Neuroradiology (1984) 26: 381–386

Neuroradiology

Vascular steal associated with vein of Galen aneurysm

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Summary. Seven patients presented with neonatal arteriovenous malformation and vein of Galen aneurysms. Six had a large degree of vascular steal demonstrated by cerebral angiography and all had significant parenchymal loss on initial CT. Angiographically, the steal affected the ophthalmic artery and branches of the middle and anterior cerebral arteries. Repeat CT in two patients, one of whom was untreated for 7 months, revealed pronounced atrophy. Patients with neonatal vein of Galen aneurysms present with parenchymal loss on CT which is related to vascular steal and may be progressive.

Key words: Vascular steal – vein of Galen aneurysm – visual loss – atrophy

Aneurysms of the vein of Galen (VGA) are well known vascular abnormalities resulting from either a direct arteriovenous shunt, or an arteriovenous malformation that drains into and dilates the vein of Galen. In some cases there is a combination of these two lesions [1, 2]. On computed tomography (CT) the VGA appears as a round midline high absorption mass just above and behind the third ventricle. This is connected to a prominent straight sinus and torcula. Following intravenous contrast CT reveals enlarged feeding and draining vessels. These lesions may calcify or demonstrate thrombosis [3–5]. In our report, another interesting and important aspect, the vascular steal, is stressed. Problems associated with this phenomenon in VGA include visual deterioration, seizures, and significant brain parenchymal loss. Although parenchymal loss and seizures secondary to the vascular steal have been recognized previously, its extent and fulminance deserves further emphasis [6, 7].

Table 1. Clinical data

Case	Age of presentation	Sex	Clinical presenting findings	Treatment
1	birth	M	cranial bruit seizures congestive heart failure	surgery
2	13 weeks	M	cranial bruit seizures head 2 standard deviations above normal optic pallor no visual pursuit	surgery
3	birth	M	cranial bruit seizures congestive heart failure optic pallor disconjugate gaze without visual pursuit	none
4	birth	M	cranial bruit seizures congestive heart failure	surgery embolization
5	birth	M	cranial bruit seizures congestive heart failure	surgery
6	birth	M	cranial bruit seizures congestive heart failure	none
7	birth	F	cranial bruit seizures congestive heart failure	none

Table 2. Radiographic findings

Case	CT following intravenous contrast[a]	Major feeding arteries
1	parenchymal loss in insula and over convexity ventricles enlarged serpiginous vessels with dilated vein of Galen and straight vein 2 years later – marked loss of central and cortical parenchymal on side of persistent steal	posterior cerebral
2	parenchymal loss in insula and over convexity ventricles enlarged serpiginous vessels with dilated vein of Galen and straight sinus	anterior cerebral posterior cerebral
3	ventricles slightly enlarged with parenchymal loss over convexity serpiginous vessels with dilated vein of Galen and straight sinus 7 months later – marked diffuse loss of central and cortical parenchyma	anterior cerebral posterior cerebral
4	parenchymal loss in insula and over convexity ventricles slightly enlarged serpiginous vessels with dilated vein of Galen and straight sinus	posterior cerebral
5	enlarged ventricles; parenchymal loss over convexity serpiginous vessels with dilated veins of Galen and straight sinus	anterior cerebral posterior cerebral
6	parenchymal loss in insula and over convexity ventricles enlarged serpiginous vessels with dilated vein of Galen and straight sinus	posterior cerebral
7	parenchymal loss in insula and over convexity; 3rd ventricle enlarged serpiginous vessels with dilated vein of Galen and straight sinus	angiography was not performed

[a] Conray 60% (2cc/KG). Mallinckrodt Inc., St Louis, Missouri (Iothalamate Meglumine)

Subjects and methods

Table 1 summarizes the clinical findings in seven patients. Six patients had transfemoral angiography including both carotid arteries and one vertebral artery. The seventh patient died before angiography could be done. CT scans were performed on the EMI 1010, the Philips Tomoscan 300, or the General Electric 8800.

