Idiopathic Spinal Subarachnoid Hemorrhage: A Case Report and Review of the Literature

Justin M. Moore[1] Rondhir Jithoo[1] Peter Hwang[1]

[1] Department of Neurosurgery, The Alfred Hospital, Melbourne, Victoria, Australia

Global Spine J 2015;5:e59–e64.

Address for correspondence Justin M. Moore, MD, PhD, Department of Neurosurgery, 1st floor, Old Baker Building, The Alfred Hospital, Melbourne, Victoria, Australia 3004 (e-mail: justinmurraymoore@gmail.com).

Abstract

Study Design Case report.

Objective Spinal subarachnoid hemorrhage (SSAH) makes up less than 1.5% of all the cases of subarachnoid hemorrhage. Most cases of spontaneous SSAH occur in association with coagulopathy, lumbar punctures, or minor trauma. Idiopathic SSAH is extremely rare with only 17 cases published. Idiopathic SSAH presents a diagnostic dilemma, and the appropriate investigations and treatment remain a matter of controversy. We report a case of idiopathic SSAH and a review of the literature regarding its clinical presentation, diagnosis, and treatment.

Methods A 73-year-old woman presented to the emergency department after spontaneously developing severe right leg and lower back pain while bending over to vomit. After a review of the patient's history and examination, the magnetic resonance imaging (MRI) of the thoracolumbar spine revealed T1 hyperintensity and T2 hypointensity, a diffusion-restricted collection at the T11–T12 level, and a posterior collection from L3 to S1 producing a mild displacement of the thecal sac.

Results The patient was taken for an L5 laminectomy. Intraoperatively, rust-colored, xanthochromic fluid was drained from the subarachnoid space, confirming SSAH. The thecal sac was decompressed. The cultures and Gram stains were negative. Computer tomography (CT) and CT angiography of the brain were normal. She recovered postoperatively with resolution of the pain and no further episodes of hemorrhage after 2 years of follow-up. Repeat thoracolumbar MRI, selective spinal angiogram, and six-vessel cerebral angiogram did not reveal pathology.

Conclusion We suggest a clinical algorithm to aid in the diagnosis and management of such patients.

Keywords
- spinal
- subarachnoid hemorrhage
- treatment
- diagnosis
- idiopathic
- spontaneous
- guideline

Introduction

Spinal subarachnoid hemorrhage (SSAH) makes up less than 0.05 to 1.5% of all cases of subarachnoid hemorrhage (SAH).[1] SSAH is associated with trauma, vascular malformations, neoplastic lesions, autoimmune disorders, coarctation of the aorta, coagulopathy, rheumatologic diseases, and drug and alcohol use and withdrawal.[1–3] Most cases of spontaneous SSAH occur in association with coagulopathy, lumbar punctures, or minor trauma.[4–6] Idiopathic SSAH is extremely rare with only 17 cases published (►**Table 1**).[2,5,7–18] We report a case of idiopathic SSAH.

Case Report

A 73-year-old woman presented to the emergency department of the Alfred Hospital after spontaneously developing severe right leg and lower back pain while bending over to

received
February 11, 2014
accepted after revision
December 31, 2014
published online
April 29, 2015

DOI http://dx.doi.org/10.1055/s-0035-1546416.
ISSN 2192-5682.

© 2015 Georg Thieme Verlag KG Stuttgart · New York

License terms

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

vomit. There was no history of trauma or use of anticoagulants or antiplatelet agents. She had no headache or other features of cranial SAH. She had a history of hip replacement and hypertension that had been successfully treated. On examination she was febrile and had a positive straight leg raise test on the right at 60 degrees. She had no neurologic deficit, and her coagulation profile was unremarkable.

Magnetic resonance imaging (MRI) of the thoracolumbar spine revealed T1 hyperintensity and T2 hypointensity, a diffusion-restricted collection (27 × 4 mm) at the T11–T12 level, and a posterior collection from L3 to S1 producing a mild displacement of the thecal sac (►**Fig. 1**). The collection had a suspicious sedimentary component. As the imaging characteristics were suggestive of a hematoma or an abscess and the patient was febrile, the patient was taken for an L5 laminectomy, which provided access to the posterior collection. Intraoperatively, the thecal sac was observed to be dark blue in color. A pediatric needle was introduced into the subarachnoid space and rust-colored xanthochromic fluid was drained. The thecal sac was decompressed following the procedure. As she was neurologically intact and the thecal sac was decompressed, no further intervention was deemed necessary. Cultures and Gram stain were negative. Computer tomography (CT) and CT angiography of the brain was normal. She recovered well postoperatively with resolution of her pain and has had no further episodes of hemorrhage after 2 years of follow-up. Repeat thoracolumbar MRI and selective spinal angiogram and six-vessel cerebral angiogram did not reveal pathology.

