CASE REPORT

OLGU SUNUMU

POSTPARTUM REVERSIBLE CEREBRAL VASOCONSTRICTION SYNDROME PRESENTING WITH
REVERSIBLE SPLENIAL LESION

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ABSTRACT

Co-occurrence of postpartum reversible cerebral vasoconstriction syndrome (RCVS) and reversible splenial lesion syndrome (RESLES) is highly rare, and may provide an opportunity in understanding the mechanism of these two rare entities. A 28-year-old female with postpartum psychosis in association with both RCVS and RESLES was reported. The worsening with antipsychotics and the dramatic response to oral nimodipine served as a clue to the diagnosis. RESLES and RCVS may have overlapping pathophysiology. It is possible that RESLES is one of the forms of cerebral edema resulting from RCVS.

Keywords: Reversible cerebral vasoconstriction syndrome, reversible splenial lesion, postpartum psychosis.
GEÇİCİ SPLENİAL LEZYON İLE PREZENTE POSTPARTUM REVERSİBİL SEREBRAL VAZOKONSTRİKSİYON SENDROMU

ÖZ
Postpartum reversibıl serebral vazokonstrıksión sendromu (RSVS) ile geçici splenial lezyon birlikteliği oldukça nadır bir durum olup, bu iki nadır hastalığın altında yatan mekanizmaların anlaşılabilmesi için bir fırsat doğmaktadır. Bu çalışmada, 28 yaşında RSVS ve geçici splenial lezyon birlikteliği ile ilişkili bir postpartum psikoz olgusu tartışılmıştır. Hastın antipsikotikler ile kötüleşmiş olması ve oral nimodipin'e dramatik yanıt vermesi hatta tam için ipucu teşkil etmiştir. Geçici splenial lezyon ile RSVS'nin orta k patofizyolojik özellikleri olduğu düşünülebilir. Geçici splenial lezyonun RSVS sebebiyle gelişen serebral ödem tiplerinden birisi olması mümkündür.

Anahtar Sözcükler: Reversibıl serebral vazokonstrıksión sendromu, geçici splenial lezyon, postpartum psikoz.

INTRODUCTION
Reversible cerebral vasoconstriction syndrome (RCVS) comprises a group of disorders characterized by fully reversible narrowing of the cerebral arteries, usually presenting with acute-onset, severe and recurrent headaches, with or without additional neurologic signs and symptoms (1). It has been associated with several diverse conditions including pregnancy, vasoconstrictive drugs, and primary headache syndromes (2).

Postpartum reversible cerebral vasoconstriction syndrome (PPRCVS) is a distinct clinical entity comprising up to 10% of all RCVS cases (3-5). Even though the underlying mechanisms contributing to PPRCVS are not uncovered yet, alterations in female reproductive hormones and pregnancy-related changes in vascular physiology are thought to play a role in its pathophysiology (6).

Reversible splenial lesion syndrome (RESLES) is a rare benign clinic-radiological syndrome characterized by reversible, focal, and usually diffusion-restricted lesion in the central region of the splenium of corpus callosum (sCC) (7). It is associated with a plethora of diverse etiologies, yet the pathophysiologic mechanisms are not completely understood (8). These two syndromes can rarely coexist. (9-11) We revisit the physiopathology of this association based on a new case reported herein and three other cases published in the germane literature.

CASE REPORT
A 28-year-old woman was consulted for acute onset disorientation and behavioral abnormalities that started two days after an uneventful delivery by cesarean section. Her relatives and physicians declared absence of significant medical issues and headache prior to her mental deterioration. We were informed that intramuscular methylergonovine was administered after the delivery to prevent postpartum bleeding. On examination, she was non-cooperative, disoriented and inattentive. Her speech was slow with perseverations. No other abnormality was detected in the neurological examination.