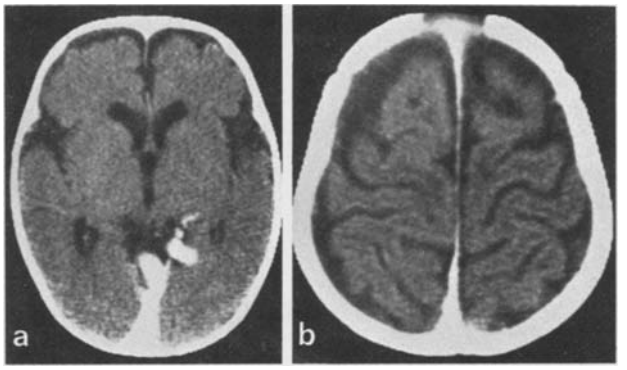

Fig. 1 a and b. Case 6. **a** Contrast enhanced CT showing loss of parenchyma in the frontal and insular regions. Mild ventricular dilatation is also present. Enhancing vascular structures are noted posteriorly. **b** Higher CT slice exhibiting cortical parenchymal loss

Results

The radiographic findings are summarized in Table 2. There were six males and one female who presented at birth with cranial bruits and seizures. Six patients had congestive heart failure. All showed significant parenchymal loss and ventricular dilatation on CT (Fig. 1). In two patients follow-up CT scans (7 months and 2 years later) revealed increase in the parenchymal loss.

In all patients undergoing angiography there was enlargement of major cerebral arteries (anterior and/or posterior cerebral) draining into a dilated vein of Galen. In all cases the ophthalmic artery filled predominantly retrogradely via internal maxillary artery collaterals or bidirectionally rather than antegrade from the internal carotid artery (Figs. 2 and 3). Optic pallor was noted in two patients (cases 2 and 3) which appeared to be reversed following surgery (case 2). In all cases a steal was also noted from branches of the middle and anterior cerebral arteries (Figs. 2–4). This corresponded to the pattern of parenchymal loss seen on CT (Fig. 1). Four patients (cases 1, 2, 4 and 5) were surgically treated with ligation of feeding arteries surrounding the vein of Galen. Postoperative angiography in cases 2, 4, and 5 exhibited a partial reversal of the vascular steal with antegrade filling of the ophthalmic artery and improved blood flow in the middle and anterior cerebral arteries (Fig. 4). In case 1 a repeat arteriogram performed approximately 2 years after surgery still

