Intraoperative Ultrasound-assisted Treatment of Idiopathic Spontaneous Intramedullary Hemorrhage: A Case Report and Literature Review

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Case report

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Abstract

Background: Idiopathic spontaneous intramedullary hemorrhage is a rare clinical disease. No cases of intraoperative ultrasound-assisted treatment of the disease have been reported in the literature. To present a case of idiopathic spontaneous intramedullary hemorrhage treated with intraoperative ultrasound and review the diagnosis and treatment of the disease.

Case presentation: An 11-year-old child was admitted to our department because of a sudden severe pain on the left side of her back. Magnetic resonance images of the thoracic vertebrae showed abnormal signals in T2-T3 and spontaneous intramedullary hemorrhage was suspected. Intraoperative ultrasound-assisted evacuation of the intramedullary hematoma was performed and no abnormal blood vessels or malformations were found during intraoperative exploration.

Conclusion: We report a rare case of idiopathic spontaneous intramedullary hemorrhage with intraoperative ultrasound-assisted hematoma clearance and a good prognosis.

Introduction

Unexplained spontaneous intramedullary hemorrhage is an extremely rare clinical disease characterized by a sudden onset of neurological impairment accompanied by acute, subacute, or chronic clinical processes. We report a case of idiopathic spontaneous intramedullary hemorrhage treated with intraoperative ultrasound, which has not been reported in the literature.

Case Report

An 11-year-old girl developed severe pain on the left side of her back with a constant burning sensation 20 hours before admission. She had a progressive movement disorder in both lower limbs within 2 hours, but maintained normal movement of both upper limbs and urine and stool retention. She was admitted to the Department of Pediatric Neurology at our hospital. After admission, physical examination showed paraplegia of both lower limbs, disappearance of the bilateral knee tendon reflex, bilateral Babinski's disease, hypothermia below the nipple plane, and normal bathyesthesia. The patient denied a history of trauma or a previous medical history and there was no recent history of drug use. Magnetic resonance imaging (MRI) of the cervical and thoracic vertebrae showed that the spinal cord was slightly thickened at the level of C5 to T6. The lesion showed an iso-signal shadow on T1-weighted imaging (T1WI; Fig. 1A) and a low signal shadow on T2WI (Fig. 1B). An enhanced scan did not show definite enhancement (Fig. 1C and D). Thus, hemorrhage and edema were considered. There was no obvious abnormality in the results of the laboratory examination. The patient was transferred to our department because the family refused the suggested operation and she was temporarily given symptomatic support treatment.

Eight days after admission, the patient's symptoms became aggravated and surgical treatment was planned after consultation with her family. Re-examination with MRI revealed a high signal intensity on T1WI and a low signal intensity on T2WI in the spinal cord at the T2 and T3 vertebral body level (Fig. 2A and B). Obvious edema was found in the lesion at the upper segment of the spinal cord and enhanced images (Fig. 2C and D) showed the lesion had diffuse high signal intensity. The clinical diagnosis was spinal cord hemorrhage at the

level of the T2 and T3 vertebrae, with spinal cord edema at the level of the C5 to T5 vertebrae. Because no obvious intramedullary vascular malformation or other abnormal lesions were observed in the MRI examination, myelography was not performed. T2-T3 laminectomy and intramedullary hematoma removal were performed under neurophysiological monitoring.

After laminotomy, ultrasound was used to locate the lesion and accurately cut the dura mater. The hemorrhagic segment of the spinal cord was dilated with high tension (Fig. 3A and B). Intraoperative ultrasound was used to accurately locate the lesion area on the surface of the spinal cord. Under neurophysiological monitoring, the spinal cord was cut longitudinally from the dorsal median sulcus of the spinal cord. Brown fluid with a very small amount of clotting was found in the intramedullary region and the tension on the spinal cord disappeared after clearance (Fig. 3C). The bleeding site was washed repeatedly with normal saline and no abnormal blood vessels and malformations were found.

