presented with quadriparesis and non-passage of faeces for 3 days.[9] The child was not a known case of hemophilia, and had undergone circumcision and received intramuscular injections

uneventfully. Factor VIII levels were found to be <1%. The child was managed non-operatively with recombinant factor VIII for 2 weeks followed by prophylactic factor VIII treatment for 6 months.

The patient started showing neurological improvement from 4th day of treatment and had complete neurological recovery by the end of 3 weeks.

Table 2: Case reports of spontaneous epidural hematomas in hemophiliacs.

Year	Author	Age	Level	Neurological deficit	Management	Type of hemophilia	Recovery
1993	Noth[18]	6 months		-	Non operative	-	Complete
1994	Sheikh[11]	7 years	Extensive	-	Non operative	B	Complete
1998	Meena[19]	-	Cervical	-	Operative	B	-
2006	Balkan[1]	17 years		Paraparesis	Non operative	B	Complete ~ 3 weeks
2007	Kubota[12]	4 months	C4-S1	Incomplete left limb paralysis	Non operative	-	Complete ~ 2 years
2010	Rathi[9]	3 months	C2- D5	Quadriparesis	Non operative	A	Complete ~ 3 weeks
2010	Kiehna[5]	5 months	C1 – cauda equina	UMN signs	Non operative	A	Complete
2011	Borkar[10]	5 years	L4- S2	Paraparesis (2/5)	Non operative	B	Complete ~ 1 month
2012	Sheikh[20]	6 – 20 years	Cervical	-	3 patients operated 1 non operative	-	Complete
2015	Ahn[21]	74 years	L1- L2 and L4- L5	Radiating pain, claudication and tingling	Operative	A	Complete

Borkar et al also observed the favourable outcome of nonoperative management of spontaneous epidural hematoma in children with haemophilia B.[10] They described the case of a 5 year old child, a known case of haemophilia B, who developed a spontaneous epidural hematoma in the lumbosacral spine with paraparesis. Patient showed good neurological recovery following a 2 week replacement course of recombinant factor IX. Prolonged prophylactic factor IX replacement was not given, but the hematoma did not recur.

Kiehna et al also concluded in their review of spinal EDH in pediatric hemophilia patients that correction of the coagulaopathy by factor replacement and close observation is a safe and effective treatment.[5] They stressed on timely factor replacement, maintenance of adequate factor levels, close monitoring of hemodynamic and neurological status. A multidisciplinary approach involving pediatric hematologist and neurosurgeon is recommended.

Sheikh and Abildgaard emphasized the usefulness of serial MRI scans to monitor progress of disease in patients undergoing non-operative line of management with aggressive factor replacement therapy.[11]

Kubota et al also concluded that non-surgical management is safe for spinal extradural hematoma in haemophiliacs. Surgical management can be attempted under cover of factor replacement if there is a rapidly progressive neurological deficiency.[12]

Falavigna et al and Lo et al concluded separately in their reports on spontaneous idiopathic spinal extradural hematomas that MRI should be performed and the need for surgical intervention in the form of laminectomy should be dictated by location of hematoma and speed of progression of neurological deficit.[13,14] Cervical and thoracic lesions with rapidly progressing neurological deficit is regarded as a surgical emergency. Early surgical intervention by decompressive laminectomy and hematoma evacuation is the treatment of choice in such patients. Nonoperative line of management is favoured when neurological deficit improves in the early phase of management.

Fukui et al in their series of eight patients of acute spontaneous spinal epidural hematomas recommend nonoperative management for patients with minimal neurological deficits, inspite of cord compression on MRI imaging and early surgical decompression for patients with significant neurological deficit before interference with blood supply of spinal cord.[15]

Suzer et al also similarly recommend conservative management in select patients of spontaneous epidural hematoma who either have minimal neurological deficit or show early neurological recovery.[16]

Early conservative management involves factor replacement to achieve a level of 100 U/dl and then maintenance at this level for a minimum period of 2 weeks. Factor VIII dose can be calculated using the formula: factor VIII in units = U/dl desired rise × body weight × 0.5.[17] Prognosis following early initiation of correction of factor deficiency is good. Surgical decompression under the cover of the deficient factor should be considered only in those patients who do not respond to conservative treatment or in cases of delay in diagnosis.[12]

Our study was limited by a small sample size due to the rarity of such a presentation. However, it highlights the fact that the possibility of hemophilia must be entertained even if a clear history suggestive of coagulation deficiency is not available. Epidural hematomas in hemophilia patients need not be decompressed surgically. Adequate restoration of clotting factors can also lead to spontaneous resolution of the hematoma followed by neurological recovery

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