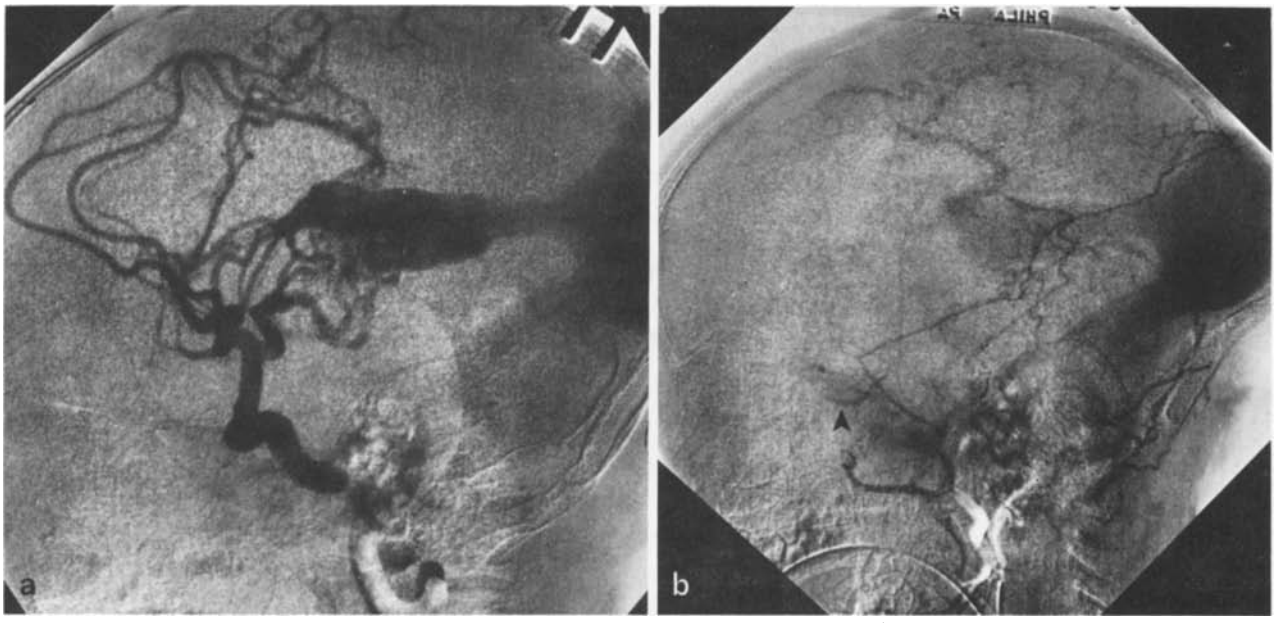

Fig. 2 a and b. Case 3. **a** Internal carotid injection without filling of ophthalmic. Note poor filling of anterior and middle cerebral branches. **b** External carotid injection with retrograde ophthalmic filling *(arrowhead)*

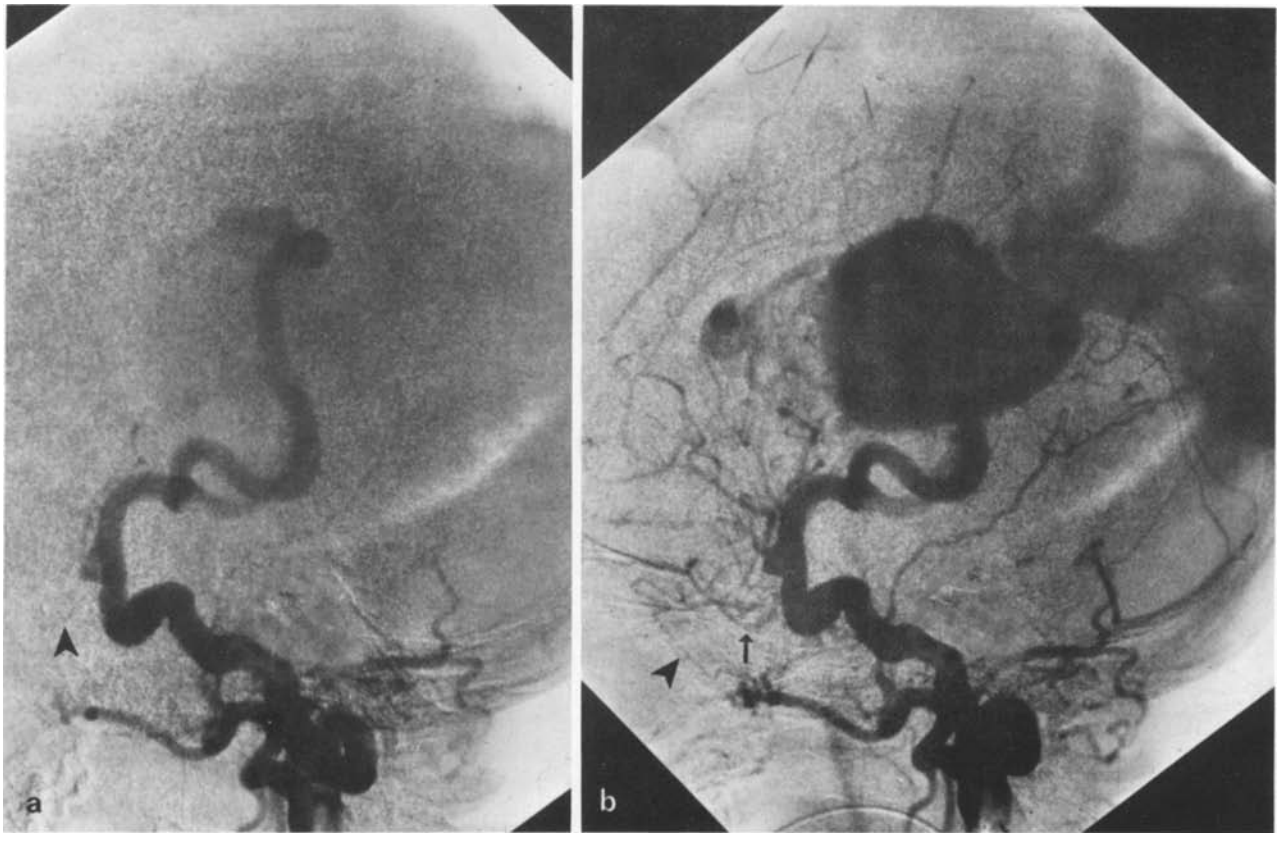

Fig. 3 a and b. Case 1. **a** Early common carotid injection demonstrating enlarged posterior cerebral artery draining into VGA. There is lack of filling of the middle cerebral artery and poor filling of the ophthalmic artery *(arrowhead).* **b** Late arterial phase exhibiting retrograde blood flow in the ophthalmic artery *(arrow)* via internal maxillary collateral *(arrowhead)*

