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Multiple Dural Arteriovenous Fistulas as the Mystery of Rapidly Progressive Dementia with Bilateral Thalamic Lesions

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Dural arteriovenous fistulas · Dementia · Embolization · Case report · Literature review

Abstract
Dementia is a gradual and irreversible loss of higher mental function, particularly memory. Dural arteriovenous fistulas (DAVFs) are one of the rare causes of a rapid decline in cognitive function, which can be curable. DAVFs are pathological shunts between the dural artery and the dural venous sinus, dural vein, or cortical vein. Here, we present a case that initially manifested nausea and dizziness and developed rapidly progressive dementia caused by DAVFs in the left transverse sinus-sigmoid sinus junction area and the sinus confluence area, combined with cerebral venous sinus thrombosis. Moreover, our case has multiple DAVFs that cause bilateral thalamic lesions and rapidly progressive dementia called thalamic dementia, which is infrequent and often misdiagnosed. His symptoms have improved after receiving endovascular embolization treatment. In addition to presenting our case, we conducted a systemic literature review to summarize how familiarity with the manifestation and early diagnosis of bilateral thalamic lesions caused by DAVFs can lead to earlier and more effective therapy.

Yu Zhan and Murad Al-Nusaif equally contributed to this work and shared the first authorship.

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Introduction

Dural arteriovenous fistulas (DAVFs) account for 10–15% of acquired cerebrovascular malformations [1] and derive from unknown causes. DAVFs are most common during the fourth and sixth decades but can occur at any age. Previous research claimed that venous system anomalies, head injuries, or cranial surgery could be potential etiologies [1]. The pathogenesis may be connected to neovascularization and the opening of arteriovenous traffic channels. Multiple shunts in DAVFs may correlate with dementia manifestations [2]; it may be that a higher percentage of cortical venous drainage increases the risk of bleeding and deep venous hypertension. DAVFs can present with pulsatile tinnitus, headache, stroke, cognitive impairment, and Parkinson's. Among them, DAVFs causing bilateral thalamic lesions leading to progressive dementia are rare. Cognitive loss can be reversed by endovascular embolization. Here, we present an intriguing case in which the patient first had nausea and vertigo before developing rapidly progressive dementia (RPD), which greatly improved after receiving endovascular embolization therapy. Furthermore, we conducted a thorough review of the relevant research on bilateral thalamic lesions induced by DAVFs to provide a synopsis of how familiarity with the symptoms of DAVFs may reduce the rate of DAVF misdiagnosis, resulting in earlier treatment and an improved prognosis.

Case Presentation

A 56-year-old male patient with a history of type 2 diabetes mellitus and chronic gastritis, two months before admission, had severe dizziness, nausea, and vomiting. Subsequently, he had a progressive inability to walk and stand. The neurological examination at the time of admission revealed fluent speech, spiritual decline, unresponsiveness, mildly increased muscle tone, muscle strength grade 5 in both upper limbs and grade 4 in both lower limbs, bilateral instability in the finger-nose test and heel-knee-shin test, and no other positive findings. Magnetic resonance imaging (MRI) of the brain revealed bilateral thalamic and right cerebellar lesions, with no apparent abnormalities in magnetic resonance angio (MRA) (Fig. 1a–h). Lumbar puncture cerebrospinal fluid pressure was 150 mmH_2O, with no abnormalities in routine, biochemistry, or pathology. At that time, he was diagnosed with "nonspecific inflammation". He was discharged after receiving anti-inflammatory (methylprednisolone, intravenous preparations, from 1 g/d, reduced the dose by half every 3 days) and fiber-lowering (fibrinolytic enzyme, intravenous preparations) medication for 21 days, during which his symptoms improved and the lesion shrank (Fig. 2a, b). He received methylprednisolone orally in a tapered-dosage manner after being discharged. A month later, he was readmitted to the hospital with sleepiness and dysphagia. Brain MRI after 50 days from baseline detected enlarged right cerebellum and bilateral thalamus lesions and new brainstem lesion (Fig. 2c–e) compared to the brain MRI after 12 days from baseline (Fig. 2a, b). In addition to transient diplopia, ataxia, and consciousness disturbance, serum IgM type sulfatide antibody and GM1 antibody were weakly positive; hence, Bickerstaff brainstem encephalitis (BBE) was considered. The patients' dysphagia and disturbance of consciousness considerably improved after treatment with low doses of anti-inflammatory hormones, anticoagulation, and dehydration therapy. However, 9 days later, the patient developed delirium and transitory psychotic symptoms, and 13 days later, RPD and significant consciousness impairment emerged.