The urine and stool functions recovered first on the second day after the operation though the muscle strength and sensory disturbance in both lower limbs were not significantly improved. Two weeks after the operation, the right lower limb recovered to grade IV/V on the muscle strength grading scale (the limb could withstand resistance, but the strength was weaker than normal), the left lower limb reached a grade of I/V (muscle contraction could be seen), and the sensory disturbance decreased at the T3-T4 level. Systematic rehabilitation treatment was given. During the follow-up at 4 months after the operation, the patient could walk freely.

Discussion

Spontaneous intramedullary hemorrhage is a rare disease and most cases have clear causes, such as oral anticoagulants, hematological diseases, etc. The incidence of intramedullary hemorrhage reported by Kreppel was 0.82% [12], including cases with a clear cause. There is no clear etiology for idiopathic spontaneous intramedullary hemorrhage and it is usually diagnosed following surgery when no potential pathology is found in macroscopic and microscopic examination [4]. Only a few cases have been reported in the literature [4–10].

Our review of idiopathic spontaneous intramedullary hemorrhage in the literature is summarized in Table 1. The reported incidence is the same for both genders (five male and five female patients). It has mostly been reported in middle aged individuals (age range from 11 to 77, with an average age of 45.5). The patients with idiopathic spontaneous intramedullary hemorrhage ranged from 45 to 65 years old, accounting for 60% of cases (six cases). Almost all of the lesions were located in the thoracic spinal cord, with the exception of one case involving the cervical spinal cord and one case involving the lumbar spinal cord. Idiopathic spontaneous intramedullary hemorrhage is usually characterized by acute onset and rapid deterioration of the neurological state of the patient, characterized by the Brown-Sequard syndrome [4–9]. A few patients showed chronic progression [10]. MRI examination of the spinal cord was performed for all patients [4–10] and was the first choice for intramedullary hemorrhage [11]. The MRI findings for intramedullary hematoma change over time. In the superacute phase (< 24 hours), MRI shows equal signal intensity on T1WI and high signal intensity on T2WI, which is sometimes difficult to distinguish from the cerebrospinal fluid. In the acute stage (24 h – 3 days), T1WI shows equal signal intensity and T2WI shows low signal intensity. In the early stage of the

subacute phase (3–7 days), T1WI shows high signal intensity and T2WI shows low signal intensity. The T1WI and T2WI signals remain hyperintense in the late subacute stage (7–30 days) and eventually change to low intensity signals in the chronic phase (>30 days) [11]. If no vascular lesions are found in spinal cord MRI, myelography is not recommended as a necessary examination, consistent with the recommendations by Karavelis [6] and other authors.

Table 1
Idiopathic spontaneous intramedullary hemorrhage reported in the literature

REPORTING YEAR	AUTHOR	GENDER	AGE	LOCATION	DURATION OF SYMPTOMS	OPERATION	OUTCOME
1996	Karavelis [6]	M	77	T9	1 month	YES	Good
2013	Chao [4]	F	59	T8-T10	10 days	YES	Good
2013	Chao [4]	F	51	T1-T5	3 days	YES	Good
2015	Choi [5]	F	48	T9-T10	7 days	YES	Good
2010	Sharma [9]	M	18	C6-T12		NO	Bad
2014	Lee [8]	M	34	T4-T5	18 hours	YES	Good
1999	Matusmura [10]	M	47	T2-T3	6 months	YES	Good
1999	Matusmura [10]	M	63	T3-T5	5 months	YES	Good
2005	Kumar [7]	F	47	T12-L1	1 day	YES	Bad
2019	Current case	F	11	T2-T3	8 days	YES	Good

Most authors recommend early diagnosis and surgical treatment for patients with intramedullary hemorrhage whose neurological function deteriorates rapidly [4–6, 8, 9]. The results shown in Table 1 demonstrate that the prognosis of the patients is not significantly correlated with sex, age, time from operation, and segment of hemorrhagic spinal cord affected. The prognosis is related to whether the patient receives surgical treatment or not. A patient with spontaneous intramedullary hemorrhage reported by Sharma [9] had a poor prognosis without operation and rehabilitation. The long-term prognosis is good for the vast majority of patients that receive surgical treatment. Among the 613 patients with spinal hematoma reported by Kreppel [12], the patients who received surgical treatment within 12 hours of onset had the highest recovery rate. The second highest recovery rate was for patients who received surgery more than 1 week after disease onset and the worst recovery rate was for patients treated with surgery within 1 week to 13 hours of onset. This may be related to the severity of the patient's clinical symptoms. The prognosis with conservative treatment is relatively good for a very small number of patients and this population of patients is characterized by mild paralysis [12].