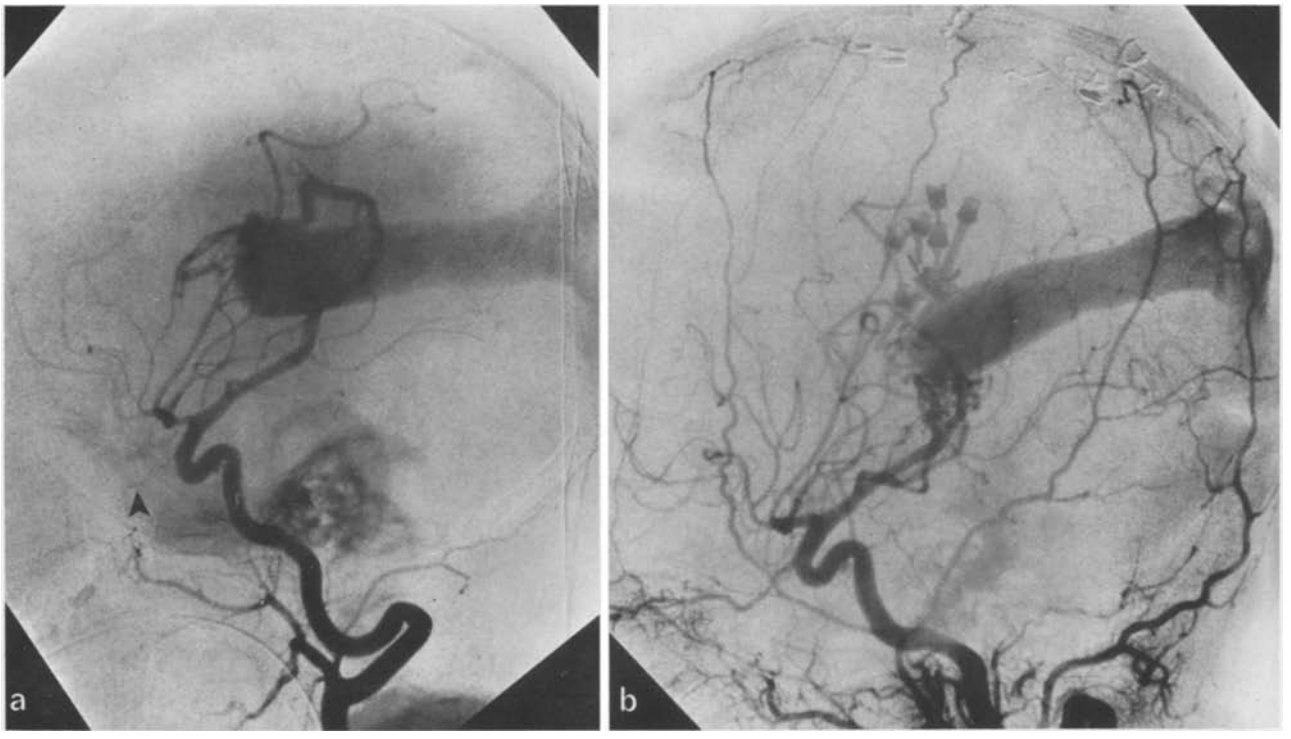

Fig. 4 a and b. Case 2. **a** Common carotid injection with dilated posterior cerebral artery and poor filling of anterior cerebral, middle cerebral and ophthalmic arteries *(arrowhead)*. **b** Postoperative common carotid injection revealing improved antegrade flow in ophthalmic artery and improved filling of the middle cerebral artery

revealed poor filling of one ophthalmic artery as well as persistent steal from the left middle cerebral artery (Fig. 5 a). There was improved filling of the right middle cerebral artery (Fig. 5 b). CT showed significant atrophic changes in the left hemisphere which correlated with the steal (Fig. 5 c). Following surgery cases 1 and 5 experienced decreased congestive heart failure, while in cases 1, 2, and 5 growth and weight gain were noted. Case 3 has made no progress in developmental milestones. Case 4 required embolization after surgery to control heart failure but is now growing and gaining weight. Case 6 has not been treated. Case 7 expired before angiography could be performed.

