

Case Report

Association of giant cell arteritis and pituitary tumor. Report of two cases

CHRISTOS C. PAPAIOANNOU
JAMES C. TRAUTMANN
FRANCIS J. KAZMIER
GENE G. HUNDER

Abstract: Two patients with giant cell arteritis and visual deficits were found to have pituitary tumors, which accounted for the optic findings. The various ophthalmologic abnormalities in these two conditions, which were apparently coincidentally associated in these two patients, are compared. These cases illustrate the importance of careful neuro-ophthalmologic examination and roentgenograms of the head in patients with giant cell arteritis who have visual field loss.

KEY INDEXING TERMS

Giant cell arteritis
Blindness

Cranial arteritis
Polymyalgia rheumatica
Pituitary tumor

Temporal arteritis
Headaches

Ocular signs and symptoms due to ischemia are a common and important manifestation of giant cell arteritis (cranial arteritis, temporal arteritis). Although blindness is the most serious complication of giant cell arteritis, various other ophthalmologic findings may also be seen.¹⁻⁸ When a visual deficit occurs in a patient with this disease, the arteritis is quickly suspected as the cause. We report two patients with giant cell arteritis whose visual deficits were *not* related to arteritis; rather, they appeared to be due to a concomitant pituitary tumor.

Case Reports

Patient No. 1

A 63-year-old white man was seen at the Mayo Clinic on May 14, 1973, with a three-day history of severe bilateral occipital headaches. The patient had noted visual blurring in the left eye on awakening on the day of admission. He had felt fatigued and had lost 9 pounds in the previous month, but there was no nausea or vomiting. Because of lassitude, cough, chills, and slight fever, he had stayed home from work for four days one week before admission.

On examination, he was alert but pale. Blood pressure was 120/85 mm Hg, pulse was 76 beats per minute, and temperature was 96 F (35.6 C). The extracranial, cervical,

From the Departments of Internal Medicine and Ophthalmology, Mayo Clinic, Rochester, Minnesota.

Requests for reprints should be addressed to Gene G. Hunder, M.D., Department of Internal Medicine, Division of Rheumatology, Mayo Clinic, Rochester, Minnesota 55901.

