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Atypical Postpartum Stroke Presenting as Opalski Syndrome: Case Report and Review of the Literature

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Key Words
Post-delivery stroke · Opalski syndrome · Lateral medullary syndrome · Central sinus vein thrombosis · Pregnancy · Arterial dissection

Abstract
Background and Purpose: We present the first case of combined arterial (vertebral artery dissection) and venous [central sinus vein thrombosis (CSVT)] diseases presenting as Opalski syndrome in a female patient following induced delivery.
Case Description: A 32-year-old woman was admitted to our institute two weeks after induced delivery with intriguing neurological findings that were finally diagnosed as a combined venous-arterial disease. Although she was referred diagnosed with CSVT, her neurological findings indicated Wallenberg 'plus' syndrome with ipsilateral hemiparesis (Opalski syndrome), further confirmed by neuroimaging revealing arterial disease (vertebral artery dissection) combined with incidental acute CSVT. Coagulation, gynecological and cardiac problems were ruled out. Treatment consisted of continuous heparin with rigorous control of her blood pressure. Nine days later, the patient was discharged with prominent improvements. Most of the symptoms resolved following 3 months of rehabilitation.
Conclusions: Atypical strokes (such as Opalski syndrome) might present in postpartum patients. This rare diagnosis should be suspected in patients with Wallenberg 'plus' syndrome, and neuroimaging studies for determining the presence of arterial disease and brain stem lesions should be performed. Concomitant CSVT is rare and might mislead. Fine diagnosis followed by immediate conservative treatment can be of great benefit.

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Introduction

Pregnant women might be in risk for central sinus vein thrombosis (CSVT) and vertebral artery dissection (VAD). Opalski syndrome, a rare variant of lateral medullary syndrome (LMS) with concomitant ipsilateral hemiparesis, results from the ipsilateral extension of the medullary infarct to the superior cervical cord caudal to the pyramidal decussation. Its etiology might be iatrogenic following induced delivery and coughing episodes [1]. A concomitant venous-arterial disease (Opalski syndrome and incidental CSVT) following induced delivery has not been reported previously. In our case, intriguing neurological findings followed by neuroimaging confirmed the diagnosis, leading to successful conservative treatment.

Case Report

A 32-year-old healthy woman with 2 previous normal deliveries and a recent induced vaginal delivery was referred from another hospital diagnosed with CSVT. Family and personal history was noncontributory, except for migraine, no smoking history and psychotherapy for anxiety. The latest pregnancy was normal up to the 28th week during which she complained of dyspnea and coughing (on supine position) that spontaneously resolved. No further complaints appeared until the 30th week when she suspected reduced fetal movement which was diagnosed as a nonviable fetus, leading to induced vaginal delivery on the following day. She was discharged 2 days later with no other complaints and no additional treatment.

One week later, headaches and neck tenderness (right side) appeared, for which conservative management was recommended. Four days later, she experienced dizziness, finger numbness and intermittent suffocation. Her unremarkable neurological examination in the emergency room (ER) led the physician to discharge her for ambulatory follow-up, diagnosed as 'mixed anxiety and depressive' reaction. Two days later, now complaining of blurred vision, vomiting and urinary retention, the patient was referred to the ER again and was admitted to the gynecology ward.

Her gynecological and ophthalmological evaluations were normal; 500 cm^3 of urine was found in a post-voiding ultrasound, and an unexplained hypertension was treated. One day after admission, a new right hemiparesis (arm > leg) appeared. A CT venography (CTV) demonstrated CSVT of the left transverse and sigmoid sinuses (**fig. 1**a), with no signs of edema or hemorrhage. Heparin was initiated, and the patient was referred to our ER for 'evaluating the option of an endovascular treatment'.

We found discrepancies between the neurological signs and the isolated diagnosis of CSVT, and suspected brain stem involvement. A reevaluation of the CTV (that unintentionally included an arterial phase) revealed right VAD (V3–4 segments; fig. 1b), further confirmed by MRI showing an ischemic infarct damage of the brain stem (lateral and anterior medullary infarcts) and cerebellum, explaining the patient's symptoms (**fig. 2**a–d). Concomitant incidental CSVT was confirmed. Aneurysms or stenosis were excluded.

