A Pathologic Correlate of the ‘Steal’ Phenomenon in a Patient With Cerebral Arteriovenous Malformation

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SUMMARY A patient died after therapeutic bucrylate embolization and resection of a right occipital arteriovenous malformation, which received significant blood supply from the right posterior cerebral artery. At autopsy, there was marked diffuse neuron loss and gliosis in all cell layers of the right hippocampus — pathology which we hypothesize was secondary to a ‘steal’ of blood from this structure into the malformation. This report represents pathologic evidence for a phenomenon believed to be of major clinical importance in the symptomaticatology of many AVMs.

ENCEPHALIC ARTERIOVENOUS MALFORMATIONS (AVM’s) are thought to be congenital lesions that arise in fetal life. With time, such malformations may acquire new characteristics as a function of increased blood flow. The concept of a ‘cerebral steal’, secondary to the large blood flow and arteriovenous shunt through an AVM, as a cause of cerebral symptoms referable to brain tissue at some distance from the malformation, provides one frequently invoked explanation for the clinical findings when no definitive evidence for hemorrhage from the lesion exists. There is functional, radiologic, and cerebral blood flow data in support of the ‘steal’ concept, but little pathological evidence of its existence to date. This report illustrates a case with pathological documentation to support the idea of a cerebral ‘steal’ in the brain of a patient with an AVM.

Case Report

The patient, a 62-year-old male who had worked for several years as a machinist, experienced a 24-hour episode of sudden loss of consciousness in June, 1983, and was subsequently confused and drowsy with left hemiparesis. Radiologic investigations showed the presence of a right occipital AVM. Two further seizures occurred but within four to five weeks he was neurologically normal. Five months later, he experienced another seizure. Three weeks prior to admission to the University Hospital (23 May 1984) he began to have increasing seizure frequency. The seizures were described as consisting of chewing and drooling and at least one was of the grand mal type. His admission medications included anticonvulsants, thiamine and chlor Diazepoxide. Past medical history was remarkable for labile hypertension, removal of a fatty suboccipital tumour in the 1960s, and resection of a colonic carcinoma in 1968. Prior to June, 1983, there had been no documented history of headache, visual disturbance, behavioural or personality changes.

On the final hospital admission, general physical examination showed a 4 x 4 cm soft, non-tender, mobile suboccipital mass in the scalp. On neurologic assessment, he was mildly drowsy and inappropriate but oriented and not aphasic. There was a left homonymous hemianopsia, a slight decrease in pinprick perception on the left side of the body but no other significant sensory abnormality. Reflexes were symmetrical and plantar responses were flexor. There was diffuse mild weakness and truncal ataxia, and he required assistance to walk.

CT scan confirmed a right occipital AVM with a large area of enhancement confined to the occipital pole, associated calcifications and enlarged vessels going toward the region of the AVM. Low density in the white matter adjacent to the right occipital horn was noted. Cerebral angiograms on the second hospital day revealed a right occipital pole AVM supplied by the right middle cerebral artery (MCA), right occipital artery, posterior branch of the right middle meningeal artery, and right posterior cerebral artery (PCA). Four days after angiography, the patient underwent bucrylate-tantalum embolization with obliteration of the right MCA and right occipital feeder arteries. A further embolization attempt on the next day was abandoned due to inability to selectively catheterize the right vertebral artery. Two days after embolization, the patient had a right parieto-occipital craniotomy for excision of the AVM. Pathologic examination of the resected specimen showed typical features of an AVM, with some unusual microvascular changes as described below, and focal ossification — indicating the longstanding nature of the lesion.

On the tenth hospital day, he developed mild transient left hemiparesis and fever. On the sixteenth hospital day, the patient became short of breath, confused and hypotensive. A CT scan revealed no evidence of cerebral hemorrhage. He had a cardiopulmonary arrest that evening secondary to a pulmonary embolus, and pulmonary artery embolectomy was attempted. Postoperatively, the patient was brain dead and expired on the eighteenth hospital day.

Pathological Findings

General necropsy findings included small bilateral pleural effusions, acute congestion of the liver, spleen and kidney, cardiomegaly, bilateral pulmonary thromboemboli, and bilateral large pulmonary infarcts. Two small hemangiomas were seen in the liver.

