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Case Report

Spontaneous spinal epidural hematoma in a pediatric hemophiliac

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ABSTRACT

Background: Spontaneous spinal epidural hematomas (SSEH), unrelated to trauma, epidural anesthesia, or surgery, are rare in the pediatric population. Here, a 1-year-old male with hemophilia presented with a magnetic resonance (MR)-documented SSEH and was successfully treated with a C5-T10 right hemilaminectomy.

Case Description: A 1-year-old male with hemophilia presented with quadriparesis. The holo-spine magnetic resonance imaging with contrast showed a posterior cervicothoracic compressive epidural lesion extending from C3 to L1 consistent with an epidural hematoma. He underwent a C5 to T10 right-sided hemilaminectomy for clot removal, following which his motor deficits fully resolved. A literature review of SSEH attributed to hemophilia revealed that 28 of 38 cases were effectively treated conservatively, while only 10 cases warranted surgical decompression.

Conclusion: Select patients with SSEH attributed to hemophilia with severe MR-documented cord/cauda equina compromise and significant accompanying neurological deficits may require emergent surgical decompression.

Keywords: Epidural, Hematoma, Hemophilia, Pediatric, Spine

INTRODUCTION

Spontaneous spinal epidural hematomas (SSEH) that develop in the extradural space without accompanying histories of trauma, epidural anesthesia, or surgery represent <1% of all space-occupying spinal lesions. In adults, SSEH occurs with a frequency of 0.1 cases/100,000/year, and account for 0.6% of all spinal hemorrhages; in children, their incidence remains unclear.[19,31,34] Conservative treatment (i.e., infusion of recombinant coagulation factors, corticosteroids) may suffice where there are no major neurological deficits, while surgery is warranted for those with significant paresis.[4,6,8] Here, a 1-year-old hemophiliac male with a significant C5-T10 SSEH

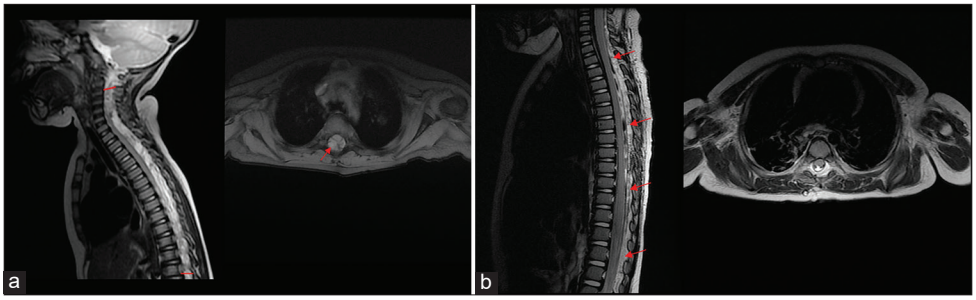

Figure 1: Preoperative holo-spine magnetic resonance imaging showing a large cervicothoracic posterior epidural hematoma, hyperintense in T2-weighted images (a, red arrow in axial plane) and hypointense in T1-weighted images (b, red arrows in sagittal plane).

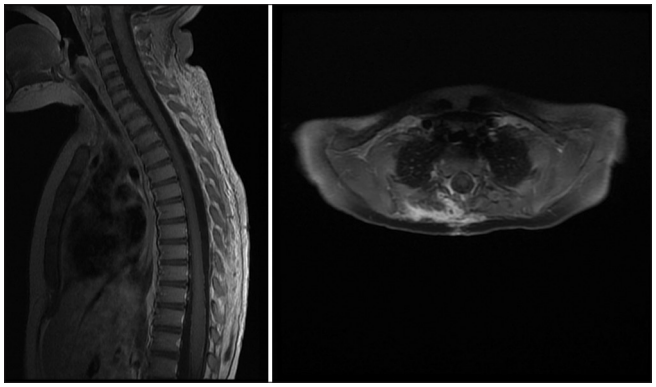

Figure 2: Postoperative holo-spine T1-weighted magnetic resonance imaging sequences showing a complete spontaneous spinal epidural hematomas removal and decompression of the spinal cord.

that contributed to a severe quadriparetic deficit warranted emergent surgical decompression.