Since brain computed tomography showed no abnormal findings, a brain magnetic resonance imaging (MRI) study was scheduled. However, MRI was postponed due to her agitation, and a psychiatry consultation was obtained for alternating agitation episodes and an ongoing increase in her negativistic behaviors. Antipsychotic treatment consisting of olanzapine 2.5 mg/day was initiated with a presumptive diagnosis of postpartum psychosis, which was followed by the further deterioration of the patient into a state of akinetic mutism. Olanzapine was then altered with risperidone 1 mg/day and lorazepam 3x1 mg/day was added but no clinical change was obtained. The MRI was then performed under sedation, and showed a nodular lesion in the center of the sCC characterized by restricted diffusion indicative of RESLES (Figure 1). The MR angiography (MRA) revealed segmental narrowing and dilatation of both anterior and posterior cerebral arteries, suggestive of postpartum angiopathy (also known as RCVS) (Figure 2). Oral nimodipine (6x30 mg/day) treatment was initiated, and clinical amelioration has observed on the second day of the treatment. The patient’s mental status returned to normal and her speech became fluent without any pathological signs in several days.

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Anti-psychotic treatments were discontinued, and the patient was discharged on the fourth day of nimodipine treatment with a normalized neurologic examination. After three weeks of nimodipine treatment, a follow-up MRI revealed resolution of the splenial lesion and the cerebral arteriopathy significantly. The treatment was subsequently stopped and she had no further health problems since then. Of note, consent was obtained for this paper from the patient.

DISCUSSION AND CONCLUSION

Postpartum cerebral angiopathy is a rare disease presenting with headache, seizures and focal neurological deficits, following labor. It occurs generally within the first 3 weeks of the postpartum period, but late onset presentations until 6 weeks postpartum have been reported (12). The most common symptom is thunderclap headache. Focal neurological deficits such as visual disturbances, dysarthria, aphasia, extremity paresis, dysesthesia, ataxia and epileptic seizures can also be present (2).

PPRCVS, as one of the severe forms of RCVS spectrum disorders, is characterized by a 75% incidence of neurological deficits or seizure, which is higher than other RCVS subtypes (3-6). Pregnancy is a known risk factor but the pathophysiological mechanisms are not yet fully elucidated. Pregnancy associated physiological changes, such as increased blood volume, systemic vasodilation, increased peripheral vascular resistance, increased arterial tonus, mild systemic inflammation and changes in the lipid profile are considered to alter the arterial wall functions, resulting in the observed diffuse cerebral arteriopathy (13). In addition, the use of vasoactive drugs such as methylergonovine during the peripartum period, which was the case in our patient, might also contribute to the pathophysiology (14).

Our detailed literature search revealed a total of 10 postpartum RESLES cases, 3 of which were associated with postpartum RCVS, making our case the fourth (Table) (9-11,15-20). The suggested etiologies included preeclampsia, eclampsia, postpartum psychosis and cerebral venous thrombosis in postpartum RESLES cases without RCVS in the literature. Overall, the imaging spectrum of RCVS includes cerebral infarction, convexity subarachnoid hemorrhage,
lobar intracerebral hemorrhage and posterior reversible encephalopathy syndrome like (PRES-like) reversible cerebral edema (3). With other three previous cases, our experience adds typical RESLES lesion to the RCVS imaging spectrum. Other associations of RESLES include infections, seizures, anti-epileptic drug withdrawal and metabolic disorders (7). The presenting symptoms are varied, and include mental status changes, seizures, vertigo, visual and motor abnormalities, ataxia, tremor, psychosis and hallucinations (21).

Unlike three previous cases with postpartum RESLES with RCVS, where the initial symptom was thunderclap headache, our patient presented with postpartum psychosis without headache. A RESLES case presenting with only postpartum psychosis was published by Udaya et al; but in that case angiography was normal (15). Our case is the first in the literature, and thereby unique, as postpartum RVCS, RESLES and psychosis altogether co-exist. It should be noted that the psychosis of the patient worsened with anti-psychotics but improved with nimodipine.

The mechanism underlying RESLES has not been unveiled yet. It is speculated that due to the high density of crossing fibers, the myelin sheaths of splenial fibers are prone to water and electrolyte deposition. It has been suggested that ion transfer mechanisms of these axon bundles, which are very tightly arranged in the central region of the sCC, are insufficient to pump intracellular fluid into the extracellular space in the setting of pre-eclampsia. This relative insufficiency may result in intra-myelin edema and myelin vacuolization (21). Another suggested mechanism is cytokine induced glutamate excitotoxicity associated cytotoxic edema formation (22). Hiraga et al, on the other hand, proposed a simpler mechanism linking arterial tonus changes originating from pregnancy-related hormonal and metabolic alterations to both RVCS and RESLES (10,13).