On physical examination, tachycardia (heart rate 93) and hypertension (166/113 mm Hg) were noted. The patient was alert, suffering from headache and presenting the following neurological findings suggestive of Opalski syndrome: ipsilateral hemiparesis; vestibular signs (vertigo, diplopia, nystagmus and vomiting); cerebellar signs (side ataxia, dysdiadochokinesia, nystagmus and dysarthria); lateral spinothalamic tract signs (contralateral deficits in pain and temperature sensation, with preserved proprioception); nucleus ambiguus signs (dysphagia, hoarseness, palate and tongue deviation to the right); spinal trigeminal nucleus signs (ipsilateral loss of pain and temperature sensation from V2–3 distribution, with normal V1 and reduced sensation of the palate); bilateral positive Babinski signs; descending sympathetic fiber signs (partial ipsilateral Horner's syndrome: myosis and ptosis), and diplopia due to limited upward and lateral gaze of the right eye.

Heparin treatment maintained a PTT range of 60–70 s, followed by unfractionated heparin treatment (80 mg twice a day) with rigorous blood pressure (BP) control (systolic 110–140 mm Hg). Gradual improvement could be noted, and the patient started to walk by day 4 and swallow by day 6.

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A thorough workup included an echocardiography (mild tricuspid regurgitation), bilateral lower limb Doppler examination, hematological workup (CRP, folic acid, vitamin B12, homocysteine, factor V Leiden, cytoplasmic and perinuclear ANCA (antineutrophil cytoplasmic antibodies), rheumatoid factor, prothrombin, complement factors, anti-thrombin 3, β-2 glycoprotein, cardiolipin IgM and IgG, proteins S and C, and lupus anticoagulant). All results were normal.

The patient was discharged after 9 days, further improving, being fully conscious and less hoarse, eating by herself, having less diplopia, nystagmus and hypoesthesia, and walking independently. After 3 months of rehabilitation, she resumed a normal lifestyle with further improvement on a recent MRI.

Discussion

Opalski Syndrome

Opalski [2] first described this phenomenon in 1946 as a variant of LMS with concomitant ipsilateral hemiparesis explained by the ipsilateral extension of the medullary infarct to the superior cervical cord caudal to the pyramidal decussation. Others have explained these combined findings as a result of vertebral artery (VA) occlusion/stenosis or VAD [3, 4], compromising the medullary penetrating arteries that arise from distal VAs or anterior spinal arteries [5]. Opalski syndrome might be due to either ischemic disturbances (such as distal VAD) and/or due to hemodynamically vulnerable areas such as the 'borderline zone' between the anterior and posterior spinal arteries and between the vertebral and spinal arteries [4].

Concomitant LMS with ipsilateral hemiparesis, supported by neuroimaging, led us to this rare diagnosis. Either the induced delivery and/or the unexplained coughs could have caused the VAD leading to brain stem lesions.

CSVT, Stroke and Arterial Dissection during Puerperium

Our patient presented a rare combination of VAD causing Opalski syndrome with incidental CSVT. Cases of dissections with or without concomitant CSVT following deliveries have been reported [6–12, 13–17]; however, our case differs from those found in the literature in that: (1) the gynecological course was more complicated [intrauterine fetal death (IUFD) and induced delivery]; (2) our patient experienced unexplained dyspnea and coughing 2 weeks before IUFD diagnosis; (3) our patient suffered from concomitant VAD, Opalski syndrome and CSVT, and (4) there were suspected findings of bilateral damages in the brain stem-spinal cord junction.

Women of child-bearing age have a higher incidence of stroke compared to age-matched men [6, 10–12], with only mild differences between pregnant and nonpregnant patients [18]. Nonpregnant women have a lower incidence of hemorrhagic than ischemic strokes; however, these frequencies become similar during pregnancy, suggesting pregnancy as a risk factor for hemorrhagic strokes [11, 12]. Strokes usually occur during the third trimester and the puerperium [18], up to 1 month postpartum [19]. Eclampsia/preeclampsia remains the main cause for both types of strokes (up to 47%). Hemorrhagic strokes carry a poor prognosis [20, 21].