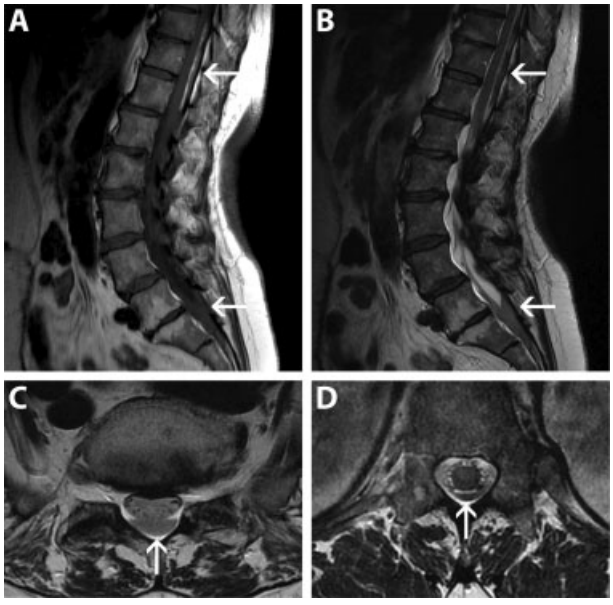

Fig. 1 (A) T1-weighted sagittal magnetic resonance imaging (MRI) reveals hyperintensity at the T11 and T12 levels (top arrow) and a hypointense collection at the lumbosacral level (bottom arrow). (B) T2-weighted sagittal MRI reveals hypointensity at the T11 and T12 (top arrow) levels and a hypointense collection at the L5–S1 level (bottom arrow). (C) T2-weighted axial MRI reveals hypointensity at the L5–S1 level (arrow). (D) T2-weighted axial MRI reveals T12 hypointensity (arrow).

Discussion

Idiopathic SSAH is extremely rare with only 17 case reports in the literature. Of these 17 reports, a number have not been completely investigated, generally due to patient refusal to undergo a spinal angiogram (►**Table 1**).[7,19] The diagnosis of spontaneous SSAH is a diagnosis of exclusion. The most common causes of SSAH are secondary to trauma, vascular malformation, and tumors.[1] The majority of idiopathic SSAH occurs in the thoracic and lumbar spine (►**Table 1**).[1] These patients typically present with acute back pain often in association with sensory disturbance, paralysis, and sphincter abnormalities.[1,11,13] Of the 17 previously published cases, 12 reported sudden-onset back or neck pain (►**Table 1**). In our case, the patient's symptoms developed while bending over to vomit. Several case reports have described an idiopathic SSAH in the setting of squatting or other activities, which, like our case, increase venous pressure.[1]

To safely make this diagnosis, we recommend complete cranial and spinal angiograms and a spinal MRI to rule out small spinal arteriovenous malformations. Additionally, an MRI can be helpful in defining the level and position of the SSAH as well as ruling out small tumors and trauma as possible causes.[1] Acutely, the SAH is hyperintense or isointense on T1 and hyperintense or hypointense on T2 MRI.[11,19] A subacute hemorrhage becomes hyperintense or isointense on T1-weighted MRI and T2 reveals a hyperintense signal due to the strongly paramagnetic methemoglobin.[10] Our case highlights the limitations of MRI in the presence of SSAH where hyperacute SAH may be difficult to distinguish from a subdural hematoma or an abscess.