The brain magnetic resonance venography + susceptibility-weighted imaging (SWI) + enhancement found bilateral thalamus SWI show significant patchy enhancement, multiple dotted stripes SWI with significant low signal, multiple venous sinuses filling defects, and many tortuous stripes in the cerebellum and bilateral basal ganglia area, as well as in the left

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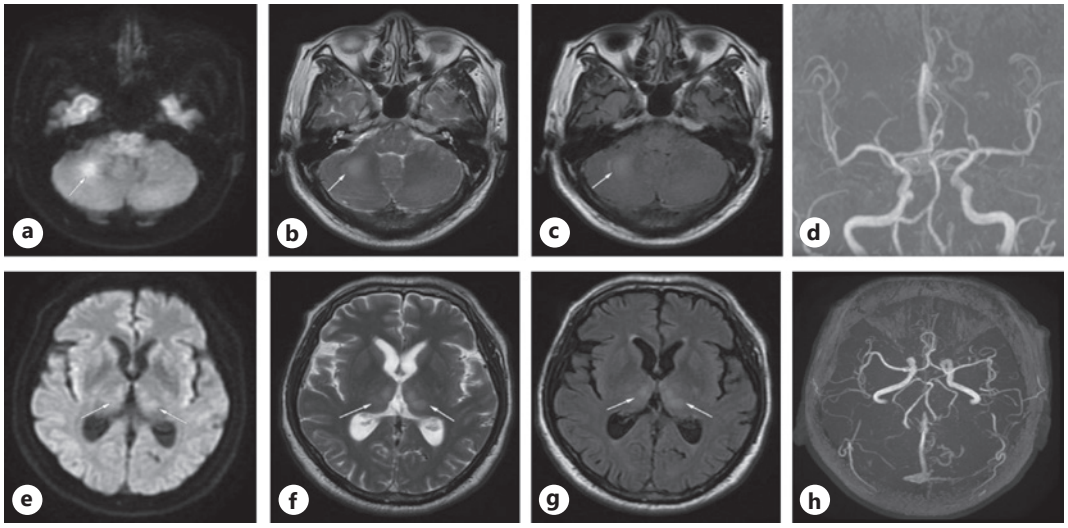

Fig. 1. Baseline MRI of the brain. **a**–**c** The right cerebellar pontine arm shows a slightly increased DWI signal and a slightly high signal on T2WI and T2 FLAIR; **e**–**g** Bilateral thalamus with multiple patchy abnormal signal shadow, slightly high signal on DWI, and slightly high signal on T2WI and T2 FLAIR; **d**, **h** No significant abnormalities in head MRA. DWI, difusion weighted imaging.

