

# Abdominal Total Hysterectomy in a Patient with Isolated Angiitis of the Central Nervous System

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Isolated angiitis of the central nervous system (IAC) is an extremely rare disease characterized by vasculitis restricted to the vessels of the central nervous system (CNS) without other apparent systemic vasculitis. A patient with IAC suffers from recurrent cerebral strokes leading to death within months, if untreated<sup>1-5</sup>. It appears to be dangerous for the patient with IAC to undergo an operation under general anesthesia because the patient's cerebral blood flow is remarkably restricted. We report a case with IAC who had to be performed the abdominal total hysterectomy because of massive genital bleeding.

## Case Report

A 48-year-old female (weight 47 kg, height 155 cm) complained of genital bleeding. Gynecologists made a diagnosis of myoma uteri, about 10 cm in diameter, by echographic examination. Laboratory tests revealed extremely severe anemia; red blood cell (RBC) 870,000, hemoglobin (Hb) 1.5 g·dl<sup>-1</sup>,

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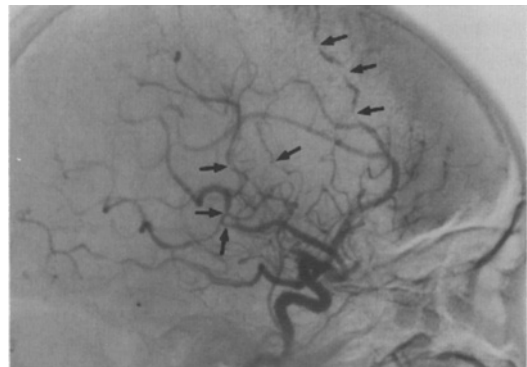


Fig. 1. Right internal carotid angiogram.

Arrows indicate multiple segmental areas of narrowing in both the anterior and middle cerebral arteries. Similar findings are also present on left internal carotid and vertebral angiograms.

and hematocrit (Ht) 5.4%. She was transfused 1,800 ml blood for 4 days and her laboratory tests were improved; RBC 3,330,000, Hb 9.1 g·dl<sup>-1</sup>, Ht 31.3%.

Seven days after admission, she complained of headache and developed a left hemiparesis. Computed tomography (CT) scan revealed a round hypodense mass, 2 cm in diameter, in the right cerebral posterior region. Cerebral angiography demonstrated multiple segmental areas of narrowing in

both the anterior and middle cerebral arteries, and in the basilar artery (fig. 1). Angiography revealed no other vascular abnormalities in arteries such as neck, kidney, and abdominal region. Laboratory data showed no autoimmune deficits. Although there was no leptomeningeal biopsy, the patient was diagnosed to be IAC ruling out some of the other possible diagnoses and was treated with i.v. betamethasone (12 mg per day for 2 weeks and 8 mg per day for 2 weeks). Her left hemiparesis began to improve 6 days after the beginning of the treatment. On the day before surgery, she could move her left arm and fingers slowly but not so skillfully, and she could manage to bend her left knee.

As her myoma prolapsed into the uterus, which might induce much more bleeding, she was scheduled for an abdominal total hysterectomy under general anesthesia 4 weeks after the onset of her CNS symptoms.

On arrival in the operating room, her blood pressure was 150/80 mmHg. Anesthesia was induced with fentanyl 0.1 mg, diazepam 5 mg, thiamylal 3 mg·kg<sup>-1</sup>, and vecuronium 0.1 mg·kg<sup>-1</sup> intravenously, and maintained with 60% nitrous oxide, enflurane (about 0.5%), and incremental doses of fentanyl. Intraoperative monitoring consisted of ECG, blood pressure, pulse oxymetry, capnometer, and temperature probe. Mean blood pressure was maintained between 80 and 100 mmHg, and end-tidal CO<sub>2</sub> between 38 and 43 mmHg. Nicardipine 0.1–1.0 μg·kg<sup>-1</sup>·min<sup>-1</sup> was continuously infused to increase cerebral blood flow. After the operation, the patient was extubated in the operating room.

No postoperative deterioration of her neurologic findings occurred. Betamethasone therapy was tapered. Her postoperative course was stable and uneventful.

## Discussion

IAC is an idiopathic, recurrent vasculitis confined to the CNS involving primarily small, and sometimes medium-sized, blood vessels. Early manifestations of disease include severe headache, altered mental function, and focal neurologic deficits, but clinical symptoms such as fever, myalgia, arthralgia, and arthritis, which occur frequently in other vasculitic syndromes, are generally not present in patients with IAC. IAC was originally described as invariably fatal, but recent studies have demonstrated that treatment with corticosteroids alone or in combination with cyclophosphamide may induce prolonged remission. Tissue biopsy is the most specific diagnostic test, but antemortem tissue diagnosis of intracranial vasculitis is very difficult. Therefore, cerebral angiography is the earliest and sometimes the only abnormal neurodiagnostic study<sup>1–5</sup>.

In our case, the patient had an abdominal total hysterectomy under general anesthesia only 4 weeks after the onset of her CNS symptoms. Some techniques of anesthetic management were necessary to prevent the patient from neurological deterioration.

Cerebral blood flow (CBF) is regulated by autoregulation to be almost constant at which mean arterial pressure is between 60 and 150 mmHg, but association with early ischemia is an alteration or loss of autoregulation. It is said that an alteration or loss of autoregulation lasts about 4 to 6 weeks<sup>6</sup>. This phenomenon involves pressure-dependent CBF, thus an anesthesiologist must carefully maintain perfusion pressure and blood volume to promote perfusion to areas of early ischemia<sup>7</sup>. In addition, the relationship between PaCO<sub>2</sub> and CBF is well documented<sup>8</sup>, but effects of ventilation of CBF in the presence of intracranial disease are complex<sup>9</sup>. The phenomena, called in-

tracerebral steal phenomenon<sup>10-12</sup> and inverse steal phenomenon or Robin Hood syndrome<sup>11</sup>, have been described. Generally, spinal or epidural anesthesia have the risk of rapid decrease in blood pressure. Furthermore, there was a possibility of much bleeding during the operation in this patients. Thus, we chose general anesthesia for this patient. In order to keep cerebral perfusion pressure, we maintained mean blood pressure between 80 and 100 mmHg. We also monitored end-tidal CO<sub>2</sub> and maintained it normocarbica, as we thought hypocarbica or hypercarbica was detrimental to our patient because of above reasons.

Additionally, we administered nicardipine, 0.1–1.0  $\mu\text{g}\cdot\text{kg}^{-1}\cdot\text{min}^{-1}$ , continuously to increase CBF, because nicardipine is a new calcium antagonist and this dose increases CBF without changing arterial blood pressure significantly<sup>13-16</sup>. Nicardipine appears to be very useful to control arterial blood pressure and increase CBF in such a patient with cerebral ischemic disease<sup>15,16</sup>, though whether intracerebral steal phenomenon occurs or not has not been proven.

In conclusion, we have described a case of the abdominal total hysterectomy under general anesthesia in a patient with isolated angiitis of the CNS. Some techniques of anesthetic management for keeping CBF were effective to prevent the patient from neurological deterioration.

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