Giant suprasellar aneurysm associated with optic stalk agenesis and unilateral anophthalmos

Case report

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The authors report a patient with unilateral anophthalmos who had a large suprasellar aneurysm associated with aplasia of an optic nerve and the chiasmal commissure. The anatomical findings at craniotomy are described.

Key Words • optic nerve • optic chiasm • anomaly • microphthalmia • aneurysm

Congenital absence of an optic nerve and chiasmal commissure is one of the developmental consequences of agenesis or regression of an optic stalk. These anomalies of the anterior visual system have been reported in necropsy specimens, but their appearance at operation has not been recorded. This report describes the anatomical observations during craniotomy in a patient with unilateral anophthalmos who had a large suprasellar aneurysm associated with aplasia of an optic nerve and the chiasmal commissure.

Case Report

A 49-year-old man with anophthalmos on the right was admitted to San Francisco General Hospital with complaints of headache, memory loss, and unsteadiness.

Examination. Vision in his only eye (the left) was 20/40, with defective color perception and a total temporal hemianopia. The eye was of normal size, moved fully in all directions, and exhibited constant horizontal nystagmus of irregular amplitude that increased in gaze toward the right. The nystagmus remained horizontal during upward and downward gaze and was not influenced by gaze at a nearby object. Pupillary reactions of the eye were normal in response to light and accommodative stimuli. The ocular media were clear. There was no retinopathy and the retinal vessels appeared normal. The optic disc was abnormally small, slightly pale temporally, flat without an optic cup, and vertically oval with the retinal vessels emerging from its nasal margin at 9 o'clock. The nerve fiber layer surrounding the disc was abnormally thinned in the upper and lower sectors and absent in the temporal and nasal sectors. These changes in the disc and the peripapillary nerve fibers were consistent with hemiopic hypoplasia described in patients.
with congenital hemiplegia by Hoyt, et al.7

Examination of the ocular adenexa in the anophthalmic orbit showed the expected appearance of enophthalmos with a small palpebral fissure. In the depths of the conjunctival sac was a small firm nodule of white tissue that might have contained congenital remnants of neuroectodermal tissue. This nodule moved back and forth in apparent synchrony with the horizontal movements of the left eye.

Neurological examination confirmed a moderate defect in recent memory, a broad-based ataxic gait, and absent position and vibratory sensation with loss of deep tendon reflexes distally in the arms and legs. There were no signs of pituitary insufficiency. The clinical diagnoses were chiasmal syndrome of uncertain etiology, congenital absence of the right eye, and peripheral neuropathy from chronic alcoholism.

Neuroradiological investigations revealed a normal-sized right orbit, a tiny (2 mm) optic foramen on the right, and a curvilinear calcification above a normal-appearing sella. Pneumoencephalography showed a round filling defect in the suprasellar cisterns (Fig. 1). This filling defect caused a smoothly curved convex indentation in the anterior inferior third ventricle. The optic and infundibular recesses and the chiasmal commissure could not be identified. The suprasellar filling defect obscured all signs of the optic nerves in the suprasellar area. Selective internal carotid angiography showed a narrow ophthalmic artery and no choroidal crescent on the right. The left internal carotid study showed a normal ophthalmic artery and choroidal crescent and a suprasellar aneurysm originating from the left anterior cerebral artery. The aneurysm had an irregular, elongated, inferiorly-pointing lumen that was smaller than the mass which was demonstrated by the pneumoencephalogram (Fig. 2 left). Vertebral injections opacified normal-appearing basilar and posterior cerebral arteries. Asymmetry of the size of the occipital lobes or their vascularity could not be detected either by angiography or air study.

Operation. After the patient's peripheral neuropathy had improved, the suprasellar
region was explored with the intention of clipping and resecting the aneurysm. At craniotomy the left optic nerve was identified first at the optic foramen. It appeared healthy but was abnormally small in diameter and was deflected upward and laterally by a midline mass (Fig. 3). The left ophthalmic artery arose 1 cm distal to the dural entry of the carotid artery into the cranial cavity. It entered the left optic foramen beneath the optic nerve. Posteriorly the left optic nerve passed directly into the left optic tract as a single pathway without a vestige of a chiasm. Filmy arachnoid adhesions formed a bridge between this nondecussating optic "nerve" and the mass. The aneurysmal mass was firm, immobile, and yellow without attachment to the carotid trunks or the right anterior cerebral artery.