The brain was diffusely swollen (fresh weight 1580
grams) and there was bilateral uncal and cerebellar tonsillar herniation. The site of the right occipital AVM resection was moderately hemorrhagic and (on coronal sections cut at a thickness of precisely 1 cm) surrounded by dilated, thrombosed blood vessels. As ascertained from the resection margin (fig. 1) the AVM had been confined to the occipital pole, extending at most 3–3.5 cm anteriorly.

Microscopic sections through the right occipital lobe near the edge of the AVM showed ferruginization of many small vessels in the cortex and adjacent white matter — the appearance was similar to that seen in cases of encephalotrigeminal angiomatosis (Sturge-Weber syndrome). Bucrylate-tantalum mixture was seen within some larger vessels in the sections. There were many dilated vessels remaining in the AVM bed, some with deficient or absent elastic tissue and others with smooth muscle proliferation in their walls. Some vessels showed thrombosis with early organization, or focal mural necrosis, presumably secondary to the presence of nearby bucrylate.

Throughout the remaining right occipital lobe there was patchy demyelination, severe gliosis, recent anoxic-ischemic change, and necrosis with lipid-laden macrophages. The corpus callosum showed moderate gliosis with an old area of cortical necrosis near the splenium. Areas of the left occipital lobe showed patchy acute anoxic change. The rest of the brain and spinal cord showed patchy but extensive changes of anoxic-ischemic encephalomyelopathy. By histologic

![Figure 1](image1.png)

**Figure 1.** Posterior view of the fixed brain shows the region of right occipital pole from which the AVM was resected (arrow).

![Figure 2](image2.png)

**Figure 2.** Whole mounts (PAS-stained) of the left (2a) and right (2b) hippocampi at the same coronal level, i.e., level of the lateral geniculate nuclei, which are identified (curved arrows). Photographs are at identical magnifications. Whereas a normal granule cell layer (arrowhead) and pyramidal cell layer (long arrow) are identified on the left, both structures are severely depleted of neurons and gliotic on the right. Scale marker represents 1 mm.
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Figure 3. Corresponding sections taken from regions roughly encompassing Spielmeyer sectors (h2 zones) of pyramidal cell layers, i.e., regions designated by long arrows in Fig. 2. Normally populated pyramidal cell layer on left (3a) contrasts with severely gliotic cell layer on right (3b). Cresyl violet stain, both \( \times 140 \).

Criteria, changes were consistent with a severe hypoxic event at the time of the cardiopulmonary arrest, two days before the patient's death. However, the hippocampi showed a marked asymmetry (Fig. 2). Whereas the left hippocampus (Fig. 2a) showed patchy slight eosinophilia of neurons (recent anoxic change) the right hippocampus showed severe diffuse longstanding loss of neurons in all portions of both the granule cell and pyramidal cell layers (Fig. 2b), with a profound reactive gliosis (Fig. 3). The right lateral geniculate nucleus was moderately depleted of neurons and gliotic.

The only other neuropathologic findings of note were a small schwannoma of the left vagus nerve, and anterior pituitary necrosis in keeping with the changes of 'non-perfused brain'.

Discussion

For decades, it has been believed that a 'steal' of blood by the AVM and away from normal cerebral tissue can produce focal neurologic signs and symptoms referable to the area of brain from which effective flow is 'stolen'. Structurally, the reasons for this seem apparent when one visualizes the massively dilated vessels (feeders and draining veins) adjacent to an AVM, as in this case. The 'steal' phenomenon has been clinically documented and verified by the use of fluorescein angiography and regional cerebral blood flow (CBF) studies. 'Steal' is one of several mechanisms by which an AVM may produce focal neurologic signs/symptoms, another important one being subclinical or 'silent' hemorrhage. The etiology of all signs/symptoms in a given patient with AVM is sometimes not even apparent.