It is worth noting that a patient with spontaneous intramedullary hemorrhage without obvious inducement reported by Hyun-Min [13] received systematic rehabilitation treatment immediately after diagnosis with good prognosis. Therefore, we suggest that patients with idiopathic spontaneous intramedullary hemorrhage should be considered for surgical treatment when their clinical symptoms are severe, such as a severe decrease in limb muscle strength or even paraplegia and loss of cystorectal function. Additionally, systematic rehabilitation should be performed after surgery. Conservative treatment can be considered if the muscle strength in the limb of the patient is partially decreased or the condition is limited. Patients managed with conservative treatment should undergo systematic rehabilitation at the early stage, as this is beneficial for improving spinal cord function [13].

All patients who undergo surgical treatment choose posterior laminectomy and intramedullary hematoma removal [4–8, 10]. Therefore, it is very important to accurately locate the focus and reduce iatrogenic injury of the spinal cord. The uniqueness of our case lies in the accurate identification of the location of intramedullary hemorrhage with the assistance of intraoperative ultrasound. The advantages of intraoperative ultrasound are: 1. Real-time guidance to identify the location and the resection range of lesions during surgery can reduce iatrogenic injury [14]. 2. The technique can be used repeatedly, the cost is low, and the economic burden on patients is reduced. 3. Intramedullary hematoma can mask the signal of abnormal lesions such as vascular malformations on MRI but such lesions can be detected in real time with the assistance of intraoperative ultrasound [15]. Using intraoperative ultrasound, surgeons can quickly determine the location of bleeding and reduce the length of the operation. With the help of neurophysiological monitoring, accurate and minimal injury incision of the spinal cord can lead to a good prognosis. In recent years, the use of intraoperative ultrasound was reported for the resection of intracranial and intraspinal tumors [16, 17] as well as the treatment of intracranial hemorrhage [18], showing good application prospects.

Conclusion

We report a rare case of idiopathic spontaneous intramedullary hemorrhage. The hematoma was removed with the assistance of ultrasound during the operation and the prognosis of the patient was good. And we believe that the application of intraoperative ultrasound is beneficial to the surgical treatment of intramedullary hemorrhage.

Declarations

Ethics approval and consent to participate

Not applicable

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editors-in-Chief of this journal.

Availability of data and materials

The datasets used and analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Author Contributions

LCY, WYB and ZXF diagnosed the patient. LCY and ZHQ analyzed literature data. All authors wrote and revised the manuscript.

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Figures

Figure 1

MRI shows a lesion located in the thoracic spinal cord at T2-T3. The lesion showed an iso-signal shadow on T1WI (A) and a low signal shadow on T2WI (B). No obvious enhancement was found on the contrast-enhanced image (C), and the axial image showed that the focus was located in the center (D) of the spinal cord.

Figure 2

Eight days later, MRI showed a lesion with high signal intensity in T1WI (A), low signal intensity in T2WI (B), and obvious edema in the upper segment of the spinal cord. Contrast-enhanced images (C, D) showed diffuse high signal intensity in the hematoma.

Figure 3

Intraoperative ultrasound showed that the hematoma was located in the spinal cord, showing low signal intensity and uniform density (A). When the dura mater was cut, the hemorrhagic segment of the spinal cord was dilated with high tension (B). After the hematoma was removed, the tension on the spinal cord disappeared. The bleeding site was explored, and no vascular abnormalities and malformations were found (C).