The requirement for cranial imaging is currently a matter of debate, with some authors suggesting that in the absence of cranial symptoms intracranial imaging may not be mandatory.[10] We, in light of recent literature, would recommend cranial imaging, either MR angiography, CT angiography, or a formal angiogram, which may identify an underlying abnormality and enables assessment for intracranial SAH and the potential for vasospasm.[4]

Case management depends on the neurologic status of the patient, not on the extent and location of the hematoma. The management of these patient has usually involved an operation.[1] Komiyama et al postulated that ventral hematomas were typically benign, whereas dorsally located hematomas typically resulted in symptoms as a result of a dynamic interaction between the cerebrospinal fluid and the hematoma and required surgical management.[1] Our case supports a benign natural history for at least some of these dorsal lesions.[19] Surgical decompression is warranted only if the spinal cord is compressed by the mass effect from hematoma, if the patient has severe or deteriorating neurologic symptoms, or if there is a diagnostic dilemma as to the pathology of the lesion on imaging studies, as in our case.[1] The outcome of treatment for patients with satisfactory neurologic status on presentation is generally good for those treated medically or surgically in over 90% of cases (►**Table 1**).[5] Sunada et al emphasized the importance of early decompression once the patient deteriorates neurologically.[13] For extensive

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

Table 1 Summary of cases of spinal SAH reported in the literature

Presentation	Age	Sex	Location	Labs	Anti-coagulant	Diagnostic imaging	Spinal DSA	Cranial	LP	Site	Management	Outcome	Author	Year
Sudden-onset back pain	73	F	T11–S1	Normal	Nil	MRI	DSA: Neg	CTB/CTA/DSA: Neg	SAH (intraop)	P	Operative	Asymptomatic, resolution on MRI at 6 mo	Our case	2015
Sudden-onset back pain and headache postcoughing	66	F	L1–L2	Normal	Nil	MRI	DSA: Neg	CTB/DSA: Neg	SAH	A	Conservative	Asymptomatic, resolution on MRI at 1 mo	Oji et al[8]	2013
Sudden-onset back pain, numbness, and paraplegia	37	F	C6–T6	Normal	Nil	MRI	Not completed	CTB/MRI: Neg	SAH (intraop)	A	Operative	Paralysis persisted, numbness improved	Sasaji et al[9]	2013
Subacute back pain (1 wk), headache, and lower limb pain	66	M	L1–L5	Normal	Nil	MRI	Refused	CTB: Neg	SAH	A	Conservative	Asymptomatic, resolution on MRI at 5 mo	Kakitsubata et al[10]	2010
Subacute-onset back pain (over hours)	58	F	L2–S2	Normal	Nil	MRI	Not completed	CTB: Neg	Not completed	P	Conservative	Asymptomatic, resolution on MRI at 6 mo	Kim and Lee[7]	2009
Sudden-onset back pain, lower limb paresthesia	48	M	T12–L3	Normal	Nil	MRI	Not completed	Not completed	SAH	A	Conservative	Asymptomatic, resolution on MRI at 1 mo	Kim et al[11]	2004
Sudden-onset neck pain, occipital headache	55	M	T8–T12	Normal	Nil	MRI	DSA: Neg	Not completed	SAH	P	Conservative	Asymptomatic, resolution on MRI at 1 mo	Ruelle et al[2]	2001
Sudden-onset back pain, occipital headache, urinary retention	61	M	L1–L2	Normal	Nil	MRI	DSA: Neg	CTB: Neg	SAH	A	Conservative	Asymptomatic, resolution on MRI at 1 mo	Ruelle et al[2]	2001
Subacute-onset neck pain (7 d), quadriparesis, and urinary retention	43	M	C4–C5	No reported	Nil	MRI	DSA: Neg	Not completed	Not completed	P	Operative	Died from respiratory complications	Romano et al[12]	1999
Sudden-onset back pain	30	F	C7–T6	Normal	Nil	MRI	DSA: Neg	CTB/DSA: Neg	SAH	A	Conservative	Asymptomatic, resolution on MRI at 1 mo	Komiyama et al[1]	1997
Sudden-onset back pain	56	F	T11–L2	Normal	Nil	MRI	DSA: Neg	CTB/MRI/DSA: Neg	SAH	A	Conservative	Asymptomatic, resolution on MRI at 1 mo	Komiyama et al[1]	1997
Sudden-onset back pain, paraplegia, and voiding difficult	66	F	T2–T6	Not reported	Nil	MRI	DSA: Neg	DSA: Neg	Not completed	P	Operative	Mobilizing independently	Sunada et al[13]	1995
Sudden-onset back and neck pain, sudden spastic paresis, and bowel/bladder abnormalities	56	F	T12–L3	Not reported	Nil	MRI/ myelography	DSA: Neg	Not completed	SAH	P	Operative	Mobilizing independently	Hiyama et al[14]	1990

(*Continued*)

