Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/survophthal

Clinical challenges

See no evil, hear no evil…

Sarah Driscoll, MD[a], *Ann P. Murchison, MD, MPH*[b,*], *Jurij R. Bilyk, MD*[b]

[a] *Wills Eye Institute, Philadelphia, Pennsylvania, USA*
[b] *Skull Base Division, Neuro-Ophthalmology Service, Wills Eye Institute, Philadelphia, Pennsylvania, USA*

ARTICLE INFO

Article history:
Received 18 January 2013
Revised 4 March 2013
Accepted 5 March 2013
Available online 1 August 2013
Peter Savino and **Helen Danesh-Meyer,** Editors

(In keeping with the format of a clinical pathologic conference, the abstract and key words appear at the end of the article.)

1. Case report

A 32-year-old black woman with a history of ovarian cancer presented to the emergency room (ER) with progressive worsening of the vision in her right eye over the past 6 months and a right-sided headache. She attributed the change to old glasses and had seen an optometrist 3 months prior to presentation. Refraction failed to improve the vision, and ophthalmologic evaluation was recommended, but not obtained. The patient's mother thought her right eye had looked "swollen" for the past few months.

Does this compilation of symptoms worry you?

2. Comments

2.1. *Comments by Jurij R. Bilyk, MD*

Progressive unilateral vision loss, especially in a 32-year-old, is certainly a matter of concern. The decline over a relatively short period of time could indicate a progressive process with potential to worsen further.

Headache is a ubiquitous complaint, but can be helpful in certain clinical scenarios. Certainly, a new onset headache or change in the character of chronically recurring headaches should not be ignored. It is difficult to know what to make of the mother's report of the right eye swelling without additional information. If a symptom such as this is not apparent to the examining clinician, reviewing old photographs may aid in the detection of any subtle changes in external physical appearance.

In any patient with a positive oncologic history, the concern for recurrent malignancy must remain high in the differential diagnosis. A more detailed cancer history should be obtained, including date of diagnosis, method(s) of treatment, whether the disease is in remission, what surveillance the patient is under to monitor for disease activity, and date of last oncology visit.

3. Case report (continued)

The patient's ovarian cancer was treated surgically with a total abdominal hysterectomy and bilateral salpingo-oophorectomy 10 years previously without adjuvant chemotherapy or radiation. She was notably negative for BRCA gene mutation (BRCA is a tumor suppressor gene and a mutation is

* Corresponding author: Ann P. Murchison, MD, MPH, Wills Eye Institute, 840 Walnut Street, Philadelphia, PA 19107.
E-mail address: amurchison@willseye.org (A.P. Murchison).
0039-6257/$ – see front matter
http://dx.doi.org/10.1016/j.survophthal.2013.03.003

associated with an increased risk for breast and ovarian cancer). She had no other known medical problems and was on no systemic or topical medications. During further discussion, she mentioned decreased hearing in the right ear.

Does this additional information influence your approach to this patient?

4. Comments (continued)

With regard to ovarian cancer the rate of orbital metastasis is exceedingly low. Retrospective reviews totaling 345 patients with orbital metastasis showed only one case (0.3%) of an ovarian primary[6,29]; adjuvant treatments for solid tumors, however, including chemotherapy and radiation, increase the risks for secondary solid and hematologic malignancies (breast, lung, lymphoma), which can certainly metastasize to the orbit.[27] Breast cancer metastatic to the orbit is much more frequent and, although still a possibility in this case, would be of greater concern if the patient had a positive BRCA mutation.

5. Case report (continued)

Visual acuity was 20/400 on the right and 20/30 on the left. A relative afferent pupillary defect (RAPD) was noted in the right eye. She correctly identified 1/8 Ishihara color plates in the right eye and 8/8 in the left eye. Confrontation visual field testing revealed a superotemporal defect in the right eye. She appeared to be exophthalmic on the right with an otherwise quiet appearance, and exophthalmometry confirmed 4 mm of right proptosis. There was questionable increased resistance to retropulsion of the right globe. Ductions and versions were normal in both eyes. She had dysesthesia in the distribution of the frontal (V1) and infraorbital (V2) nerves on the right side. There was right-sided hearing loss by gross examination. Anterior segment examination was unremarkable. Dilated funduscopic examination showed temporal pallor of the right optic disc (Fig. 1). There were no other posterior segment abnormalities.

Does this process localize? What imaging is indicated? What would be on your differential diagnosis?

6. Comments (continued)

The examination confirms the clinical suspicion of a process that is not simply an optic neuropathy. At least three cranial nerves—the optic (II), trigeminal (V), and auditory (VIII)—are involved. The dyad of cranial nerve II and V1/V2 abnormality localizes the process to the orbital apex or cavernous sinus, and possibly more posteriorly. The addition of CN-VIII involvement raises the specter of a more widespread process involving the skull base and almost certainly intracranial structures.

The case thus far illustrates an important point for ophthalmologists: It is critical to ask about and check all cranial nerves when one is found to be abnormal. It is quite common for patients to inadvertently compartmentalize their symptomatology depending on the medical professional they are seeing. In this case, the chief complaint to the eye doctor is visual loss and headache; the hearing loss was elicited only on direct questioning and is a critical piece of information because the differential diagnosis now shifts from a possible intraorbital process to one involving the deeper skull base.

Should this patient be imaged emergently and, if so, what modality would you recommend?

7. Comments (continued)

The presence of temporal disc pallor points to a subacute or chronic process of at least 4 weeks duration. If this were an isolated finding, an argument could be made for an outpatient work-up. In this particular case, the history and examination point to the possibility of an intracranial process, and it remains unclear whether the subacute tempo of visual loss can be correlated with her presumed intracranial disease. An assumption of a slowly progressive intracranial process based on the finding of disc pallor is potentially dangerous. In this case, the patient should be immediately imaged.

The optimal imaging modality of the head in an emergency is a bit of a sticking point. The ubiquity and easy availability of computed tomography (CT) in all hospitals makes this an attractive choice. Certainly, if an acute neurologic process

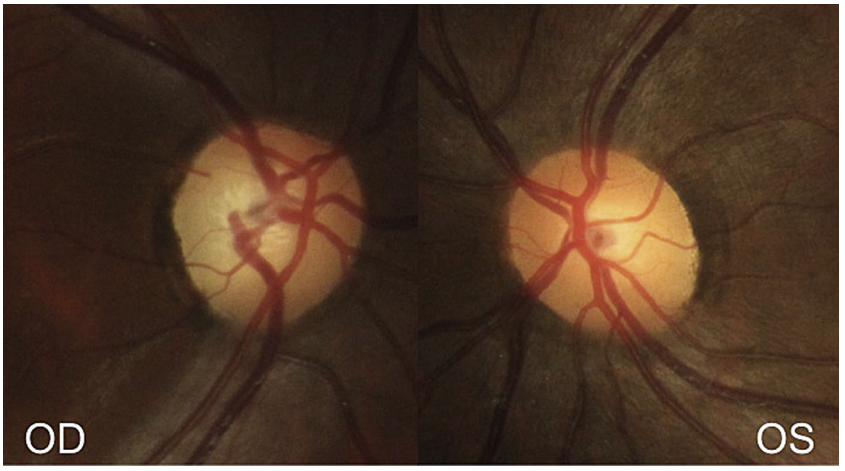

Fig. 1 – Optic disc photos. Note the right temporal disc pallor with corresponding loss of the nerve fiber layer reflex.