Case 42

History of Present Illness

A 75-year-old retired administrative assistant with a history of cataracts and previous corneal erosion many years before was doing well until one day she noticed she had a deep throbbing pain over her right eye and that her eye lid seemed droopy. Within 2 weeks she was seeing double. She remembers a severe headache 2 weeks before. She has a history of migraines all of her life, but recently she was getting more headaches. She is worried, since things seem to be progressing—she was sent for urgent evaluation by her ophthalmologist.

Past medical and ocular history Hypertension Left bundle branch block	*Past surgical history* None
Medications Hydrochlorothiazide 50 Lopressor 50 Potassium 20 Aspirin 625	*Family history* Father and mother heart disease and hypertension
Social history Widowed with three children and now retired	*Review of systems* Dizziness

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Examination
Acuity with correction Right eye: 20/50 Left eye: 20/40
Pupils 7.5 mm OD in darkness to 7.0 mm OD light 5.5 mm OS in darkness 4.5 mm OS light Near 7 mm OD and 3 mm OS NO RAPD by reverse technique
Intraocular pressure Right eye: 15 mmHg Left eye: 15 mmHg
External exam 3 mm Ptosis OD
Eye alignment Limited elevation and adduction-3 and depression-2. Good abduction (see Fig. 42.1); she has 25 diopters of right exotropia and eight diopters of hypotropia
Slit lamp examination Conjunctiva 1 + injection OU; 1 + nuclear sclerosis OD, and 1 + posterior subcapsular cataract OD
Visual field Normal
Fundus examination Normal
Neurologic examination Aside from eye findings she was normal

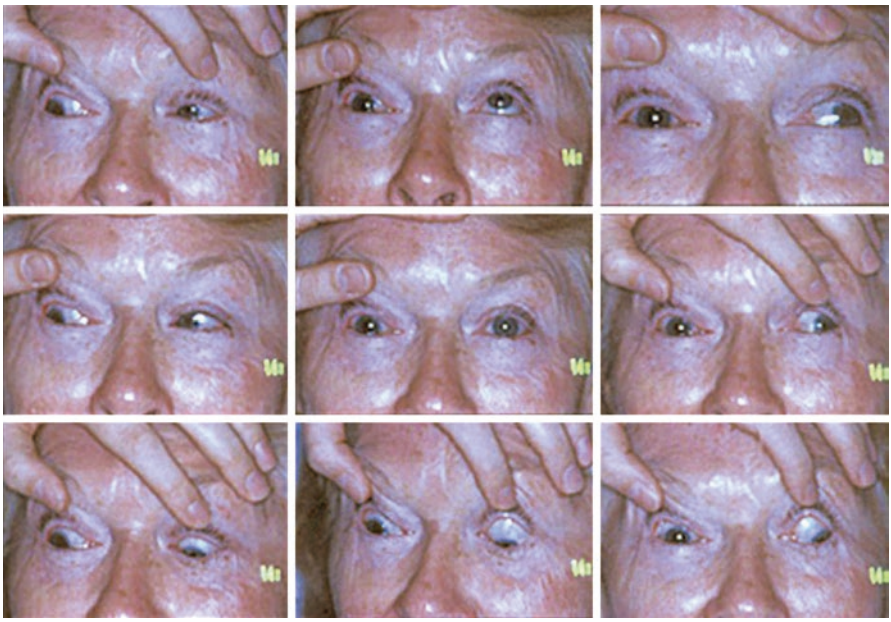

Fig. 42.1 These were her cardinal positions of gaze. Note, that we had to raise her lid to get these photos. The findings were typical for a pupil involving, third nerve palsy

Discussion

Neurologic Perspective—Dr. Digre

When a person over 60 starts having new headaches, diplopia, and ptosis—you have to pay attention! First, she had a previous headache 2 weeks ago, and you have to find out what happened with that. Also careful query about other constitutional symptoms (fatigue, pain with chewing her food)—looking for giant cell arteritis. The ptosis and diplopia could be a symptom of ocular myasthenia, but there is pain—and that is not myasthenia. The new pain with ptosis, a slightly larger pupil on the right, and a pattern of a third nerve palsy is really disturbing.

