Cryptogenic Recurrent Spontaneous Epidural Hematoma: A Case Report and Literature Review

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Case report

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Abstract

Background

Spontaneous epidural hematoma (EDH) has been suggested associated with adjacent infective pathologies, dural vascular malformations, extradural metastasis, or coagulopathies. Cryptogenic spontaneous EDH is extremely rare.

Case presentation

We reported a cryptogenic spontaneous EDH case in a young woman following sexual intercourse. She occurred consecutively EDH at three different sites within a short time. After three timely operations, a satisfactory outcome was achieved.

Conclusion

EDH should be investigated when a young patient develops headaches and signs of increased ICP after emotional hyperactivity or hyperventilation. If early diagnosis and surgical decompression can be carried out in time, the prognosis is satisfactory.

Introduction

Intracranial epidural hematoma (EDH) is a hematoma that occurs between the inner plate of the skull and the dura mater, which accounts for about 30% of traumatic intracranial hematoma. EDH is mainly caused by skull fracture after head violence action, which induces hemorrhage by tearing meningeal arteries, veins, and venous sinuses. In contrast, spontaneous EDH is rare and mostly occurs due to adjacent infective pathologies, hematological system diseases, immune system diseases, vascular malformations, metastasis to the skull, etc.

Cryptogenic spontaneous EDH without specific underlying disease is extremely rarer in this category.(1, 2) The unidentified primary causes may lead to recurrent episodes of EDH, resulting in severe neurological dysfunction. In this study, we reported a case of cryptogenic recurrent spontaneous EDH (three times) in a 22-year-old woman who received three surgical procedures and finally achieved satisfactory outcomes. Literature focused on the underlying mechanism of spontaneous EDH was further reviewed.

Case Presentation

First EDH

A 22-year-old female was admitted with a blunt headache 1 hour after intercourse. On general physical examination, the consciousness was clear, and the speech was fluent. There was no obvious swelling or injury on the head or face. Neurologic specialist physical examination revealed that the Glasgow Coma Scale (GCS) score was 15 (eye opening, 4; motor responsiveness, 6; verbal performance 5). There were no

obvious positive signs in the nervous system except headache and dizziness. Emergent nonenhanced computed tomography (CT) scan (each layer with 5-mm-thick) revealed left temporoparietal EDH (the inner and outer diameter: 3cm, the maximum diameter: >10cm, the upper and lower diameter: 7.5 cm) with a rightward midline shift of 1cm. Bone window imaging confirmed no fracture line (**Figure 1A**). Preoperative coagulation tests showed that all coagulation parameters were within the normal range (**Table 1**). Past medical history suggested that the patient had undergone medical abortion 1 month ago, and the specific drug is unknown.

During preoperative preparation, the patient became increasingly unconscious and agitated. Before anesthesia, The GCS score was 11 (eye opening, 2; motor responsiveness, 5; verbal performance, 4). Bilateral pupils were equiround with a diameter of 3mm and had light reflection. Two hours after admission, the left temporo-occipital EDH was cleared under general anesthesia in the emergency operating room. No significant fracture lines or EDH responsible vessels were found during the operation. The intraoperative blood loss was about 300ml, and no blood transfusion was performed.

Second EDH

After the first operation, the patient's right pupil was dilated with a diameter of 5.0mm, and light reflection disappeared, while the left pupil diameter was 3.0mm, and light reflection was retarded. Immediate CT examination showed right temporo-occipital EDH (the inner and outer diameter: 3cm, the maximum diameter: >10cm, the upper and lower diameter: 8.5 cm) with a leftward midline shift of 1cm (**Figure 1B**). The right temporo-occipital EDH was removed immediately under general anesthesia. Decompression with flap removal was performed considering preoperative cerebral hernia and high intraoperative intracranial pressure (ICP). No significant fracture lines or EDH responsible vessels were found during the operation. The intraoperative blood loss was about 300ml. During the operation, 3U red blood cells and 625ml plasma were transfused.

Third EDH

After the second operation, the patient's right pupil was retracted with a diameter of 3.5mm, and light reflection was retarded. The third immediate CT examination showed bilateral frontal EDH (the inner and outer diameter of the right frontal part: 3.5cm, the hematoma volume: >30ml; the inner and outer diameter of the left frontal part: >2cm, the hematoma volume: 20ml) (**Figure 1C**). Bilateral frontal EDH removal was continued in the emergency operating room. As before, no fracture line or EDH responsible vessels were found during the operation. The intraoperative blood loss was about 350ml, and 200ml of plasma was transfused.

After anesthesia resuscitation, the patient resumed spontaneous breathing and regained consciousness. The GCS score was 14 (eye opening, 3; motor responsiveness, 6; verbal performance, 5). The postoperative CT scan showed satisfactory hematoma clearance, and the midline did not shift (**Figure 1D**). The patient was discharged two weeks after surgery with a mRS score of 0. There was no significant

abnormality in the CTA examination in the outpatient clinic 6 months after the operation (**Figure 2**), and the mRS score was 0.