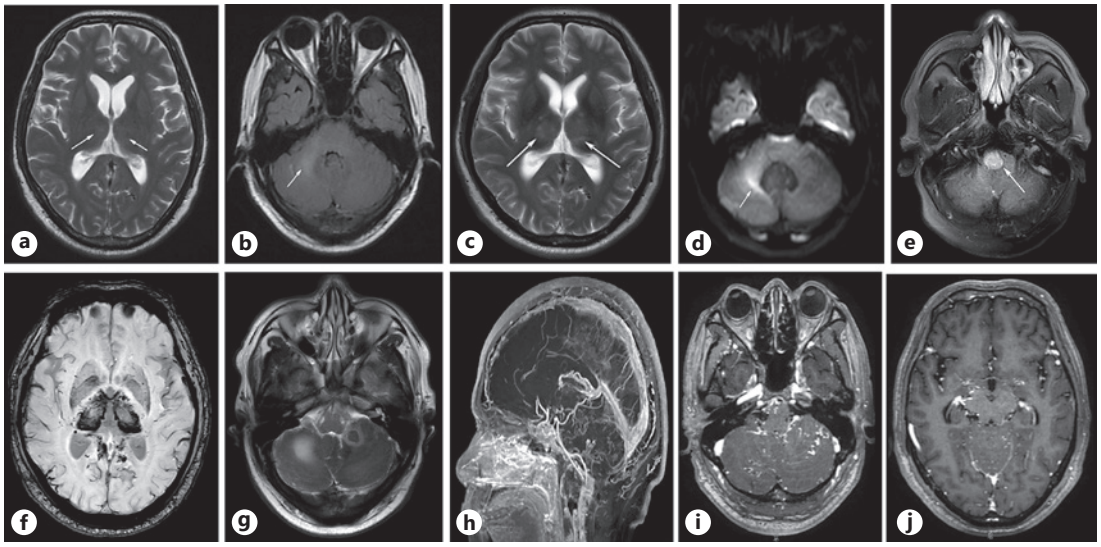

Fig. 2. Brain MRI after 12 days from baseline. **a**, **b** The left thalamus and right cerebellar bridge arm show T2WI and T2 FLAIR slightly high signal and no enhancement, and the scope of the lesion is significantly reduced compared with the previous; brain MRI after 50 days from baseline. **c**, **d** Bilateral thalamus and right cerebellar pontine arm are enlarged compared with the previous range; **e** T2 FLAIR new brainstem high signal intensity; brain MRI after 60 days from baseline. **f** Bilateral thalamus and medial temporal lobe SWI show significant patchy enhancement and multiple dotted strip SWI with the significant low signal; **g** the left pontine arm node shows T2WI low signal with surrounding ring of slightly high signal, and the right cerebellar pontine arm shows patchy T2WI slightly high signal; **h** left transverse sinus, ethmoid sinus, superior sagittal sinus filling defect; **i**, **j** many tortuous stripes in the cerebellum and bilateral basal ganglia area.

occipital lobe (Fig. 2f– j), which were the main imaging features of DAVFs. Digital subtraction angiography (DSA) revealed several DAVFs signs in the left transverse sinus-sigmoid sinus junction and sinus confluence regions. Blood supply arteries included bilateral branches of

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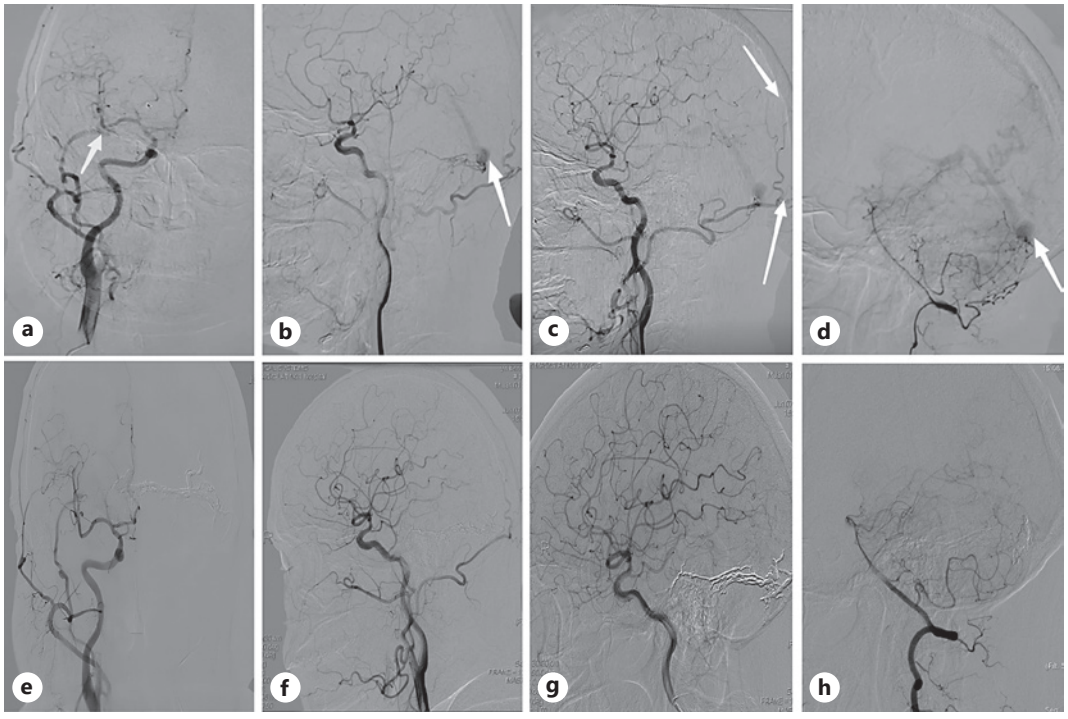