Pathologic documentation of a structural correlate for the 'steal' effect has been scanty. This may represent the relative scarcity of large AVMs in a given autopsy population and/or the rarity of the phenomenon, or at least definite neuropathologic findings thereof. Subtle neuron loss and gliosis in the territory of a vessel that also supplies an AVM may be difficult to detect, for instance, without rigorous morphometric assessment of appropriate sections. This case represents our first encounter with such specific pathology, though we have examined in detail six brains from patients who died with AVMs or after treatment of AVMs.

We feel confident in ascribing the severe right-sided hippocampal sclerosis to a 'steal' effect for several reasons. The AVM received an important component of its blood supply from the right PCA. Although there has been ongoing debate as to the detailed microvascular supply of the mammalian (including human) hippocampus, all authors agree that the PCA constitutes the main vascular supply to this structure, though branches of the anterior choroidal artery may make a small contribution in some species. Though both the AVM and degenerate hippocampus were in the same hemisphere, they were separated on coronal sections by at least 6 cm of relatively unaffected cerebral parenchyma, i.e. the hippocampal pathology cannot be ascribed to direct compression or distortion by the AVM itself. Although the patient had experienced seizures, which are known to be correlated with hippocampal sclerosis, the change of severe neuron loss and gliosis was confined to the right hippocampal formation, whereas the left was well preserved apart from acute anoxic change. Furthermore, the changes in the right hippocampus extended through all cytoarchitectural zones of the pyramidal cell layer and the dentate fascia, while cell damage related to epilepsy is in general well-localized within the pyramidal cell layer, and there is often sparing of the h2 or Spielmeyer sector with most severe involvement of the h1 and h3 zones in the pyramidal cell layer.

That the hippocampus demonstrated neuron loss and gliosis, while other right hemisphere structures remote from the AVM were relatively spared, may simply reflect the known predisposition of the hippocampal formation to show the effects of anoxic-ischemic insults.
Major Cerebral Arterial and Venous Disease in Osteopetrosis


SUMMARY Two patients with osteopetrosis were studied in whom severe stenosis of one or both internal carotid arteries was demonstrated. One patient had autosomal dominant osteopetrosis and the other patient had the autosomal recessive form of the disease. In one patient, probable occlusion of one internal jugular vein and retrograde thrombosis of the contributing dural venous sinuses was present. Venous drainage of parts of the brain occurred through dilated emissary and scalp veins. It appears that major extracranial vessels may be impinged upon by dysplastic bone in osteopetrosis, although this is the first report of such an occurrence. A posterior fossa aneurysm was present in one case, and may have been related to abnormal intracranial hemodynamics.

OSTEOPETROSIS is a rare metabolic bone disease characterized clinically by pathological fractures, bone marrow failure and neurological deficits. Cranial nerve palsies and blindness with optic atrophy are the commonest neurological manifestations of the disorder.

Cerebrovascular complications so far described include intracerebral hemorrhage, subdural hematoma, cerebral venous thrombosis, and subarachnoid hemorrhage. On occasion, dilated scalp veins of uncertain significance have been observed in affected infants. There are no reported cases of severe intrapetrous stenosis of the internal carotid arteries, or of occlusion of major veins or dural sinuses.

Case 1

K.K., a 31-year-old right-handed woman, experienced a sudden severe headache without loss of consciousness or neurological deficit. Angiography subsequently revealed a ruptured aneurysm at the superior cerebellar/basilar artery junction.

The patient had osteopetrosis as had her father (fig. 1). Her mother was unaffected and she did not know the condition of her sibling from whom she had been separated since childhood. She had no children. She had suffered over 30 pathological fractures: four fractures of the left hip occurred in an 18 month period, necessitating internal fixation. Several rib fractures had occurred during sexual activity. Poor vision had been present since birth and was nonprogressive. She had always been mildly anemic. No anosmia, diplopia, deafness, facial paresis, or facial numbness had been experienced.

On examination, she was 4'10" tall and scaphocephalic. Blood pressure was 100/70 and a soft systolic ejection murmur was present. There was no hepatosplenomegaly or lymphadenopathy. The right hip was externally rotated and both hips were limited in range of motion. All pulses were present including those of the carotid, supratrochlear, superficial temporal and facial arteries. No cervical bruits were present.

References


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