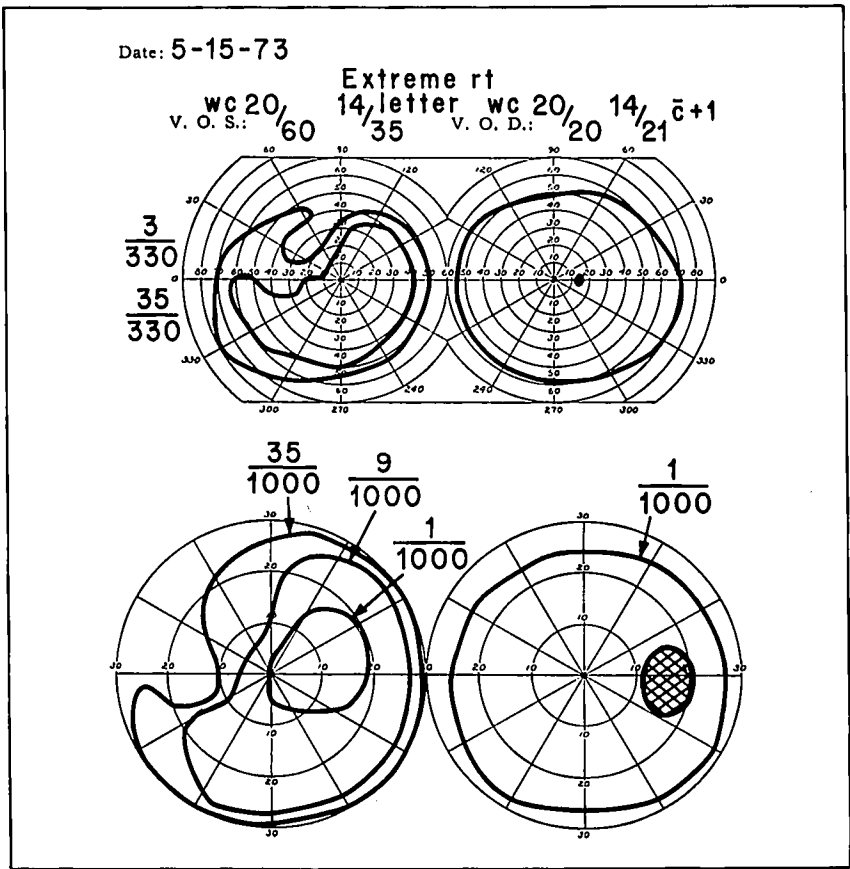


Fig. 1: (Patient No. 1). Visual fields. Normal field of vision, right eye. Optic nerve type field defect, left eye. Top figure shows visual field tested at distance from patient of 1/3 meter. Bottom figure shows central 30° portion of visual field at 1 meter distance.

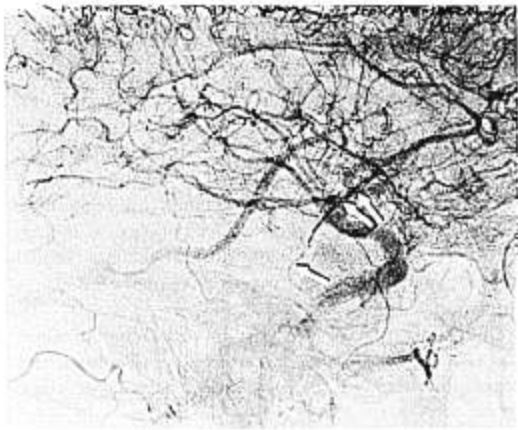


Fig. 2: (Patient No. 1). Portion of left cerebral angiogram. Superficial temporal artery with irregular, narrowed (single arrow) and dilated (pair of arrows) segments.

and peripheral arteries were normal to palpation. There was no scalp tenderness. Visual acuity was reduced to 20/60, 14/35 in the left eye and a visual field defect was present. Results of the vision and field examination of the right eye were normal (Fig. 1). The extraocular movements and optic fundi were normal except for minimal arteriolar sclerosis. The remainder of the examination was unremarkable. The erythrocyte sedimentation rate (Westergren) was 111 mm in one hour, hemoglobin concentration was 11.9 $\mu\text{g/dl}$, and α_2 -globulin was 0.98 $\mu\text{g/dl}$.

A clinical diagnosis of giant cell arteritis was made. Prednisolone (Hydeltrasol®), 100 mg intramuscularly, was given immediately and cortisone acetate, 200 mg, was started intramuscularly twice daily. The patient's headaches were relieved within eight hours. However, roentgenograms of the head disclosed enlargement of the sella turcica and this was confirmed by lateral tomograms of the sella. On further questioning, the patient admitted to progressive loss of libido and body hair over the

previous three to four years and impotence for five months. The breast areolae were depigmented, and axillary hair was absent. There was no galactorrhea. Pituitary gonadotropins were 0 rat unit per 24 hours (normal male, 3 to 20 units/24 hr), serum testosterone was less than 10 ng/dl (normal male, 300 to 1,200 ng/dl). Levels of total serum thyroxine and thyroid-stimulating hormone were normal.

On May 21, 1973, a retrograde left brachial arteriogram was obtained which showed a vascular pituitary tumor and also pronounced irregularity of the left superficial temporal artery consistent with temporal arteritis⁹ (Fig. 2). Left temporal artery biopsy taken in an arteriographically abnormal area showed giant cell arteritis. On May 24, 1973, a chromophobe pituitary adenoma was removed trans-sphenoidally. Ten days later, the patient was dismissed on a regimen of 40 mg of prednisone daily, intramuscular long-acting testosterone every three weeks, and 0.2 mg of *l*-thyroxine daily. On a follow-up visit on Feb 15, 1975, the visual fields were normal on perimetry. The visual acuity was normal. The erythrocyte sedimentation rate was 23 mm in one hour.