CASE DESCRIPTION

A 1-year-old male with hemophilia A (i.e., severe deficit of factor VIII) presented with the acute and spontaneous onset of severe quadriparesis. Laboratory tests revealed a low hemoglobin (8.5 g/dL), low hematocrit (25.6%), a mildly elevated international normalized ratio (1.34) and partial thromboplastin time (37 s), with reduced prothrombin activity (65 s). The holo-spine magnetic resonance (MR) with contrast showed a large cervicothoracic posterior epidural compressive lesion extending from C3 to L1 that was consistent with an epidural bleed; it was hypointense on T1, and hyperintense on T2-weighted images [Figures 1a and b]. An urgent C5-T10 right hemilaminectomy was performed along with the simultaneous administration/infusion of VIII replacement. The 48-h postoperative cervicothoracic magnetic resonance imaging documented complete epidural hematoma removal with no increased signal in the cord (i.e., consistent with the absence of a significant ischemic injury) [Figure 2]. Postoperatively, the factor VIII deficit was confirmed and he was treated with continued VIII factor infusions (i.e., 500 UI twice a day, for a total of 6 days). Five years later, he exhibited no residual deficit and demonstrated no spinal deformity.

DISCUSSION

Hemophilia is one of the most common hereditary X-linked bleeding disorders attributed to a deficiency of factor VIII. Some patients with hemophilia acutely develop SSEH and present with the rapid onset of paralysis correlated with the levels of MR-documented spinal hemorrhages.[20,22,30,34] Kiehna *et al*. strongly suggested conservative treatment for SSEH in hemophiliac pediatric patients; they cited their high success rates for medical therapy alone.[19] Other studies also advocated conservative treatment of SSEH utilizing factor VIII replacement infusions; these measures avoided potential surgical complications.[21] Further, Carlhan-Ledermann *et al*. presented a case of SSEH successfully treated conservatively, but also noted that some patients' hemorrhages/severe neurological deficits warranted acute surgical intervention.[8]

Surgery

Most adults with SSEH will undergo decompressive laminectomies. However, children may occasionally be managed with less extensive partial laminectomies or single/multiple short-segment laminoplasties.[23] Notably, in the pediatric population, multilevel surgical decompressions post a greater risk of future instability/deformity. In our case, the urgent C5-T10 surgery consisted of a right-side partial hemilaminectomy, and the patient, 5 years later, exhibited no deformity. In a review of the literature, 28 of 38 pediatric patients with SSEH were successfully managed conservatively, while only 10 cases warranted surgery [Table 1].[1-3,5,7,9-18,24-29,32,33,35-40] Notably, there was just one death (i.e., expiration due to a postoperative complication) and just one other patient who requiring additional surgery to address a recurrent spinal epidural hematoma [Table 2].

Table 1: Literature review of reported cases of hemophilic pediatric patients with SSEH.

S. No.	Author	Year	Number of patients	Age	Sex	Hemophilia	Signs of myelopathy	Extension of the hematoma	Surgery	Adverse events	Outcome
1.	Jones and Knighton[17]	1956	1	12 y	M	A	Yes	C6-T2	Yes	No	Persistence of paraplegia
2.	Stanley and Mccomb[35]	1983	1	13 y	M	A	Yes	T5-T8	Yes	No	Improvement with residual mild distal weakness
3.	Narawong *et al*.[24]	1988	2	4 y	M	A	No	T1-T5	No	-	Complete recovery
				8 m	M	A	No	C2-T3	No	-	Complete recovery
4.	Faillace *et al*.[11]	1989	1	3 m	M	-	Yes	T8-L4	Yes	No	Improvement with residual paraplegia with Th5 sensory level
5.	Hamre and Haller[12]	1992	1	9 y	M	B	Yes	C2-T6	No	-	Neurological improvement
6.	Noth *et al*.[26]	1993	1	6 m	M	A	Yes	C2-T6	No	-	Complete recovery
7.	Sheikh and Abildgaard[32]	1994	1	7 y	M	B	Yes	C3-L2	No	-	Neurological improvement
8.	Abdelaal *et al*.[1]	1994	1	5 y	M	A	Yes	C0-T8	Yes	No	Complete recovery
9.	Travis *et al*.[38]	1996	1	20 m	M	A	No	L4-L5	No	-	Complete recovery
10.	Hutt *et al*.[14]	1996	1	11 m	M	A	No	T9-T12	No	-	Complete recovery
11.	Varsha *et al*.[39]	2000	1	10 y	M	A	Yes	C3-C7	No	-	Complete recovery
12.	Iwamuro *et al*.[16]	2004	1	7 m	M	B	Yes	T1-L1	No	-	Complete recovery
13.	Balkan *et al*.[3]	2006	1	17 y	M	B	Yes	C6-T12	No	-	Complete recovery
14.	Cuvelier *et al*.[9]	2006	1	10 m	M	A	Yes	C2-T8	Yes	No	Complete recovery
15.	Bisson *et al*.[5]	2007	1	7 y	M	B	-	C2-T3	No	-	Complete recovery
16.	Kalina *et al*.[18]	2007	1	6 m	M	A	No	C2-L4	No	-	Complete recovery
17.	Kubota and Miyajima[21]	2007	1	4 m	M	A	Yes	C4-S1	No	-	Complete recovery
18.	Heer *et al*.[13]	2008	1	12 y	M	A	Yes	C5-C6	Yes	Yes: re-do surgery	Neurological deterioration, severe diparesis and paraplegia
19.	Rois *et al*.[30]	2008	1	13 y	M	A	Yes	D5-D6	Yes	No	Complete recovery
20.	Morsing *et al*.[23]	2009	1	1 y	-	A	No	C1-S1	No	-	Complete recovery
21.	Kiehna *et al*.[19]	2010	1	5 m	M	A	No	C1-cauda	No	-	Complete recovery