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

Table 1 (*Continued*)

Presentation	Age	Sex	Location	Labs	Anti-coagulant	Diagnostic imaging	Spinal DSA	Cranial	LP	Site	Management	Outcome	Author	Year
Subacute-onset back pain (2 mo) and progressive paralysis (10 d)	55	M	T12	Normal	Nil	Myelography	Not completed	Not completed	Not completed	P	Operative	Mobilizing independently	Gambacorta et al[15]	1987
Sudden-onset paresthesia and voiding difficulties	40	M	T11–L1	Normal	Nil	Myelography	Not completed	Not completed	Not completed	P	Operative	Mobilizing independently	Khosla et al[16]	1985
Sudden-onset back pain with paralysis and urinary retention	–	F	T1–T5	Not reported	Nil	Myelography	DSA: Neg	Not completed	SAH	A and P	Operative	Mobilizing independently, full recovery	Owaki et al[17]	1975
Sudden-onset thoracic pain and voiding difficulties and paralysis	48	M	T6–T9	Normal	Nil	Myelography	Not completed	Not completed	SAH	P	Operative	Asymptomatic	Plotkin et al[18]	1966
Sudden-onset neck and abdominal pain with complete motor/sensory loss and voiding difficulty	81	M	T8–L3	Normal	Nil	Myelography	Not completed	Not completed	SAH	P	Operative	No improvement, paralyzed	Plotkin et al[18]	1996

Abbreviations: A, anterior location; CTA, computer tomography angiography; CTB, computer tomography of the brain; DSA, digital subtraction angiography; intraop, intraoperatively; MRI, magnetic resonance imaging; Neg, negative; P, posterior location; SAH, subarachnoid hemorrhage.

SSAH, the possibility of cerebral vasospasm needs to be considered in the posthemorrhagic period.[4]

The literature suggests that idiopathic SSAH is a nonrecurrent pathology (►**Table 1**).[1,13] Our case, with over 2 years of follow-up and no further episodes of hemorrhage, would support this observation. Some authors have suggested that the pathogenesis of the condition, particularly in iatrogenic SSAH, involves rupture of the radicular arteries and veins.[6] Others have hypothesized that a minor trauma and changes in intrathoracic and intra-abdominal pressure lead to increased luminal pressure within the vessels of the subarachnoid space with subsequent rupture of the vessels.[1] We hypothesize that some of these hemorrhages may be venous in nature as a result of a transient increase in venous pressure; however, this will be difficult to prove, given the rarity of the condition.

It may be difficult to distinguish SSAH, spinal subdural hematoma (SSDH), and spontaneous spinal epidural hematoma (SEDH) clinically, as all can present with sudden-onset back pain with sensorimotor deficits. However, meningism, headache, and metal status changes are more common symptoms of SSAH.[2] The incidence of spontaneous idiopathic spinal hematomas has not been delineated due to the rarity of the condition. However, Kreppel et al performed a large review of all spinal hematomas and found that SSDH makes up less than 5% of spinal hematomas, SSAH makes up 16%, and SEDH makes up 79%.[20] The incidence of spontaneous SEDH has been estimated at ~0.1 per 100,000 patient-years.[21] The pathophysiology of spontaneous SSDH and SEDH is not well understood. It has been theorized that spontaneous SSDH arises following the rupture of the valveless radiculomedullary veins due to increased intrathoracic pressure or a minor trauma, with SSDH that can subsequently break through the arachnoid to enter the subarachnoid space.[22] In regard to spontaneous SEDH, the literature provides evidence of both epidural venous plexus and arterial origin.[13] In general, a progressive neurologic decline, a severe neurologic deficit, and an expanding hematoma on imaging are indications for surgical management. Improving symptomatology or mild stable neurologic deficits can be managed conservatively.[22,23]

To our knowledge, no guidelines exist for the management of these patients. Therefore, we recommend the algorithm in ►**Fig. 2** for the management of patients in whom SSAH is strongly suspected.

Conclusion

Our case highlights the importance of systematically imaging the spinal cord and the brain, in the presence of idiopathic SSAH. We present a clinical algorithm to enable the safe diagnosis and management of patients whose presentation is suggestive for SSAH.