Patient No. 2

A 69-year-old white woman experienced left-sided scalp tenderness, ear pain, throbbing headaches, and night sweats in August 1974. She was started on triamcinolone (Aristocort®, Kenacort®), 4 mg four times daily, which promptly relieved all symptoms. The dosage of triamcinolone was gradually reduced and use of the drug was discontinued in October 1974. Two weeks later, the patient noticed bilateral blurring of vision. A right visual field defect in the right eye was noted. In the first week of November 1974, a left temporal artery biopsy was done which showed giant cell arteritis. Therapy with triamcinolone was resumed, in a dosage of 4 mg twice daily; there was no improvement in vision. Subsequent re-examination revealed a visual defect in the left eye, in addition to the previously noted right field defect, in spite of continued triamcinolone treatment.

Past pertinent medical problems included arterial hypertension and arteriosclerotic cardiovascular disease with stable angina pectoris for 15 years. In 1973, a right radical mastectomy had been performed for adenocarcinoma of the breast.

On Feb 10, 1975, she was seen at the Mayo Clinic. Blood pressure was 168/70 mm Hg, pulse was 72 beats per minute, and temperature was 97.6 F (36.4 C). Her cheeks were puffy as a result of the triamcinolone treatment. The temporal and occipital arteries were normal to palpation except for the localized area of the previous biopsy. The pupils and extraocular movements were normal; visual acuity was 20/60 OD and 20/25 OS. The fundi showed minimal pallor of both optic discs. An old healed chorioretinitis scar was noted in the temporal retina of the right eye. Intraocular tensions were normal. An aortic systolic ejection murmur (2/6) was heard. Bruits were audible over the carotid and femoral arteries. The

erythrocyte sedimentation rate and hemoglobin concentration were normal.

The following day, perimetry showed a bitemporal hemianopsia, right greater than left, suggesting a chiasmal lesion (Fig. 3). Skull roentgenograms revealed an enlarged sella turcica, and tomograms suggested an intrasellar tumor with retrosellar and left parasellar extension. Levels of serum thyroxine, follicle-stimulating hormone, luteinizing hormone, and 24-hour urinary gonadotropins were normal. There was no galactorrhea nor evidence of adrenocorticotrophic hormone (ACTH) deficiency. On Feb 20, 1975, a carotid arteriogram was obtained which showed a faintly vascular intrasellar tumor with suprasellar extension. On Feb 24, 1975, trans-sphenoidal removal of a chromophobe pituitary adenoma was performed. On March 3, 1975, the patient was dismissed on a regimen of triamcinolone, 4 mg twice daily. On follow-up visits there was pronounced improvement in the right visual acuity (OU 20/25), and the hemianopsia had significantly lessened on perimetric examination. There was no evidence of a recurrence of the breast cancer.

Discussion

Since the recognition of ocular involvement in giant cell arteritis by Horton and Magath¹⁰ and Jennings,¹¹ loss of vision has come to be considered the most common serious manifestation of the disease.¹⁻⁶ Ischemia of the optic nerve and the retina leading to loss of vision is usually secondary to inflammatory involvement of the posterior ciliary and proximal ophthalmic arteries.¹² The arteritis usually causes an abrupt, partial, or complete visual loss in one eye which may go on to affect the other eye within days or weeks. Impairment of less than one-half of one or both visual fields is uncommon.^{3,4} The most frequent ophthalmoscopic appearance that accompanies visual impairment is ischemic optic neuritis with its characteristic findings of pale papilledema, a few splinter hemorrhages around the disc, and occasional cotton-wool exudates.

Involvement of the intracranial arteries posterior to the optic nerve, causing ischemia of the visual pathways, is rare, but cortical blindness and visual hallucinations have been described.^{13,14} Crompton¹⁵ found minimal arteritic changes in the region of the optic chiasm and pituitary stalk in a meticulous autopsy of a patient with giant cell arteritis, and Walsh and Hoyt¹⁶ write of a patient with

