H20 A Tremor in the Hand That Rocks the Cradle: Fatal Consequences of Postpartum Angiopathy

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After attending this presentation, attendees will better understand fatal outcomes of postpartum angiopathy, its variable presentation, differential diagnosis, and uncertainties of its incidence, pathogenesis, and treatment. Attendees will also be able to describe its importance as an often unrecognized cause of maternal death and the role of autopsy (especially neuropathological examination) in diagnosis.

This presentation will impact the forensic science community by fostering recognition and optimal examination of cases of postpartum angiopathy which is of benefit to family, clinicians, and public health.

Maternal mortality is of singular importance, usually scrutinized by public health authorities. Acute neurological symptoms occurring after a seemingly uncomplicated pregnancy and delivery are particularly alarming. A fatal intracerebral hemorrhage in an otherwise healthy postpartum woman is reported; the proximate cause of death was not recognized by clinicians and the case was almost not reported for definitive investigation.

A 26-year-old woman underwent uncomplicated term vaginal delivery and was discharged five days later. Two days later, she developed endometritis; following intravenous antibiotics, she was discharged after being prescribed oral antibiotics. On the 13th postpartum day, she complained of feeling "peculiar" and had two brief episodes of dysphasia. She later sustained a tonic-clonic seizure, recovered fully, and was transported to the hospital where a Computed Tomography (CT) scan of the head showed no acute changes. She was transferred to a tertiary center for monitoring and further investigation.

A chest X-ray, CT scan of the pelvis and abdomen, and abdominal/pelvic ultrasound were normal. A lumbar puncture was unsuccessful, but soon afterward she developed a severe headache. She vomited, became incoherent, and rapidly became comatose. A CT of the head showed a large hemorrhage within the left frontal lobe. Subsequent scans showed no structural cause of the hemorrhage. She deteriorated and met brain death criteria six days after initial neurological symptoms.

Autopsy revealed a healthy-appearing woman with features of medical intervention and critical illness. Degenerative endometrial changes consistent with her postpartum status were present. A probe-Patent Foramen Ovale (PFO) was noted. The brain showed a large hemorrhage apparently arising in the left striatum extending into the frontal white matter and rupturing into the ventricle (with acute hydrocepahalus), with secondary features of edema (transcranial/tonsillar herniation, left-sided sinus thromboses, and pituitary infarction). There was no identifiable structural etiology. No choriocarcinoma or septic foci were identified. The vessels showed no vasculitis or other abnormalities.

Postpartum angiopathy (Call-Fleming syndrome) usually presents within two weeks following normal pregnancy and parturition. It presents with an abrupt (maximum intensity within minutes), severe global or occipital headache (described as a "thunderclap" or "the worst") and/or focal, transient neurological signs. Definitive diagnosis consists of angiographic demonstration of multiple segmental narrowing of large- and medium-sized cerebral arteries. Vasospasm is transient and initial angiogram may be normal.

No incidence numbers are available, although fatal cases and small studies are widely reported. Multiple reports suggest under-recognition, "more common than appreciated," with a spectrum of outcomes, including disability. It is usually a benign condition, often grouped with "benign angiopathies" (a heterogeneous group of reversible cerebral vasospastic conditions), although this would seem to neglect uniquely postpartum physiology with hormonally mediated susceptibility to vasospasm and labile hypertension. Admissions for pregnancy-related strokes have increased since the early 1900s.

Pathogenesis is unclear. Vasospasm as a mechanism is favored by the typically benign outcome. Overlap with eclampsia is hypothesized and the use of vasospastic drugs (ergonovine, sumatriptan, bromocriptine) has been implicated in some cases.

Pregnancy is associated with an increased risk of cerebral events (venous thrombosis, subarachnoid hemorrhage, Intracerebral hemorrhage), coagulopathy, vasculitis, migraine, and mechanical complications due to epidural (spontaneous cerebrospinal fluid leak, meningitis), as well as eclampsia (25% developing postpartum). The differential diagnosis is large, but most (except for vasculitis) can be excluded by imaging and laboratory studies.

No definitive treatment is established. Favored strategies include calcium channel-blockers and steroids; others include immunosuppression, aspirin, MgSO4, hypervolemic therapy, and endovascular procedures (calcium channel blockers, balloon angioplasty, stents).

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Autopsy was helpful in this case in suggesting the etiology of this death, educating physicians, and excluding other causes, particularly given the PFO and potential for septic emboli.

Postpartum Angiopathy, Maternal Mortality, Postpartum Stroke

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