In evaluation, the first step would be emergent imaging—MR scan and MRA to rule out a mass lesion or aneurysm as well as ESR and CRP (since a painful third could also be giant cell arteritis) (Case 32).

Aneurysms—especially posterior communicating (PComm) aneurysm compressing the third nerve are one of the most feared causes of a new onset headache and a third nerve palsy especially with pupil involvement such as is in this case. Failure to diagnose correctly can lead to a deadly outcome. Sometimes the symptoms are vague and not easy to diagnose, and sometimes the symptoms are sudden onset of a severe headache related to a sub-arachnoid hemorrhage. A sentinel headache, like the headache she had 2 weeks before can be a warning of an aneurysm. When thinking of thunderclap headache (Case 33), it is worth remembering that aneurysm is one of the feared causes along with reversible vasoconstriction syndrome, arterial dissection, and venous thrombosis among others. The sentinel headache can occur without evidence of a bleed. Headaches commonly occur with both ruptured and non-ruptured aneurysm. The ICHD 3beta criteria for headache with saccular aneurysm are in Table 42.1.

Table 42.1 Headache attributed to unruptured saccular aneurysm

Diagnostic criteria:
(A) Any new headache fulfilling criterion C
(B) An unruptured saccular aneurysm has been diagnosed
(C) Evidence of causation demonstrated by at least two of the following:
1. Headache has developed in close temporal relation to other symptoms and/or clinical signs of unruptured saccular aneurysm, or has led to its diagnosis
2. Either or both of the following:
(a) Headache has significantly worsened in parallel with other symptoms or clinical or radiological signs of growth of the saccular aneurysm
(b) Headache has resolved after treatment of the saccular aneurysm
3. Either or both of the following:
(a) Headache has sudden or thunderclap onset
(b) Headache is associated with a painful IIIrd nerve palsy
(D) Not better accounted for by another ICHD-3 diagnosis, and intracranial hemorrhage and reversible cerebral vasoconstriction syndrome have been excluded by appropriate investigations

Headache Classification Committee of the International Headache Society. The International Classification of Headache Disorders: 3rd edition (beta version). *Cephalalgia.* 2013;33:629-808

While aneurysms may lie dormant for years and never rupture, risk factors for rupture of aneurysms include: female sex (check), age (check), hypertension (check) aneurysm size (over 7 mm), and location. The most common symptom of an unruptured aneurysm is headache, sometimes stroke like symptoms, and cranial neuropathy. The most common symptom of a ruptured aneurysm is sudden onset of headache with nausea and photophobia (symptoms of dural irritation), followed by change in mental status and possibly abnormal neurological examination.

Third nerve palsy (TNP) is the most common cranial neuropathy seen with an aneurysm followed by other cranial nerve palsies such as a fourth, sixth, fifth (first and second division). The pain involved in this TNP comes from small first division twigs innervating the third nerve—so eye pain occurs in association with the third nerve compression from aneurysm in over a quarter of the cases.

Some of the causes of misdiagnosis include: diagnosing a primary headache (thunderclap or migraine), diagnosing stroke (if there are neurological deficits), diagnosing meningitis or infection if there is fever, or microvascular cranial neuropathy (Case 18).

Evaluation should proceed with imaging: CT to look for acute blood and CTA for an aneurysm or MR (acutely blood may not show up) and MRA. When an aneurysm is detected, the size is very important, since as the size increases so does the risk of hemorrhage. For example, aneurysms in the anterior circulation less than 6–7 mm are considered incidental and often not treated since the risk of bleeding is so rare. Whereas larger ones or ones found in the posterior circulation have a greater risk of bleeding.