The right ophthalmic artery also arose 1 cm beyond the point where the right internal carotid artery entered the cranial vault and coursed anteriorly alone into the tiny right optic foramen. The tuberculum sellae, planum sphenoidal, anterior clinoid processes, and sphenoid wings appeared normal.

An atherosclerotic neck of the aneurysm was identified at the portion of the left anterior cerebral artery adjacent to its junction with the anterior communicating artery. A clip was applied across the neck and the fundus of the aneurysm was resected. At this point in the operation the lamina terminalis of the anterior third ventricle was visible as a thin-walled midline structure that did not contain any recognizable chiasmal commissure. A compressed infundibular stalk marked the posterior limit of the chiasm-free wall of the third ventricle. Histopathological examination of the mass confirmed its vascular origin.

Postoperative Course. After operation the patient recovered promptly. Angiography on the tenth day after operation showed no evidence of the aneurysmal sac (Fig. 2 right). Three weeks thereafter the patient was transferred to a special facility for alcoholics.

Discussion

The optic vesicles, from which the eyes and their optic nerves develop, appear during the neural-plate stage of embryogenesis, in the third week after conception.1 Experimentally, failure of vesicle formation or destruction of the vesicle from effects of toxins, infection, anoxia, or trauma, causes anophthalmos or severe microphthalmos with associated aplasia of the optic pathways.5,8 Clinically, causes of these optic vesicle defects are seldom defined, though heredity accounts for some.10

Mann8 distinguished three forms of anophthalmia: a primary form in which there is failure of formation of the optic vesicle from the anterior end of the neural tube, a secondary form resulting from developmental abnormality of the whole of the forebrain,
and a degenerative form in which the optic vesicles regress and disappear completely. The degenerative form merges imperceptibly with severe forms of microphthalmos in which the eye is no more than a nodule of connective tissue containing a few nests of undifferentiated neuroepithelial cells. All forms of bilateral anophthalmos are associated with absence of optic nerves, chiasm, optic tracts, and with gross anomalies of the geniculostriate system. 

Unilateral anophthalmos, depending on its cause, may be associated with partial or total absence of the chiasmal commissure. If growth of ganglion cell projections from the normal eye proceeds centrally without impediment to decussation through the forebrain anlage of the chiasmal commissure, the patient will have both crossed and uncrossed visual projections to the brain. The chiasm will then be hypoplastic. If decussation of ingrowing visual fibers from the normal eye is blocked by the forebrain abnormality that caused arrest of one optic vesicle, or if these decussating fibers degenerate after embryonic injury to the optic pathway above the chiasm, the patient will have only an uncrossed, or ipsilaterally projecting, visual system that operates through afferent hemianopic projections to one occipital lobe. In this circumstance unilateral anophthalmos and optic nerve agenesis may be associated with agenesis of the chiasmal commissure and retrograde hemiopic hypoplasia of the functioning optic nerve. Additional visual system anomalies may include absence of a recognizable lateral geniculate nucleus on the functioning side, and aplasia of the striate cortex in the blind occipital lobe. Our patient had unilateral anophthalmos with chiasmal agenesis. While we believe that his optic vesicle formed initially and then degenerated, our conclusions are speculative.

Congenital cerebral aneurysms may arise soon after mesodermal elements become tubular structures. The internal carotid arteries can be recognized in the embryo as early as the fourth week of gestation; components of the circle of Willis usually appear by the sixth week. Our patient's aneurysm was saccular with a well-formed neck and a normal parent vessel, the anterior cerebral artery. These facts strongly suggest its congenital origin. Despite the juxtaposition of vascular and optic anomalies in our patient, we cannot support or reject the possibility that both resulted from a single teratogenic event affecting cell differentiation in the first weeks of gestation.

References
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