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Case Report

Cavernous Hemangioma of Optic Chiasm, Optic Nerves and Right Optic Tract

Case Report and Review of Literature*

Herbert J. Manz[1], Larry H. Klein[1], Joseph Fermaglich[2], Jorge Kattah[2], and Alfred J. Luessenhop[3]

Departments of Pathology[1], Neurology[2] and Surgery[3] (Division of Neurosurgery), Georgetown University Medical Center, Washington, D.C., USA

Summary. Diminishing right ocular visual acuity for three weeks in a 30 year old man was confirmed by examination. Bilateral scotomata and bitemporal hemiachromatopsia indicated a chiasmal lesion; reduced visual acuity and Marcus Gunn pupil of the right eye and left relative temporal hemianopia indicated asymmetric involvement. Erythrocytes in the CSF verified a suspected subarachnoid bleed; contrast-enhanced CAT scan demonstrated a suprasellar mass. A cystic, multiloculated, bluish mass distorted the right optic nerve, tract, and chiasm. A hematoma was evacuated and biopsy revealed a cavernous hemangioma of the right optic nerve. Post-operatively, visual acuity has recovered in the right eye but a left homonymous temporal hemianopia has developed.

Key words: Cavernous hemangioma – Vascular malformation – Visual system lesions.

Introduction

As congenital anomalies of blood vessel formation, vascular malformations are tumor-like, but non-neoplastic errors of tissue development or architectural organization (Willis, 1962). Despite their non-proliferative, non-neoplastic nature, they may cause progressive signs and symptoms (Russell and Rubinstein, 1977). Ongoing dilatation of the abnormal vascular channels causes enlargement of the mass and pressure atrophy of neural parenchyma; anastomoses with vessels in the vicinity may result in ischemia due to shunting of blood ("steal" phenomenon); recurrent small or massive hemorrhage may destroy adjacent

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Address offprint requests to: Dr. H.J. Manz, Department of Pathology, Georgetown University, 3900 Reservoir Road, N.W., Washington, D.C. 20007, USA

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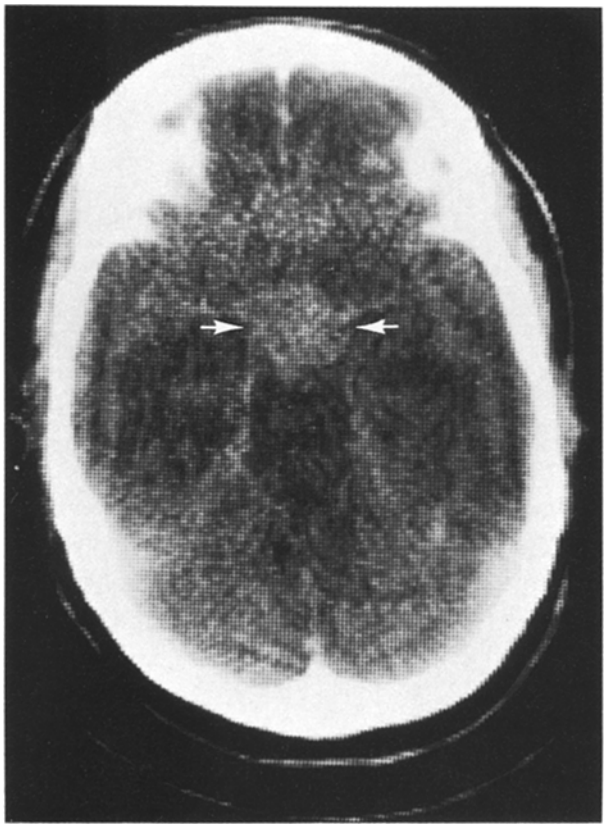


Fig. 1. Following injection of Hypaque, a large enhancing suprasellar mass is observed

tissue; thrombosis may lead to infarction (Russell and Rubinstein, 1977). The inability of the adult human central nervous system to regenerate, but rather to respond to structural injury by Wallerian degeneration and astrogliosis, also accounts for the failure of functional recovery (Robertson and Dinsdale, 1972). However, cerebral edema (including edema of optic nerves) and biochemical or metabolic abnormalities may be amenable to appropriate measures so that restitution of some function may result (Fishman, 1975); evacuation of a hematoma may reduce the severity of deficits.

In the previous report of 71 patients with intracranial and cranial arteriovenous malformation (AVM) at Georgetown University Medical Center, almost 50% had signs and symptoms referable to the visual system (Forman et al., 1975). In all but two patients could the visual field loss be explained by direct impingement of the arteriovenous malformation on optic tract, radiation or visual cortex. Of the two other patients, one had altitudinal defects in both visual fields compatible with retinal vascular occlusion, the second had a visual defect larger than predictable from the angiographic appearance and size of the AVM. In none was the optic nerve or chiasm directly involved.