Occasionally, digital subtraction angiography is needed to adequately define the aneurysm before clipping. However, when an aneurysm is *symptomatic* like in our case, it will require emergent treatment—with either clipping or coiling.

What happens to the pain and to the TNP after treatment? Guresir et al found that clipping brought greater resolution to the TNP than coiling (complete improvement 55% compared to 32% and partial improvement 92% vs 74%). Others have found 90% improve no matter what the treatment. The pain generally resolves after treatment, but I have seen post-clipping pain continue—usually behind the eye or on the ipsilateral side of the head. Anti-inflammatories or anticonvulsants such as gabapentin can be very helpful.

Ophthalmic Perspective—Dr. Lee

The rule of thumb is that a TNP is an aneurysm until proven otherwise. Sometimes, as Dr. Digre points out, the TNP can be very subtle. In any patient with ptosis, I look to see if there is anisocoria. If a bigger pupil is on the same side as ptosis, then I am concerned about a TNP. If it is smaller on the same side of ptosis, then I am worried about Horner syndrome (Case 17). If the patient with ptosis does not complain of double vision, I have the patient look up to see if I observe limited elevation and I ask the patient if they see double in upgaze. Generally, patients with diplopia only in upgaze do not notice it, because we do not look up that often in daily life. In many ophthalmic practices, a technician checks eye movements and pupils then dilates

the patient, so it is critical to instruct your staff about this scenario. Lastly, I would not be concerned if the patient has isolated ptosis (without anisocoria or ophthalmoplegia) or has an isolated large pupil. However, if the eye movements are consistent with a partial TNP then it is important to rule out aneurysm.

There was a super interesting and scary paper about underdiagnosis of PComm aneurysms causing TNP. It turns out that patients underwent good CTA or MRA, which showed the aneurysm but the aneurysms were missed. The reason…either a neuroradiology-trained radiologist did not read the film or the request did not include language about a TNP. So, if you are going to get a scan, make sure that you include a history of TNP and evaluate for PComm aneurysm. If the scan is normal, then call the radiologist to make sure they are neuroradiology trained and comfortable ruling an aneurysm out.

Non-ophthalmic/Non-neurologic Perspective

The lesson here is an older person with a new onset headache deserves immediate attention especially if there are any findings of neurological involvement. Besides listening to the history, examining the cranial nerves carefully and looking at the pupil in this case was key. Often, an isolated dilated pupil is NOT an emergency, but any hint of ptosis or diplopia should trigger an emergent CT and/or MR imaging.

Follow-up

She had an emergent CT which was negative, but an MR and MRA and digital subtraction angiogram (Fig. 42.2) showed a posterior communicating artery aneurysm that was clipped by neurosurgery. She did very well and is currently alive in her 90s.

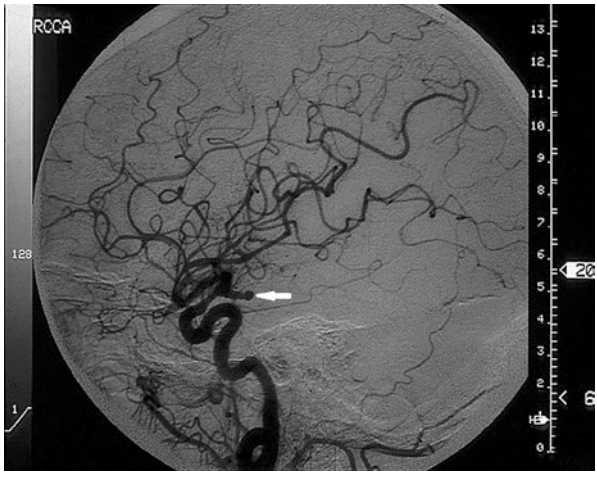

Fig. 42.2 Right internal carotid angiogram shows the PComm aneurysm (*arrow*)

She still has residual right hypertropia which appears to be stable. Her headaches completely resolved. *Final diagnosis: Posterior Communicating aneurysm causing 3NP*.

For Further Study

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