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During the peripartum period, the overall risk for strokes (13.1–10.3/100,000) and CSVT (11.6/100,000) is similar. Among nonpregnant aged-matched females, CSVT occur less frequently [6, 18], suggesting that in the peripartum period, women are prone to develop CSVT (approx. 5–20% of all CSVT occur during this period [11]), mainly during the first 14 days postpartum [13]. Notably, CSVT by itself is considered as a cause for stroke in up to 38% of cases during the postpartum period [6, 9, 14–16].

Another cause for ischemic strokes in young females is arterial dissection during pregnancy [18]. These might be associated with connective tissue disorders, trauma and aneurysms. Extracranial dissections commonly present as neck pain and headache (occipital and posterior cervical regions), and might be proceeded by ischemic symptoms within the first weeks of the dissection [22]. This gradual evolution can delay diagnosis.

Unlike carotid dissections, VAD more frequently extend intracranially and involve the subarachnoid space causing meningism. Less than 30% of the cases have simultaneous dissections of two or more vessels, for which an underlying connective tissue disorder should be suspected [17, 18, 31].

Cervicocephalic arterial dissection (CCAD) is a rare postpartum complication, with only 16 reported cases to date [7–11, 13–17]. The rate of spontaneous CCAD in the general population is 2.6/100,000, of which 2.4% can occur during the postpartum period, meaning that there are ~6.2/10,000,000 annually [9, 23, 24], representing ~6% of spontaneous CCAD in women under 50 years of age. Extracranial dissections seem to occur more frequently among pregnant women [12]. Up to 90% of VAD might be complicated by embolic stroke [1, 29].

Reversible cerebral vasoconstriction syndrome was associated with stroke during the postpartum period among 4/6 patients compared with only 3/96 aged-matched controls. Contrary to reversible cerebral vasoconstriction syndrome which has a favorable outcome, intracranial dissections in postpartum patients are characterized by a worse outcome [7, 30].

The different occurrence between intra- and extradural dissections could possibly be explained by histological changes that appear once arteries (VAs and internal carotid arteries) become 'intradural': 'The change is characterized first by a diminution in thickness of the adventitial and medial coats, and second, by a very gross reduction or loss of elastic fibers in the media and external elastic lamina' [25].

There are two issues regarding the therapeutic course. The first issue concerns BP control. Our patient needed a tradeoff between BP reduction (due to the possible hemorrhagic transformation in the stroke area) and the general permissive hypertension (for dissections). The second issue concerns the usage of antiplatelet/anticoagulation drugs, to which caution was given due to the possible hemorrhagic transformation of a stroke. Once anticoagulation treatment is initiated, it will be generally replaced by antiplatelet therapy for approximately 6 months, due to the fact that most CCAD resolve after 3–6 months [26]. Neither therapy (antiplatelet vs. anticoagulant) was found superior for treating acute dissection [26, 27]. Once the possibility of a connective tissue disorder is excluded, the risk of recurrence is 1% within 1.4–8.6 years and up to 2% within the first months following the dissection [28, 32].

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Induced Delivery Complications and Concerns about Coagulopathies

Our patient presented vascular pathologies of two different origin: one of venous origin (CSVT) and one of arterial origin (VAD). Both could have been complications of the peripartum period and of the induced delivery. Hypercoagulability should also be considered since it can either cause or be a consequence of IUFD. Since no postmortem examination of the fetus was done, these issues as well as the reason for the IUFD remained unsolved.

Despite two previous normal pregnancies/deliveries and noncontributory personal/familial coagulopathies, transient hemodynamic as well as hematologic changes during pregnancy and puerperium might be considered as additional causes. Pregnancy might alter all three factors of Virchow's triad (blood flow, blood vessel wall and constitution of blood) [33], and patients are therefore prone to develop vascular diseases, as seen in our patient.