In conclusion, we present a rare case with combination of postpartum psychosis, RESLES and RVCS. This case adds RVCS-induced arteriopathy among the causes of postpartum RESLES.

REFERENCES


Postpartum RCVS presenting with RESLES

Ethics
Informed Consent: The authors declared that informed consent form was signed by the patient.

Copyright Transfer Form: Copyright Transfer Form was signed by the authors.

Peer-review: Internally peer-reviewed.


Conflict of Interest: No conflict of interest was declared by the authors.

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<table>
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<tr>
<th>Authors</th>
<th>Year</th>
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<th>Comorbidities</th>
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<tr>
<td>Chen et al. [16]</td>
<td>2012</td>
<td>27</td>
<td>None</td>
<td>3 weeks</td>
<td>Recurrent headache, nausea and bilateral transient visual impairment</td>
<td>RESLES, normal MRA</td>
<td>Late postpartum preeclampsia</td>
<td>N/A</td>
<td>Complete resolution</td>
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<tr>
<td>Sekine et al. [9]</td>
<td>2012</td>
<td>34</td>
<td>Migraine, hypertension</td>
<td>Immediately</td>
<td>Thunderclap headache, seizure</td>
<td>T2 hyperintensities including the</td>
<td>Eclampsia</td>
<td>Olmesartan, magnesium sulfate, glycerol</td>
<td>Complete resolution</td>
</tr>
<tr>
<td>Takahashi et al.</td>
<td>2012</td>
<td>33</td>
<td>None</td>
<td>17 days</td>
<td>Thunderclap headache, transient speech disturbance</td>
<td>RESLES; severe bilateral proximal</td>
<td>Intravenous steroid treatment</td>
<td>N/A</td>
<td>Complete resolution</td>
</tr>
<tr>
<td>Curtis et al. [17]</td>
<td>2013</td>
<td>28</td>
<td>None</td>
<td>9 days</td>
<td>Recurrent bilateral transient visual impairment</td>
<td>RESLES; normal MRA and MRV findings</td>
<td>Possible subclinical postpartum preeclampsia</td>
<td>N/A</td>
<td>Complete resolution</td>
</tr>
<tr>
<td>Udaya et al. [15]</td>
<td>2014</td>
<td>21</td>
<td>None</td>
<td>2 weeks</td>
<td>Behavioral abnormalities, mutism, psychosis</td>
<td>RESLES; normal arteriograms and venograms</td>
<td>None</td>
<td>Lorazepam, Escitalopram, Olanzapine</td>
<td>Complete resolution</td>
</tr>
<tr>
<td>Altunkas et al. [18]</td>
<td>2015</td>
<td>19</td>
<td>None</td>
<td>20 days</td>
<td>Headache, nausea, vomiting</td>
<td>RESLES</td>
<td>None</td>
<td>N/A</td>
<td>Complete resolution</td>
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<tr>
<td>Hiraga et al. [10]</td>
<td>2016</td>
<td>28</td>
<td>None</td>
<td>Immediately</td>
<td>Intermittent Thunderclap headache</td>
<td>RESLES; segmental constriction of the left MCA and bilateral PCAs</td>
<td>None</td>
<td>None</td>
<td>Complete resolution</td>
</tr>
<tr>
<td>Liu et al. [19]</td>
<td>2017</td>
<td>29</td>
<td>None</td>
<td>42 days</td>
<td>Persistent right temporal headache</td>
<td>RESLES</td>
<td>None</td>
<td>Warfarin, mannitol, anti-epileptics</td>
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<tr>
<td></td>
<td></td>
<td>27</td>
<td>None</td>
<td>7 days</td>
<td>Parieto-occipital headache, increased perspiration, left sided numbness</td>
<td>RESLES</td>
<td>None</td>
<td>Interventional thrombolytic therapy, heparin, warfarin</td>
<td>Complete resolution</td>
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<tr>
<td>Yang et al. [20]</td>
<td>2019</td>
<td>23</td>
<td>None</td>
<td>10 days</td>
<td>Thunderclap headache, dizziness, blurred vision</td>
<td>PRES on the initial MRI, RESLES on the follow up MRI 10 days later (while asymptomatic)</td>
<td>Eclampsia-associated PRES</td>
<td>None</td>
<td>Complete resolution</td>
</tr>
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