In this report we describe the neuropathologic findings in a patient with a cavernous hemangioma of the optic chiasm, nerves and tract. The literature is reviewed and the clinicopathological correlation discussed. A brief communication with emphasis on clinical aspects has been published (Fermaglich, Kattah, and Manz 1978).

Case Report

Progressive visual failure of the right eye over a three week period brought a previously healthy 30 year old white man to medical attention. Intermittent, pulsating frontal headaches, more pronounced with recumbency, were at times accompanied by nausea and vomiting. There was no loss of weight, libido, or potency.

Vital signs were normal. While visual acuity in the left eye was 6/6, there was only perception of hand motion with the right eye. Goldman perimetry demonstrated bilateral central scotomata, a left paracentral scotoma and a relative left temporal hemianopia. The Marcus Gunn pupillary reaction was elicited in the right eye. Ishihara colors could not be identified with the right eye, and the red color could not be identified in either temporal field. Ophthalmoscopy revealed bitemporal pallor and slight blurring of the right optic disc. Range of eye movements was normal. Apart from nuchal rigidity, the neurological and general physical examination were unremarkable.

Neuroradiologic studies, including carotid and vertebral arteriography, disclosed no abnormality. Only after injection of Hypaque did the CAT scan demonstrate a suprasellar mass (Fig. 1). Serum levels of pituitary hormones, testosterone, and cortisol were within normal range.

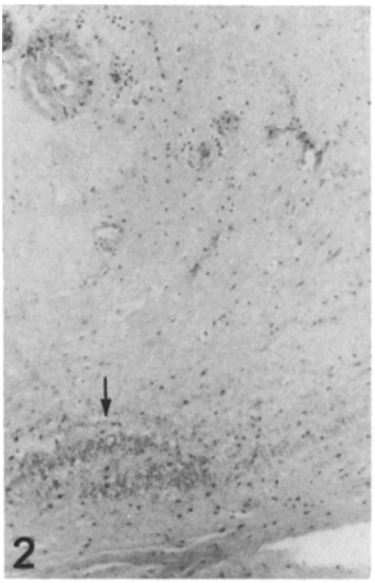
The *CSF* was under normal pressure and xanthochromic, containing 786 erythrocytes and 39 lymphcytes per cmm. CSF protein and glucose were normal and cultures and serologic studies for infective organisms negative.

At *right frontal craniotomy*, the right optic nerve was distorted by an intrinsic lobulated blue, multicystic mass extending across the chiasm to involve the left optic nerve and the right optic tract. Following evacuation of blood clot and necrotic debris, the chiasm became flat. Biopsy of the right optic nerve was performed for histopathologic study. Post-operatively, central vision in the right eye has improved to 20/70, but a permanent left homonymous hemianopia has developed.

Neuropathological Examination

The right optic nerve biopsy specimen was embedded in paraffin. Sections were stained with: hematoxylin-eosin (H & E); luxol-fast-blue hematoxylin-eosin (LFB-H & E) for myelin; Bodian's protargol for axons; phosphotungstic acid hematoxylin (PTAH) for astroglial fibrils and fibrin; Verhoeff-van Gieson for elastic tissue; Masson's trichrome for connective tissue and smooth muscle; and Mallory's for iron.

