Pupillary sparing in oculomotor palsy from internal carotid aneurysm

Case report

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The authors report a patient with a carotid-posterior communicating artery aneurysm; although the oculomotor palsy accompanying such an aneurysm is almost universally recognized as being complete, with pupillary involvement, this patient had pupillary sparing in the absence of subarachnoid bleeding. A few similar cases have appeared in the literature. The mechanism of pupillary sparing appears to be based on the position of the parasympathetic pupilloconstrictor fibers within the subarachnoid portion of the third nerve and on the anatomic relationship between the third nerve and the junction of the carotid and posterior communicating arteries.

KEY WORDS: intracranial aneurysm • oculomotor palsy • pupillary sparing

The clinical presentation of oculomotor nerve palsy associated with an aneurysm of the internal carotid artery has been recognized since late in the 19th century. The third nerve palsy is most commonly produced by a ruptured or nonruptured aneurysm at the junction of the internal carotid artery and the posterior communicating artery, a condition not characterized by pupillary sparing. Several authors have stressed that the weakness must involve the pupilloconstrictor fibers of the oculomotor nerve. Indeed, Walsh and Hoyt emphasized that an isolated oculomotor palsy with pupillary sparing will not usually occur as a result of intracranial aneurysm and stated that angiography should not be performed in such cases.

We are reporting the case of a patient with an oculomotor palsy and sparing of the pupil; he was subsequently found to have an internal carotid-posterior communicating aneurysm. We propose that this condition, although infrequent, can occur, and recommend angiography in cases where aneurysm appears likely.

Case Report

A 54-year-old man developed a sudden, severe, right-sided, retro-orbital headache followed 3 days later by a drooping of the right eyelid and diplopia. He was admitted to the Neurosurgical Service of the North Carolina Baptist Hospital 7 days after the initial onset of pain. On physical examination, he demonstrated a marked ptosis of the right
Eye and extraocular muscle palsies of the third nerve (Fig. 1 upper). Both pupils were equal, normal, and reactive (Fig. 1 lower). The cerebrospinal fluid showed no evidence of blood; a 5-hour glucose tolerance test was normal. Cerebral arteriography revealed a right internal carotid-posterior communicating artery aneurysm that measured 5 mm in diameter, with a neck 4 mm long. The fundus extended posteriorly, inferiorly, and laterally (Fig. 2). A right frontotemporal craniotomy allowed excellent visualization of the aneurysm which had compressed the medial aspect of the third nerve; no localized hemorrhage was found. The aneurysm was clipped at the neck.

The operation was well tolerated and the patient was discharged on the 12th postoperative day. Within 2 months he had returned to work full time; by the end of 5 months, the third nerve palsy had completely cleared with no evidence of aberrant regeneration.

Discussion

In the presence of classical subarachnoid hemorrhage, the third nerve palsy lends itself to a fairly simple diagnostic exercise. However, when third nerve palsy occurs as an isolated lesion in an otherwise healthy individual, we are faced with a greater spectrum of clinical possibilities.

The exact frequency with which oculomotor nerve palsy occurs with an unruptured internal carotid-posterior communicating artery aneurysm is not known. In a highly detailed description of signs and symptoms of intracranial aneurysm, Walker cited a frequency range of 35% to 65% in all cases of unruptured supraclinoid aneurysms.
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Pool and Potts\textsuperscript{17} reported an over-all frequency of 78\% from supraclinoid and intracavernous, and ruptured as well as unruptured aneurysms. Considering the reverse of the problem, Rucker,\textsuperscript{18} in an analysis of 335 isolated third nerve palsies, found that aneurysms of the circle of Willis were the most common cause with an over-all frequency of 20\%. Jefferson\textsuperscript{14} had previously made a similar qualitative observation.

The existence of an aneurysm in any location is most often heralded by subarachnoid hemorrhage. However, the importance of an oculomotor palsy as a first manifestation was illustrated by Okawara.\textsuperscript{26} In his study on the warning signs preceding aneurysmal rupture, he found that extraocular muscle impairment was second in frequency to localized headache when the etiology was based on expansion of the aneurysm. However, when it occurs, the third nerve palsy may be an ominous sign of impending aneurysmal rupture. Extraocular muscle impairment preceded frank subarachnoid hemorrhage by an average of 29.6 days.

Various authors, in discussing ocular findings associated with carotid posterior communicating artery aneurysms, have reported a small number of oculomotor palsies with pupillary sparing.\textsuperscript{2,4,10,19,21} One may wonder whether pupillary sparing had occurred in other series of cases where ocular findings were not as well documented.