Discussion

VGA has been classified according to the age of presentation of symptoms [8]. All of our cases occurred in the neonatal period which accounts for their fulminant symptomatology. Each patient presented with seizures and cranial bruit while six of seven patients had congestive heart failure. The presumptive clinical diagnosis of VGA was confirmed by CT scanning and subsequent cerebral angiography.

Our cases are most interesting when CT is analyzed with respect to the angiographic findings. In all cases CT revealed significant cortical parenchymal loss which correlated with the vascular steal from the middle and anterior cerebral arteries. Predominantly retrograde flow in the ophthalmic artery via internal maxillary artery collateral occurred as a result of the sumping of blood flow by the VGA. The ophthalmic artery steal may be responsible for the pale optic disc seen in one patient and has previously been reported in a patient with a large intracranial arteriovenous malformation [9].

Following surgery (cases 1, 2, 4 and 5) a repeat arteriogram showed partial reversal of the intracranial steal with normal filling of many middle and anterior cerebral artery branches that initially had not filled. The angiographic findings correlated with the concurrent clinical examination which included improvement in developmental milestones and growth.

Case 3 represents the most extreme example of vascular steal. The first CT scan performed shortly after birth showed mild parenchymal loss (Fig. 6 a). Angiography confirmed the enormous vascular steal from the anterior and middle cerebral vessels (Fig. 2). Notice that there is no filling of the ophthalmic artery. If one compares this scan with the second study