Table 1. Coagulation and blood routine parameters during hospitalization

Parameters	Normal reference range	Preoperative	Postoperative day 1	Discharge
Coagulation parameters				
Thrombin time (TT), s	16-18	16.6	15.5	15.5
Prothrombin time (PT), s	9-13	12	12.2	11.4
Prothrombin time ratio	0.72-1.24	1.04	1.06	0.99
International Normalized Ratio (INR)	0.8-1.3	1.04	1.06	0.99
Partial prothrombin time, s	20-35	26.4	26.8	26.4
Activated partial thrombin time (APTT), s	23-27	26.4	26.8	26.4
Fibrinogen, g/L	2-4	2.39	2.73	4.37
Fibrinogen degradation products, mg/L	0-5	18.7	5.7	6.4
Antithrombin III activity, %	75-125	84.9	85.5	108.7
D-dimer, mg/L FEU	0-0.55	9.34	1.67	1.87
Blood routine parameters				
Erythrocyte, $10^{12}/L$	3.8-5.1	3.6	2.27	3.45
Hematocrit	0.35-0.45	0.346	0.216	0.325
Hemoglobin, g/L	115-150	114	71	104
Blood platelet, $10^9/L$	125-350	169	151	288
White blood cells, $10^9/L$	3.5-9.5	10.2	12.5	7.01
Thrombocytocrit	0.108-0.271	0.2	0.18	0.3

Discussion

EDH can be classified as traumatic or spontaneous, depending on the presence or absence of violence. Traumatic EDH is mostly caused by skull fracture or laceration of meningeal arteries, veins, and/or venous sinuses. Spontaneous EDH can be divided into primary spontaneous EDH and

secondary spontaneous EDH. The former is the EDH without relevant underlying diseases. The latter is usually caused by infectious disease, vascular malformation, blood system diseases, metastases, etc. The patient we reported had no history of trauma, and preoperative and intraoperative examinations ruled out possible primary underlying diseases mentioned above. Therefore, we diagnosed this patient as cryptogenic spontaneous EDH (primary).

Cryptogenic spontaneous EDH (primary)

Previous studies reported a total of 3 cryptogenic spontaneous EDH cases.(1-3) Chen et al. proposed that restlessness can induce elevated blood pressure and diffuse micro-bleeding of the dural vessels, which can stop spontaneously under normal circumstances.(1) When patients suffer from persistent irritability accompanied by acute hyperventilation, hypocapnia, and intracranial alkalosis will cause dramatic contraction of the arterial vessels, resulting in decreased cerebral blood flow. These physiological reactions eventually decrease ICP. The dissection of the dura and skull increases the accumulation of microbleeds and eventually forms the cryptogenic spontaneous EDH.(1, 4) In our case, we noticed that the patient's first EDH occurred one hour after intercourse, so we speculated that the underlying mechanism might be similar to the above hypothesis, that is, an intense physiological reaction cause a sharp drop in ICP and eventually leads to the dissection of the dura from the skull.

Unfortunately, the patient experienced another two consecutive EDH at different sites during the acute phase after the first operation. The underlying mechanism of the recurrent EDH may be the sharp drop in ICP due to removal of hematoma and dura dissection due to brain tissue displacement. In addition, the dura mater of young people does not adhere closely to the skull, and it may be easier to peel off than the elderly.

Spontaneous EDH with specific underlying disease (secondary)

Infectious diseases such as periodontitis, maxillary sinusitis infect the meningeal arteries and vessels between skull diploe by retrograde infection, leading to vascular inflammation changes. On the one hand, accumulation of inflammatory exudate, pus, and air in the epidural space may cause separation of the dura from the skull. On the other hand, the infiltrating effects of inflammation lead to thinning and increased permeability of the diploe vessel wall, resulting in a breakthrough hematoma in the epidural space.(5-7) Dural arteriovenous malformations have been reported as a possible cause of EDH.(8) Chen et al. reported a case of spontaneous EDH caused by Langerhan cell histiocytosis (LCH), which was mainly due to osteolytic changes in the skull.(9) Over 20 cases of spontaneous EDH caused by sickle cell disease (SCD) have been reported in previous studies. The main mechanism was thought to be the rapid proliferation and expansion of bone marrow tissue (hematopoietic), which destroys the normal anatomical structure of the skull.(10) Some brain metastasis (BMS) could invade the dura mater and destroy the adjacent bone, and even cause coagulation dysfunction, ultimately leading to the occurrence of EDH.(11-13) Complement activation and immune complex deposition in the vascular wall are also involved in the onset of spontaneous EDH.(14) These substances will increase the permeability of the blood-brain barrier (BBB) and then stimulate vascular endothelial cells through inflammatory cytokines or

autoantibodies. Causes immune injury, including cerebral cortex atrophy, cerebral hemorrhage, cerebral infarction, and intracranial arteriovenous multiple sclerosis.

Conclusion

Cryptogenic spontaneous EDH is rare. EDH should be investigated when a young patient develops headaches and signs of increased ICP after emotional hyperactivity or hyperventilation. If early diagnosis and surgical decompression can be carried out in time, the prognosis is satisfactory.

Declarations

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Ethics approval and consent to participate

All procedures described in this study were conducted according to the Declaration of Helsinki and approved by the Ethics Committee of the Third People's Hospital of Yancheng. All participants provided written informed consent before entry into the study.

Consent for publication

Written informed consent for publication of their clinical details and clinical images was obtained from the guardian of the patient. A copy of the consent form is available for review by the editor of this journal.

Availability of data and materials

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

MX,YX and XFC prepared the original draft. ZLC, YJW, XH and XJ analyzed the patients' information. HSW supervised the entire research process. All authors agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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Figures

Figure 1

Preoperative and postoperative CT images. A. Preoperative CT. B. CT scan after the first operation. C. CT scan after the second operation. D. CT scan after the third operation.

Figure 2

CTA at 6 months postoperatively