The oculomotor nerve, in its subarachnoid portion, consists of a group of large caliber (10 to 16 \( \mu \)) somatic fibers that innervate the extraocular muscles and the levator palpebrae superioris, and a group of smaller caliber (1 to 5 \( \mu \)) parasympathetic fibers that are destined for the ciliary ganglion.\textsuperscript{15,19} Some investigators have also found sensory fibers that mediate the stretch reflexes in the extraocular muscles.\textsuperscript{21,23}

The location of the parasympathetic pupilloconstrictor fibers within the third nerve has been clarified through the investigations of Sunderland and Hughes\textsuperscript{19} and Kerr and Hollowell.\textsuperscript{15} Sunderland and Hughes, working with cadavers, found that the small parasympathetic fibers were concentrated at the periphery of the superior aspect of the nerve just beneath the epineurium. Kerr and Hollowell, working primarily with dogs in experiments that included stimulation and transections of the third nerve, localized the pupillomotor fibers in the superficial dorsomedial arc of the nerve. In addition, by progressively transecting the fibers, beginning medially and moving laterally across the nerve, they found that a small number of pupillomotor fibers sometimes traveled in the lateral portion of the nerve, sufficient in number to maintain pupillary tone following ablation of the more numerous dorsomedially placed fibers. Kerr and Hollowell emphasized that these pupillomotor fibers are particularly vulnerable due to their location, as opposed to previous theories suggesting that these fibers are inherently more sensitive to pressure than other fibers. In cadaver studies of third nerves, we have confirmed the dorso- peripheral location of the small-caliber parasympathetic pupilloconstrictor fibers (Fig. 3).

When seen on arteriograms, most carotid-posterior communicating artery aneurysms are found to be directed posteriorly, inferiorly, and laterally.\textsuperscript{2,9,13} If we now consider the anatomic relationship between the oculomotor nerve in its subarachnoid course and the carotid-posterior communicating artery junction, it becomes apparent that

![Fig. 3. Photomicrograph showing cross section of third nerve from a cadaver. Note smaller fibers concentrated in superior aspect of nerve. Bielschowsky's axon stain, \( \times \) 200.](image-url)
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Fig. 4. Anatomical drawing showing relationship of the third nerve to the carotid artery and almost inevitable impingement of a carotid aneurysm (hatched areas) on the third nerve. (Reproduced by permission from Jefferson G: Isolated oculomotor palsy caused by intracranial aneurysm. Proc R Soc Med 40:419-431, 1947).

many of these aneurysmal sacs will approach and impinge upon the nerve in its dor-somedial aspect (Fig. 4), thus illustrating the particular vulnerability of the pupilloconstrictor fibers in these cases.

Aneurysmal involvement of the third nerve in the absence of frank subarachnoid hemorrhage most commonly occurs because of an acute aneurysmal dilation that may be accompanied by a small localized leakage. The nerve injury may be secondary to the dilation, the extravasation, or both. 2,12

The phenomenon of pupillary sparing may be explained by the location of the parasympathetic fibers. In our case, we postulate the presence of a functional, laterally placed bundle of parasympathetic fibers, as described by Kerr and Hollowell,15 or an impingement of the aneurysm upon the third nerve which did not violate some more dorsally placed fibers. In other cases, the aneurysm may compress the nerve in an area relatively remote from the main concentration of parasympathetic fibers. Jefferson's description of internal carotid-posterior communicating artery aneurysms indicates that they may approach the third nerve inferiorly as well as superiorly. 14

The two other diseases most commonly considered in discussions of isolated third-nerve palsy are diabetes mellitus and ophthalmoplegic migraine. The clinical presentation of either disease may bear a striking resemblance to that of an internal carotid artery aneurysm, except for the more common demonstration of pupillary sparing in diabetes. The over-all incidence of ophthalmoplegia in diabetes is probably below 1%. 21

The pathology of the diabetic oculomotor lesion has been located in the intracavernous portion of the nerve. Dreyfus, et al.,8 demonstrated extensive necrosis of axon cylinders and myelin sheaths of the larger fibers along the central axis of the nerve but not usually in the periphery; Asbury, et al.,1 demonstrated demyelinization in the same distribution. We can thus understand the pupillary sparing so commonly seen in this disease. However, in a study of 22 patients with diabetic ophthalmoplegia, Goldstein and Cogan8 found five cases with pupillary involvement; it therefore appears somewhat inaccurate to attribute a pathognomonic quality to pupillary sparing or involvement.

Ophthalmoplegic migraine has frequently been an area of some controversy. Its rarity is attested to by Friedman, et al.,7 who found only eight patients meeting the criteria of this disease from 5000 cases studied. The diagnosis of ophthalmoplegic migraine should be made only after all other possible
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structural causes have been eliminated. Because the clinical picture is so controversial, Walsh and O'Doherty advocate angiography in these cases.

Conclusion

After examining the underlying pathology and briefly looking at other nonsurgical diseases able to mimic the ophthalmoplegic presentation of an intracranial aneurysm, we urge a less rigid approach in weighing the importance of pupillary sparing. Because of the potential catastrophic consequences inherent in aneurysmal disease, we urge that aggressive diagnostic efforts, including angiography, be undertaken when intracranial aneurysm seems a likely diagnosis. The neurosurgeon who is able to approach a symptomatic intracranial aneurysm in the absence of frank subarachnoid hemorrhage will have a distinct advantage.

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References


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