A small peripheral sector of optic nerve was fairly well preserved, being composed of campactly arranged, myelinated axons (Fig. 2); major portions consisted of haphazardly disposed, reactive fibrillary astrocytes, among which only isolated unmyelinated axonal fragments remained. Large numbers of hemosiderin-laden macrophages were present throughout, either singly or in large clusters. Chronic inflammatory cells lay scattered in the gliotic optic nerve, commonly as perivascular cuffs (Fig.3). Erythrocytes and fresh blood dissected along tissue planes; larger blood clots were present near conglomerate masses of irregular vascular channels (Fig. 4). The vascular spaces were haphazardly arranged and separated by glial scar and hemosiderophages. The vascular struc-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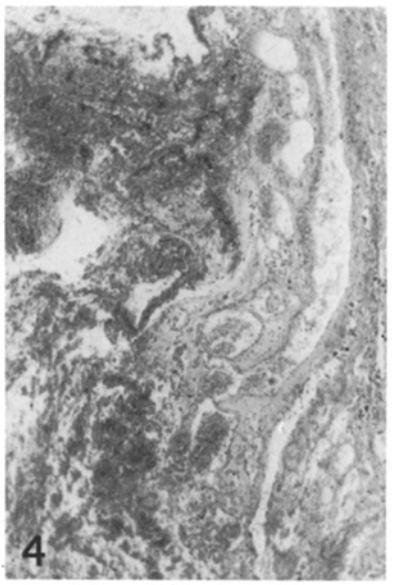
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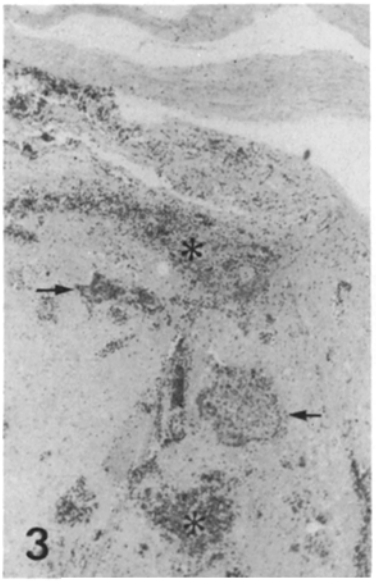
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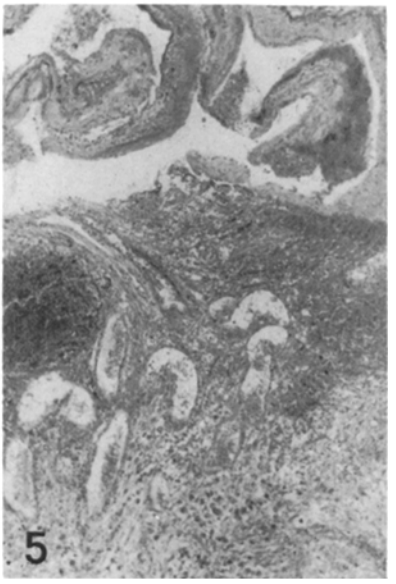
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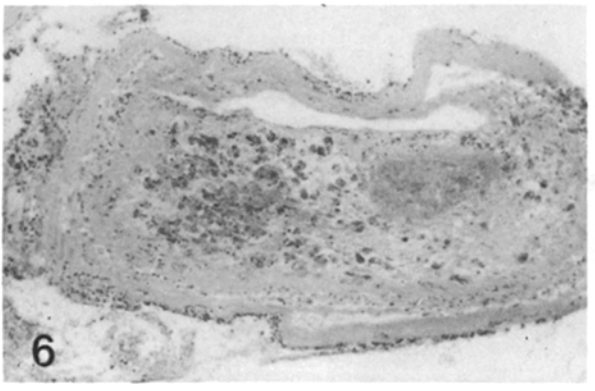
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tures ranged from small, tortuous venules having only rare smooth muscle cells embedded in their thin fibrous walls, to large, now collapsed, cavernous spaces with densely hyalinized, collagenous walls (Fig. 5). In none of the abnormal channels was an internal elastic lamina present and communications between arteries and veins could not be demonstrated on semi-serial sections. Vessels variably contained blood, fresh, or old and organized thrombus (Fig. 6).

Discussion

The differential diagnosis of the sudden onset of a neuro-ophthalmologic syndrome accompanied by meningeal reaction in a previously healthy young man offered various possibilities. Bilateral scotomata and bitemporal hemiachromatopsia indicated an optic chiasmal lesion, while the severe reduction in visual acuity of the right eye, right Marcus Gunn pupil and left relative temporal hemianopia indicated asymmetric involvement (Harrington, 1976). Space-occupying lesions from structures contiguous to the chiasm include meningioma of the tuberculum sellae, carniopharyngioma, and pituitary adenoma, as well as non-neoplastic masses such as berry aneurysm of the internal carotid or anterior communicating arteries (Jefferson, 1937; Harrington, 1976). They are more likely to compress than infiltrate. The neoplasms could produce sudden symptoms by undergoing central necrosis or hemorrhage, including extravasation of debris or blood into the CSF. Classically, pituitary apoplexy manifests as sudden headache, cavernous sinus syndrome with ophthalmoplegia, endocrinopathy, and meningeal irritation (David, Gargano, and Glaser, 1975; Banna, 1976). Primary meningeal diseases of subacute nature which could secondarily involve the chiasm include sarcoidosis (Delaney, 1977), and other granulomatous or carcinomatous meningitides (Ellner and Bennett, 1976). Rupture of epidermoid or dermoid cysts with resulting granulomatous menigitis has also been reported (DeKlerk and Spence, 1974). Intrinsic chiasmal lesions comprise gliomas, including glioblastoma (Condon and Rose, 1967; Gibberd, Miller and Morgan, 1973), demyelinating syndromes, such as acute multiple sclerosis (Walsh