Fig. 3. Whole-brain baseline DSA **a**–**d** External carotid artery blood supply forms arteriovenous fistula at the confluence of transverse sinus and sigmoid sinus; through cortical venous return, superior sagittal sinus and transverse sinus are not visible; in left carotid arteriogram, left external carotid artery is the main blood supply artery; through occipital artery branches, superficial temporal branches blood supply, right vertebral artery small amount of blood supply, superior sagittal sinus venous return to the sinus confluence area are not visible. Whole-brain DSA after 6 months from baseline **e**–**h** complete fistulas embolization.

the middle meningeal artery, bilateral branches of the occipital artery, bilateral branches of the posterior meningeal artery, and bilateral branches of the posterior inferior meningeal artery of the cerebellum. Drainage veins were noticeably thickened, and reverse drainage occurred through dilated cortical veins and straight sinuses, with the superior sagittal sinus, left transverse sinus, and sigmoid sinus filling defects (Fig. 3a–d). Three days later, endovascular embolization with an Onyx (ethylene-vinyl alcohol copolymer) injection was carried out, and subsequent imaging revealed complete embolization of the DAVFs with intact venous drainage. Six days after surgery, the patient's consciousness and intellect improved slightly, but he was still unable to stand. In a rough test, the patient had 5-grade muscle strength in both extremities, slightly improved ataxic movement, moderate memory and orientation, and was less clear in speech 3 months after surgery. After 6 months of therapy, the patient had normal muscle strength; ataxia showed a significant improvement; the Mini-Mental State Examination score was 24/30; the Montreal Cognitive Assessment score was 21/30; and multiple DAVFs disappeared (Fig. 3e–h).

Discussion

When DAVFs are combined with venous sinus thrombosis, the invasive damage is severe. Manifestations of cerebral venous sinus thrombosis (CVST) vary depending on the location. The only sign of superior sagittal sinus thrombosis may be cranial hypertension, but diplopia,

dysphagia, and choking on water may be present in lateral sinus thrombosis affecting cranial nerves. In this case, baseline brain MRI has shown bilateral thalamic and right cerebellar pontine arm lesions, and venous phase vascular shadow was seen in brain MRA, which is considered to have formed CVST and DAVFs, resulting in intracranial deep venous congestion and venous hypertension, with reverse drainage to the thalamus via the dilated cortical venous and straight sinus. CVST pathogenesis may be related to coagulation disorders and inflammation [3]; short-term high-dose hormone shock therapy to reduce inflammation and relieve brain edema may explain the improvement of symptoms and shrinkage of the lesions. In our case, the sequential long-term oral hormone administration after discharge aggravated the CVST and DAVFs and led to the development of new lesions such as the brainstem, severe cranial hypertension, and bulbar palsy symptoms. Patients with superior sagittal sinus fistula and dementia may have a "vicious circle" of severe venous hypertension leading to multiple fistulas and more severe venous hypertension [2]. Progression of venous hypertension causes arterialized veins to rupture, resulting in severe deep venous congestion and cognitive impairment.

Bilateral thalamic lesions can be caused by cerebrovascular lesions, malignancies, infections, demyelination, autoimmune diseases, vitamin deficiencies, toxicities, genetic disorders, and metabolic diseases. Complex neurological symptoms in bilateral thalamic lesions vary according to the nuclei involved. One study in 2016 reviewed 19 cases and discovered that all patients had numerous cognitive domain impairments, ataxia, and aphasia [4]. We reviewed twelve cases of bilateral thalamic lesions induced by DAVFs published in PubMed after 2016 (Table 1). Of these patients, eleven were men, and one was a woman. They presented with cognitive and consciousness impairment, ataxia, Parkinson's, and dysphasia. These symptoms primarily seen in neurodegenerative diseases require extra vigilance for nonhemorrhagic neurological dysfunction due to DAVFs' involvement of deep brain structures such as the thalamus and brainstem when the symptoms develop acutely or subacutely rather than progressing chronically. All patients exhibited cognitive or consciousness disturbances, and eleven underwent endovascular therapy; nine had a good prognosis, two incompletely recovered, and one died. In our case, the fistulas were completely embolized after endovascular treatment. Despite the higher risk, surgical resection may be an option when the blood supply vessel is tiny or the venous sinus is narrowed. DAVFs trigger dementia due to edema of deep gray and white matter structures. Cortical dementia may be secondary to impaired cerebrospinal fluid absorption due to occlusion of normal draining sinuses or deep venous congestion with venous hypertension [2]. The potential anatomical pathways of thalamic dementia may be the papillary thalamic tract, the dorsal lateral nucleus, and the intraplate nucleus are disrupted in DAVFs [5], mainly caused by bilateral thalamic venous hypertension. Damage to the papillary thalamic tract can result in limbic system neural circuit failure, characterized by rapidly progressive cognitive dysfunction, including disorientation, executive dysfunction, attention impairments, memory problems, and disinhibition. Our patient had eight blood supply arteries in DAVFs drain retrograde to the thalamus via the straight sinus and cortical veins typed as Borden type III or Cognard type IV [1]. Brief psychiatric symptoms and preoperative signs of fast global cognitive impairment, ataxia, and aphasia, the symptoms of our case are consistent with earlier findings [6], and fluctuating cognitive abnormalities may be related to changing venous hypertension [7].