Anatomical Considerations (Posterior Circulation)

Opalski syndrome is caused by compromised blood supply and/or hemodynamic vulnerability, both likely occurring during puerperium and/or induced delivery. It is well accepted that nearly all posterior inferior cerebellar artery (PICA) occlusions and more than 50% of VA occlusions (mainly if a PICA origin is included) might manifest as LMS. Importantly, 75% of all LMS cases are related to VA occlusion and 12% to pure PICA occlusions [22].

The last peridural VA segment is V3. The intradural V4 segment ascends to the lower pontine border and forms the basilar artery, giving rise to the PICA en route (10 mm distal to the dura perforation). Our patient's right V3–4 dissection included the PICA, presenting as right medullary and cerebellar infarcts. This dissection might explain both constituents of Opalski syndrome: (1) LMS signs might be caused by the PICA-VA-related brain stem strokes, while (2) concomitant ipsilateral hemiparesis might be caused by the involvement of the anterior spinal artery arising from V4 that normally supplies the ventral two thirds of the spinal cord, including the pyramidal tract.

Conclusion

Combined venous-arterial disease should be considered among patients at high-risk for both pathologies, such as women during puerperium, even when their medical history is unremarkable. Discrepancies between neurological findings and incidental neuroimaging findings can mislead.

A clear-cut mechanism for the combined pathology could not be found; however, the combination of induced delivery combined with hypercoagulability and/or hemodynamic vulnerability during puerperium seems a reasonable option. It seems plausible that either the coughs and/or the induced delivery have caused the VAD that, in turn, led to Opalski syndrome. Pregnancy-related hypercoagulability might have been the reason for the concomitant CSVT.

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Conservative treatment combining BP control and anticoagulation seems an appropriate first step for these kinds of combined pathologies.

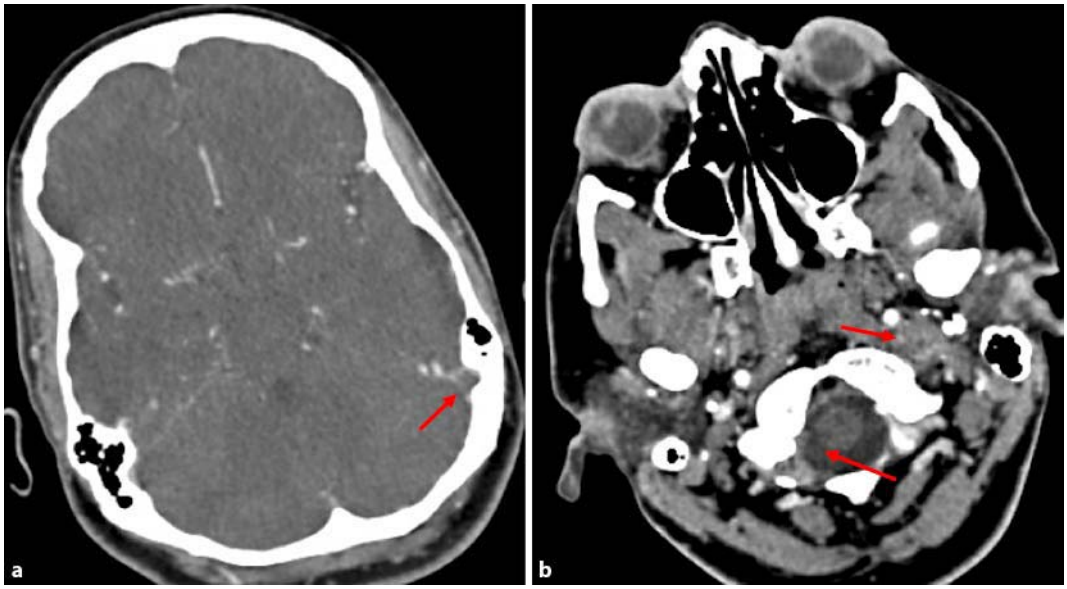

Fig. 1. a CTV showing left vein thrombosis. **b** CTV showing right VAD (V4 segment) and left jugular thrombosis.