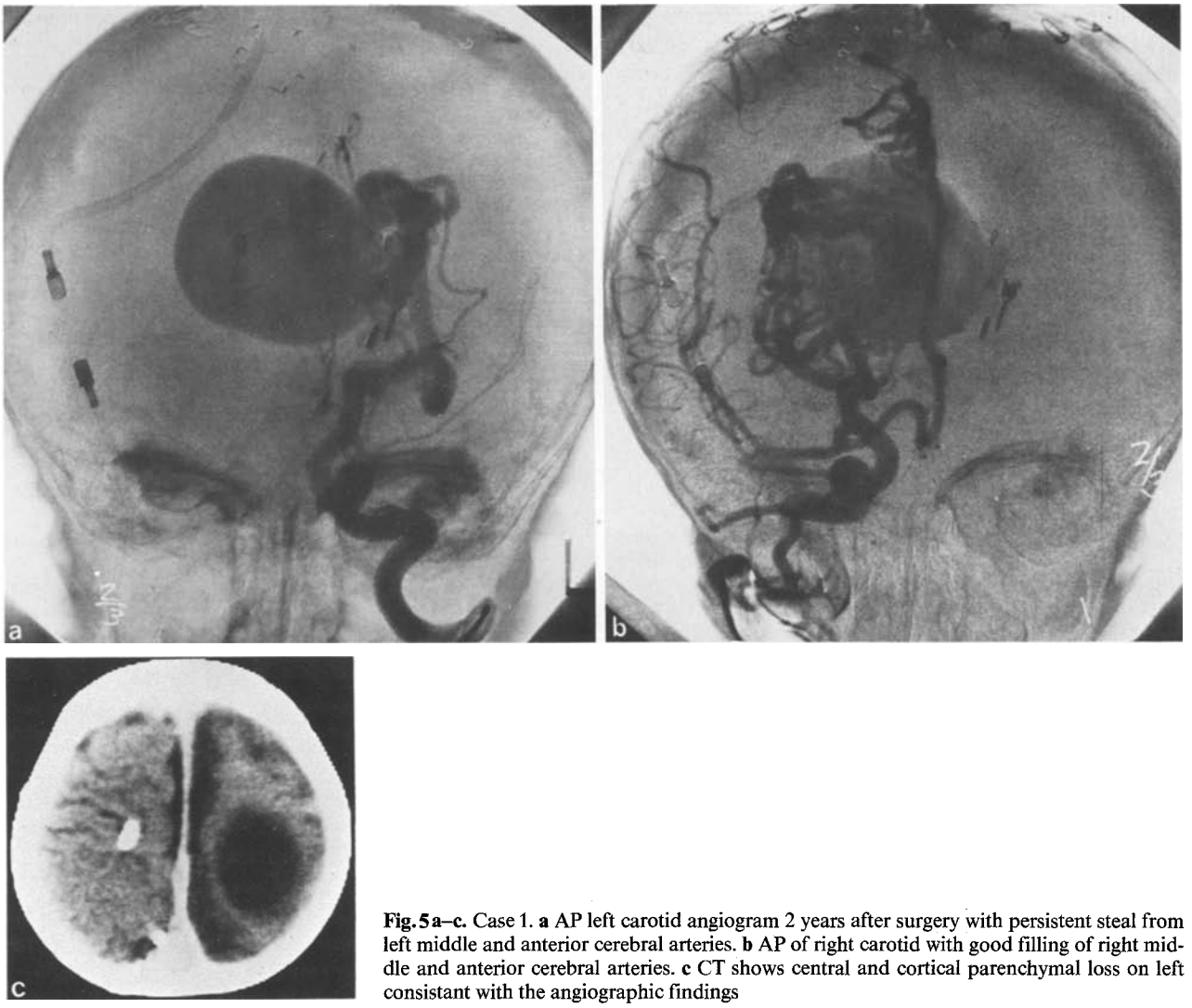

Fig. 5a–c. Case 1. **a** AP left carotid angiogram 2 years after surgery with persistent steal from left middle and anterior cerebral arteries. **b** AP of right carotid with good filling of right middle and anterior cerebral arteries. **c** CT shows central and cortical parenchymal loss on left consistant with the angiographic findings

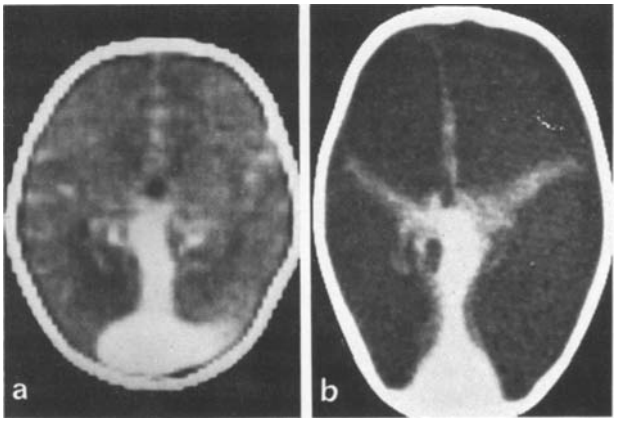

Fig. 6a and b. Case 3. **a** Enhanced CT performed at 1 week of age displaying VGA. **b** Enhanced CT at 7 months of age with marked parenchymal loss

done at age 7 months the amount of parenchymal loss is marked (Fig. 6b).

There are many complications associated with VGA including spontaneous thrombosis, hemorrhage, hydrocephalus, and congestive heart failure [10–15]. In general, the earlier the malformation presents, the more dramatic its course [16]. Surgeons have been reluctant to operate on these very young patients and their prognosis has been poor. All seven of our patients demonstrated parenchymal loss on their initial CT scan as well as seizure activity clinically. These findings may be attributed to focal ischemia produced by the vascular steal. In very young children with developing brains the effect of this vascular steal may become more pronounced as the child ages. We believe that there is a risk of significant brain parenchymal loss secondary to vascular steal as exemplified in cases 1 and 3.

Although surgery alone did not obliterate the VGA's in any of our operated cases, it clearly changed the hemodynamics. Postoperative angiography revealed a decrease in the vascular steal and a concomitant increase in blood flow to the orbit and brain parenchyma (Fig. 4b). Case 3 is a noteworthy example of what may happen when VGA's are left untreated. In this patient blindness and cerebral atrophy ensued rapidly (7 months) precluding surgical intervention. Failure to institute proper therapy may result in cerebral atrophy and possibly visual impairment. Early recognition and correction of vascular steal in VGA may prevent progressive loss of brain parenchyma.

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Received: 29 August 1983

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