Various nuclei govern the complex functions of the thalamus. The thalamus's ventral lateral (VL) nucleus is the principal transmitter of information from the cerebellum to the motor cortex [8]. Thus, symptoms of ataxia and mild motor weakness appear due to disruption of the VL's ventral section after thalamic strokes. Language impairments may result from the left paramedian and tuberothalamic lesions that include the ventrolateral nucleus [8]. The involvement of the VL nucleus, left paramedian, and left tuberothalamic lesions in our patient can be responsible for bilateral ataxia and speech impediment, respectively. High signals

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Table 1. Summary of cases of DAVF with bilateral thalamic lesions

Author	Gender/age	Position	Clinical symptoms	MRI characteristics (on T2 FLAIR weighting)	Supply artery, fistula draining vein	Treatment	Prognosis
Hwang et al., 2017 [6]	Male/43	TSS	Inattention, transient episodic memory loss, hypersomnolencet	Bilateral thalamic signal change	RMMA, ROA, LPMA	Multiple stages of TAE	GR
Colorado et al., 2018 [9]	Female/62	Tentorium (SS)	Subacute cognitive decline, ataxia, parkinsonism, acute deterioration following LP	Bilateral thalami, right dorsal midbrain	PMA, LOA, with drainage into variceal superior hemispheric veins and the vein of Galen	TAE with Onyx	IR
	Male/64	Bilateral petrous apices	Acute confusion, agitation, memory impairment	Bilateral thalami, mammillary bodies, tectum	RMHT, ROA	TAE with Onyx (left), surgical clipping of the fistula (right)	GR
	Male/51	Tentorium (SS)	Subacute cognitive decline, memory impairment, disorientation, parkinsonism, hypersomnolence	Bilateral thalami, midbrain, pons	STA, MMA, OA	Onyx embolization in stage one, Onyx and NBCA embolization in stage two	GR
	Male/42	Tentorium Cerebelli	Subacute cognitive decline, memory impairment, disorientation, generalized seizure	Bilateral midbrain, pons, medulla, and upper cervical cord	BSCA with drainage into a dilated basal vein of Rosenthal and retrograde into lateral pontomesencephalic veins	Onyx embolization, two stages	IR
Cheng et al., 2018 [7]	Male/46	UK	Fluctuation of cognitive dysfunction	Symmetrical bithalamic hyperdensity	a branch of ICA with a tortuous the vein of Galen	Endovascular treatment	GR
Chen et al. 2020 [10]	Male/68	SS	Parkinson's symptoms and memory disorders	Bilateral thalamic high signal intensity	RICA	Open surgery	GR

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Table 1 (continued)