Disclosures
Justin M. Moore, none
Rondhir Jithoo, none
Peter Hwang, none

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

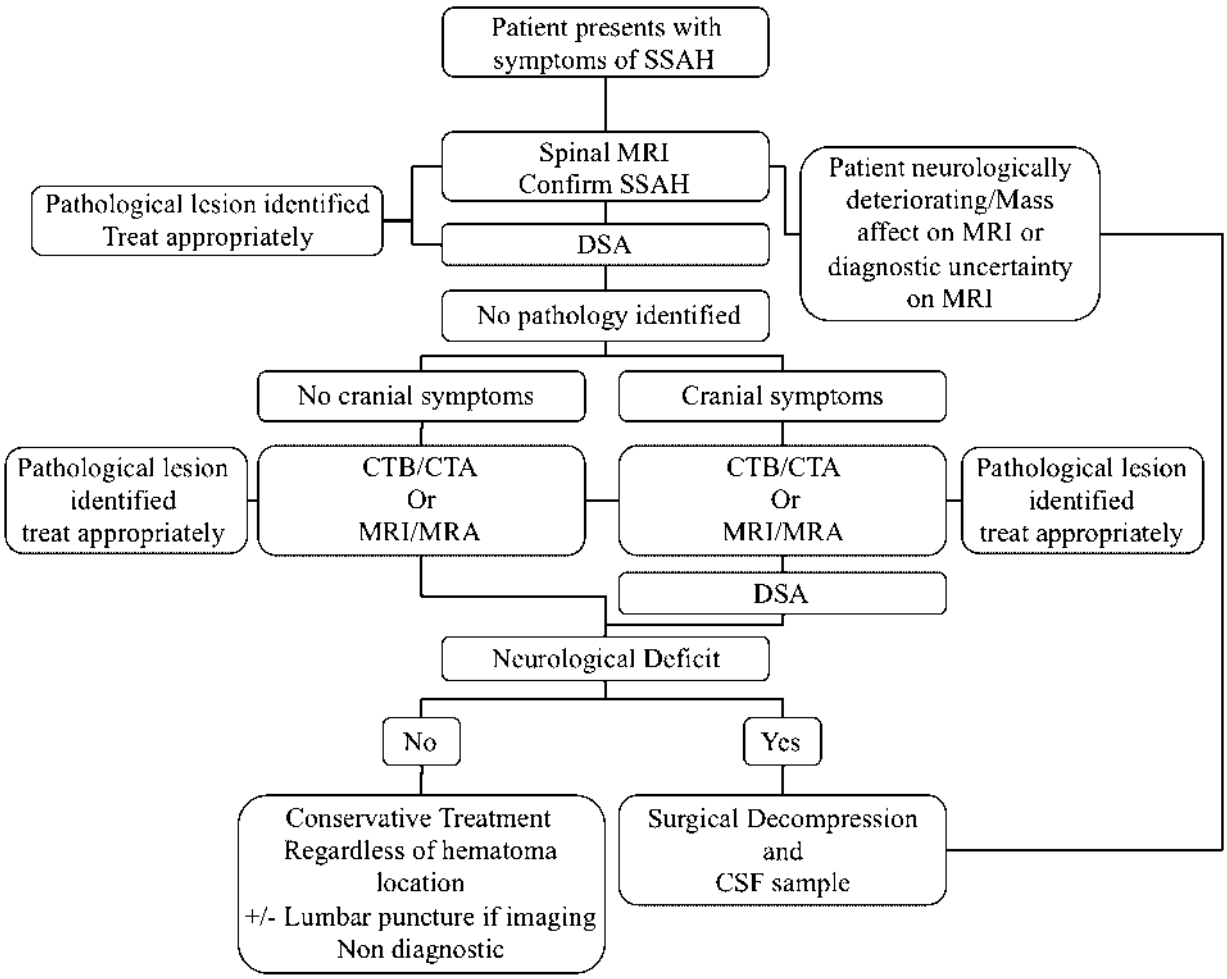

Fig. 2 Algorithm for the diagnosis and management of patients whose clinical presentation is suspicious for SSAH. Abbreviations: CSF, cerebrospinal fluid; CTA, computer tomography angiography; CTB, computer tomography of the brain; DSA, digital subtraction angiography; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; SSAH, spinal subarachnoid hemorrhage.