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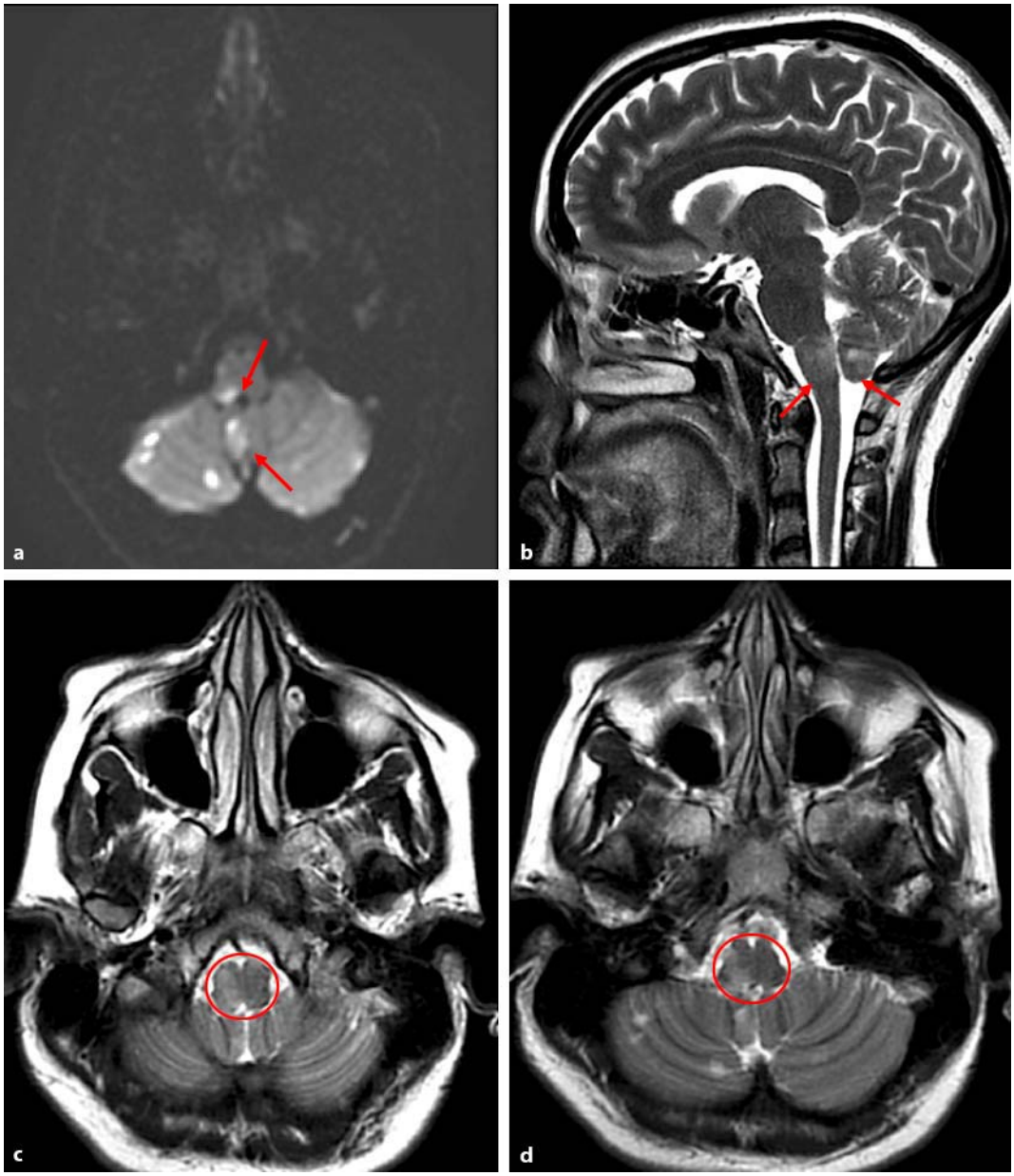

Fig. 2. **a** MRI (diffusion-weighted image) showing infarcts at the pyramidal decussating level. **b** MRI (T_2-weighted image) showing acute medullary and cerebellar infarcts. **c** MRI (T_2-weighted image) showing suspected bilateral infarcts at the pyramidal decussating level. **d** MRI (T_2-weighted image) showing acute medullary and cerebellar infarcts and CSVT.

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