Author	Gender/age	Position	Clinical symptoms	MRI characteristics (on T2 FLAIR weighting)	Supply artery, fistula draining vein	Treatment	Prognosis
Iampreechakul et al., 2020 [11]	Male/52	SS?SSS	Progressive dementia, behavior change	Bilateral thalamic edema	LAPA, LSCA, multiple tiny branches of LOA with retrograde venous drainage into the straight sinus and vein of Galen	Two stages of TAE	GR
Hall et al., 2020 [12]	Male/79	SS	Confusion, slurred speech, difficulty swallowing, headache, mobility decline	Thalamic, midbrain, tectal plate	BOA, LMMA	TAE with Onyx	GR
Zhang et al., 2021 [13]	Male/53	Tentorium Cerebelli	Memory loss and abnormal behavior	Thalamic	BMHT, BMMA RPMA, RPCA	TAE	GR
Rizzo et al., 2021 [14]	Male/61	TH	Excessive daytime sleepiness, confusion, mental slowing, memory loss, behavioral changes	Bilateral thalamic, partial midbrain, periaqueductal gray	branches of ROA, VA	TAPE	Death
Liu, et al., 2022 [15]	Male/52	Tentorium Cerebelli	Slow response, inarticulation, slow speech, poor short-term memory	Bilateral thalamic mottling signal	LMMA, draining into the basilar vein	TAE with Onyx	GR

Abbreviations: SS, straight sinus; SSS, superior sagittal sinus; TSS, transverse, sigmoid sinus; TH, torcular herophili; BMHT, bilateral meningohypophyseal trunks; RMHT, right meningohypophyseal trunks; MMA, middle meningeal artery; BMMA, bilateral middle meningeal arteries; LMMA, left middle meningeal artery; RMMA, right middle meningeal artery; OA, occipital artery; BOA, bilateral occipital arteries; LOA, left occipital artery; ROA, right occipital artery; BSCA, bilateral superior cerebellar artery; LSCA, left superior cerebellar artery(tentorial branch); RPCA, right posterior cerebral artery; ICA, internal carotid artery; RICA, right internal carotid artery; PMA, posterior meningeal artery; LPMA, left posterior meningeal artery; RPMA, right posterior meningeal artery; LAPA, left ascending pharyngeal artery; STA, superficial temporal artery; VA, vertebral artery; LP, lumbar puncture; TAE, transarterial embolization; TAPE, transarterial partial embolization; NBCA, N-butyl-2-cyanoacrylate; GR, good recovery; IR, incomplete recovery; UK, unknown.

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were observed on T2-weighted imaging (T2WI) and T2-weighted fuid-attenuated inversion recovery (T2 FLAIR), predominantly bilateral thalamus, cerebellum, and brainstem. The occurrence of several lesions on imaging in this instance led to a false-positive diagnosis of BBE since the BBE patient's brain MRI revealed T2WI high signals in the upper midbrain, cerebellum, thalamus, or brainstem that was comparable to DAVFs. Noninvasive angiography can reveal vascular anomalies, but DSA is still the gold standard for determining cerebral vascularity, the location of fistulas, blood supply arteries, and drainage patterns, which are necessary for treatment plan design.

Conclusion

Dementia is frequently misdiagnosed in patients with DAVFs. Consequently, DAVFs should be considered in progressive dementia associated with bilateral thalamic lesions. A DSA is recommended for an accurate diagnosis, and early selective endovascular treatment could improve the outcome.

Statement of Ethics

The research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. This research proposal has been reviewed and approved by the Ethics Committee of the First Affiliated Hospital of Dalian Medical University, approval number PJ-KS-KY-2022-233. Written informed consent was obtained from participants for publication of the medical case details and any accompanying images.

Conflicts of Interest Statement

The authors declare that they have no conflict of interest.

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Author Contributions

Yu Zhan contributed to conceptualization, work drafting, literature review, and editing; approved the final version; and agreed to accept responsibility for all aspects of the work. Murad Al-Nusaif contributed to conceptualization, work drafting, and English editing of the manuscript; approved the final version; and agreed to be accountable for all aspects of the work. Chang Xu contributed to the acquisition of the patient's clinical data and revising; approved the final version; and agreed to be accountable for all aspects of the work. Jiahao Li contributed to the acquisition of the imaging data; reviewed the manuscript; approved the final version to be published; and agreed to be accountable for all aspects of the work. Li Zhao contributed to conceptualization; reviewed and edited the article; approved the final version; and agreed to be accountable for all aspects of the work. Feng Wang contributed to acquisition

and supervision of the interventional management and editing; approved the final version; and agreed to be accountable for all aspects of the work. Chunbo Dong designed the manuscript; supervised clinical management and editing; approved the version; and agreed to be accountable for all aspects of the work.

Data Availability Statement

All data that support the findings of this study are included in this publication.

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