Fig. 2. Peripheral rim of well-myelinated optic nerve is seen inferiorly (arrow) adjacent to subpial gliosis. Most of the optic nerve is gliotic and the vessels (left upper) are slightly hyalinized or surrounded by inflammatory cells. (LFB-HE; ×100)

Fig. 3. Irregular collections of blood (*) and clusters of hemosiderin-laden macrophages (*arrows*) are distributed throughout the gliotic optic nerve. In the upper field, large collapsed vascular spaces are surrounded by thick, fibrous walls. (LFB-HE: ×63)

Fig. 4. A multitude of vascular channels is sectioned transversely and longitudinally on the right, while blood clot and fresh blood are present on the left. (Trichrome ×63)

Fig. 5. Large, collapsed cavernous vascular space containing fresh thrombus is seen in the upper field, while in the center fresh hemorrhage surrounds smaller anomalous vessels. Hemosiderophages are scattered singly in the lower third of the field (Trichrome: ×63)

Fig. 6. An irregular venous channel surrounded by a hyalinized fibrous wall is virtually completely occluded by old thrombus in which macrophages have phagocytosed hemosiderin. Lymphocytes and erythrocytes are sprinkled along the adventitia. An elastic membrane cannot be identified. (Verhoeff-van Gieson elastica. ×100)

and Hoyt, 1969) or Devic's disease (Oppenheimer, 1976), inflammatory angiopathy (Cullen and Coleiro, 1976), and vascular anomalies (Brock and Dyke, 1932; Krug and Samuels, 1932; Wyburn-Mason, 1943; Fermaglich, Kattah and Manz, 1978). A definitive answer can frequently only be obtained by surgical exploration and neuropathologic study.

The most useful classification of abnormalities of blood vessels causing neurologic manifestations on the basis of their mass effect is that of Russell and Rubinstein (1977). They omit vasculitides, berry aneurysm, and thrombotic or embolic events with associated hemorrhage, infarction, edema and resultant mass effect. The true neoplasms include hemangioblastoma with proliferative activity of endothelial cells, as in the von Hippel-Lindau syndrome. The hemangiomas are comprised of capillary telangiectasia, cavernous hemangioma, and venous and arteriovenous malformations. McCormick (1966) adds another category, the varix, which occurs as a single enlarged, tortuous vein or group of veins anywhere in the central nervous system, but particularly on the dorsal surface of the spinal cord. The Sturge-Weber syndrome represents a subgroup of venous angiomas. In this entity, leptomeningeal congeries of veins and calcified capillary walls in the subjacent occipital cortex are associated with ipsilateral port-wine nevus of the face in the distribution of the ophthalmic division of the trigeminal nerve, and in a significant number of cases, with ipsilateral uveal angiomatosis (Urich, 1976).

According to Reese (1956), angiomatous malformations of the optic nerve and disc are rare, but involvement of the retina by hemangiomas is more common. Von Hippel-Lindau hemangioblastomatosis characteristically involves retina (including optic disc), and/or cerebellum, medulla oblongata, or spinal cord. Capillary hemangiomas of the Lindau type have been reported confined to the optic nerve; to its intracranial portion as an incidental postmortem finding by Verga (1930) and to its intraorbital segment during orbital decompression for exophthalmos by Schneider (1942). Cavernous hemangiomas of retina and optic nerve head may be accompanied by intracranial and cutaneous vascular malformations and be familial (Gass, 1971). Arterio-venous angiomas of the retina tend to be associated with similar intracranial aneurysms, more specifically with AVM of the midbrain tectum (Wyburn-Mason,1943). Direct continuity of the vascular anomaly from retina to midbrain with involvement of the intervening ispilateral optic nerve, chiasm, optic tract and metathalamus has been demonstrated in two such cases (the case reported by Krug and Samuels (1932) represents case 3 of Brock and Dyke (1932); the second is case 7 of Wyburn-Mason (1943)). An arteriovenous malformation near the chiasm caused demyelination and atrophy, as illustrated by Lindenberg, Walsh, and Sacks (1973).

Histopathologic features of the optic nerve, chiasm, and tract in such cases depict aberrant vascular channels distributed through the neural parenchyma. Destruction of axons and consequent Wallerian degeneration result in optic nerve and chiasmal atrophy causing visual impairment (Brock and Dyke, 1932; Krug and Samuels, 1932; Lindenberg, Walsh and Sacks, 1973). Rupture of the vascular anomaly with intraneural hematoma formation, aggregation of hemosiderophages and lipophages (Krug and Samuels, 1932), and massive fatal subarachnoid hemorrhage (case 7 of Wyburn-Mason, 1943) have also been recorded.

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