References

1 Komiyama M, Yasui T, Sumimoto T, Fu Y. Spontaneous spinal subarachnoid hematoma of unknown pathogenesis: case reports. Neurosurgery 1997;41(3):691–693, discussion 693–694
2 Ruelle A, Zerbi D, Andrioli G. Spinal subarachnoid bleeding of unknown etiology. Case reports. J Neurosurg Sci 2001;45(1):53–57
3 Vishteh AG, Theodore N, Spetzler RF. Nontraumatic acute spinal subdural hematoma. J Neurosurg 2000;93(1, Suppl):180–181
4 Shakur SF, Farhat HI. Cerebral vasospasm and ischemia following a spontaneous spinal subarachnoid hemorrhage. Case Rep Med 2013;2013:934143
5 Domenicucci M, Ramieri A, Paolini S, et al. Spinal subarachnoid hematomas: our experience and literature review. Acta Neurochir (Wien) 2005;147(7):741–750, discussion 750
6 Masdeu JC, Breuer AC, Schoene WC. Spinal subarachnoid hematomas: clue to a source of bleeding in traumatic lumbar puncture. Neurology 1979;29(6):872–876
7 Kim JS, Lee SH. Spontaneous spinal subarachnoid hemorrhage with spontaneous resolution. J Korean Neurosurg Soc 2009;45(4): 253–255
8 Oji Y, Noda K, Tokugawa J, Yamashiro K, Hattori N, Okuma Y. Spontaneous spinal subarachnoid hemorrhage after severe coughing: a case report. J Med Case Reports 2013;7:274
9 Sasaji T, Shinagawa K, Matsuya S. Spontaneous thoracic spinal subarachnoid hemorrhage diagnosed with brain computed tomography. Tohoku J Exp Med 2013;231(2):139–144
10 Kakitsubata Y, Theodorou SJ, Theodorou DJ, et al. Spontaneous spinal subarachnoid hemorrhage associated with subdural hematoma at different spinal levels. Emerg Radiol 2010;17(1):69–72
11 Kim YH, Cho KT, Chung CK, Kim HJ. Idiopathic spontaneous spinal subarachnoid hemorrhage. Spinal Cord 2004;42(9):545–547
12 Romano A, Marsella M, Swamy N, de Courten-Myers G, Zuccarello M. Cervical subarachnoid hematoma of unknown origin: case report. Acta Neurochir (Wien) 1999;141(10):1115–1117
13 Sunada I, Akano Y, Kidosaki Y, Shimokawa N, Yamamoto S. Spontaneous spinal subarachnoid hematoma—case report. Surg Neurol 1995;44(2):133–136
14 Hiyama H, Shimizu T, Yato S, Kobayashi N, Ono Y, Kakinoki Y. [Wide-spread spontaneous spinal subarachnoid hematoma. Case report]. Neurol Med Chir (Tokyo) 1990;30(11 Spec No):842–847
15 Gambacorta D, Reale F, De Falco D. Spontaneous chronic spinal subarachnoid hematoma. Report of a case and review of the literature. Spine (Phila Pa 1976) 1987;12(7):716–718
16 Khosla VK, Kak VK, Mathuriya SN. Chronic spinal subdural hematomas. Report of two cases. J Neurosurg 1985;63(4):636–639
17 Owaki K, Nakazawa S, Yajima K, Sugiura K, Yabe Y. [Spontaneous spinal subarachnoidal hematoma—case report (author's transl)]. No Shinkei Geka 1975;3(7):593–597
18 Plotkin R, Ronthal M, Froman C. Spontaneous spinal subarachnoid haemorrhage. Report of 3 cases. J Neurosurg 1966;25(4):443–446
19 Kang HS, Chung CK, Kim HJ. Spontaneous spinal subdural hematoma with spontaneous resolution. Spinal Cord 2000;38(3):192–196

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

20 Kreppel D, Antoniadis G, Seeling W. Spinal hematoma: a literature survey with meta-analysis of 613 patients. Neurosurg Rev 2003; 26(1):1–49
21 Holtås S, Heiling M, Lönntoft M. Spontaneous spinal epidural hematoma: findings at MR imaging and clinical correlation. Radiology 1996;199(2):409–413
22 Payer M, Agosti R. Spontaneous acute spinal subdural hematoma: spontaneous recovery from severe paraparesis—case report and review. Acta Neurochir (Wien) 2010;152(11): 1981–1984
23 Baek BS, Hur JW, Kwon KY, Lee HK. Spontaneous spinal epidural hematoma. J Korean Neurosurg Soc 2008;44(1):40–42

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.