(*Contd*...)

Table 1: (*Continued*).

S. No.	Author	Year	Number of patients	Age	Sex	Hemophilia	Signs of myelopathy	Extension of the hematoma	Surgery	Adverse events	Outcome
22.	Rathi and Rathi[29]	2010	1	3 m	M	A	Yes	C2-D5	No	-	Complete recovery
23.	Tagaya *et al*.[36]	2010	1	4 m	M	A	Yes	C2-cauda	No	-	Complete recovery
24.	Borkar *et al*.[6]	2012	1	5 y	M	B	Yes	L4-S2	No	-	Neurological improvement
25.	Nirupam *et al*.[25]	2013	1	9 y	M	B	Yes	C6-T12	Yes	Yes	Dead
26.	Per *et al*.[28]	2014	1	2 y	M	A	No	C3-T2	No	-	Complete recovery
27.	Iizuka *et al*.[15]	2014	1	5 y	M	A	Yes	C2-T1	Yes	No	Complete recovery
28.	Erkutlu *et al*.[10]	2015	1	5 y	M	A	Yes	D9-D10	No	-	Complete recovery
29.	Oymak *et al*.[27]	2016	1	10 m	M	A	Yes	C1-cauda	No	-	Complete recovery
30.	Thora *et al*.[37]	2017	3	16 y	-	A	Yes	T7-T11;	No	-	Recovery with residual spasticity
				12 y	-	A	Yes	T5-T6	No	-	Recovery with residual spasticity
				10 y	-	A	Yes	C7-T3	No	-	Complete recovery
31.	Singh *et al*.[33]	2019	1	8 y	M	A	Yes	C6-C7	No	-	Complete recovery
32.	Boyadzhiev *et al*.[7]	2019	1	4 m	M	A	No	C1-S1	Yes	No	Complete recovery
33.	Carlhan-Ledermann *et al*.[8]	2020	1	4 m	M	A	No	C3-C5	No	-	Complete recovery
34.	Villarreal-Martínez *et al*.[40]	2021	1	4 y	M	A	No	C4-T4	No	-	Complete recovery
35.	Acharya *et al*.[2]	2021	1	12 y	M	A	Yes	C2-D8	No	-	Complete recovery
36.	Present case	2023	1	1 y	M	A	Yes	C5-D10	Yes	No	Complete recovery

SSEH: Spontaneous spinal epidural hematomas, M: Male, y: Years, m: Months

Table 2: Summary of literature review data.

Patients' demographics	Values
Total number of patients	38
Mean age and range (years)	5.5±5.25 (range 0.2–17)
Sex	34 males (89.4%), 0 females (0%), 4 not reported (10.4%)
Hemophilia	30 patients type A (78.9%), 7 patients type B (18.4%), 1 patient not reported (2.6%)
Signs of mielopathy	Present in 26 patients (68.4%), absent in 11 patients (28.9%), not reported in 1 patient (2.6%)
Surgery	10 patients (26.3%)
Conservative treatment	28 patients (73.7%)
Adverse events	In 2 patients, recurrence and exitus (5.3%)
Outcome	Complete recovery in 28 patients (73.7%), neurological improvement in 8 patients (21%), persistence of neurological deficits in 1 patient (2.6%), worsening symptoms in 1 patient (2.6%)

CONCLUSION

A 1-year-old male with hemophilia presented with acute quadriparesis attributed to a MR-documented C5-T10 thoracic SSEH resulting in marked cord compression. Following a right-sided C5-T10 hemilaminectomy, the patient was neurologically intact and remained intact for 5 subsequent years during which he developed no spinal deformity.

Ethics approval

There is no ethical issue in this paper.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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