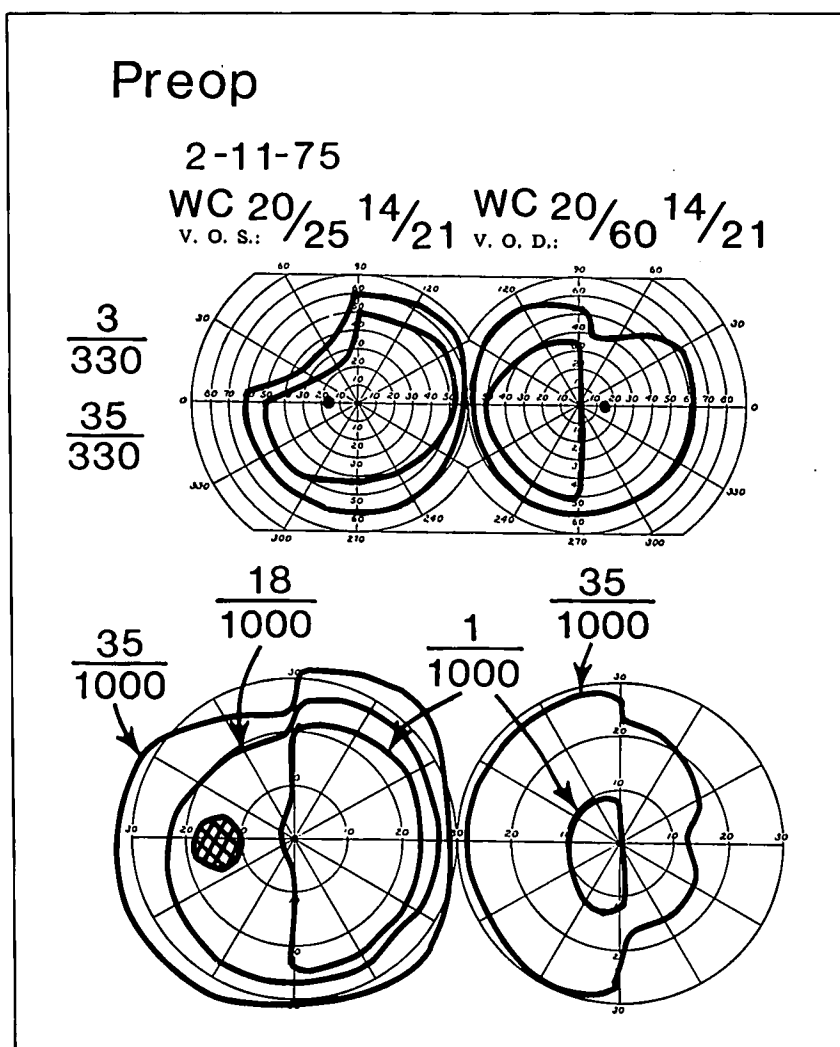


Fig. 3: (Patient No. 2). Visual fields demonstrate bitemporal hemianopsia, indicating a chiasmal lesion. Test orientation the same as for Fig. 1.

bitemporal hemianopsia and symptoms of giant cell arteritis.

Chromophobe adenomas, as were found in these two patients, are by far the most common pituitary tumor.^{17,18} Symptoms and signs of these tumors usually begin insidiously and include orbital headaches, visual disturbance and visual field loss, and hypopituitarism or hyperpituitarism.¹⁷⁻²⁰ Diagnosis may be aided by the evaluation of pituitary hormones.

The visual field defects often commence in

the upper temporal quadrant. Optic atrophy evolves slowly, and the visual acuity often remains normal in the uninvolved nasal half-fields. Among 1,000 patients with pituitary tumors evaluated at the Mayo Clinic between 1935 and 1962, approximately 70% had visual field defects and 61% of these were bilateral.²⁰ In a smaller series,²¹ bitemporal hemiscental scotomas were more common than the classic complete bitemporal hemianopsia. Currently, with the earlier diagnosis and the treatment of smaller secretory pituitary adenomas, the

incidence of visual field defects associated with pituitary adenomas is much less. Of the last 364 patients with confirmed pituitary adenomas who had surgical treatment via the trans-sphenoidal approach at the Mayo Clinic since 1972, only 99 (27%) patients had visual field defects.²²

In spite of the improvement in techniques for early diagnosis of pituitary adenomas, delay in diagnosis, in terms of years from the time of initial symptoms, continues to be noted.^{20,23} Hooper and associates²³ found neurologic complaints in 100% of their patients (85% visual disorders, 72% headaches) and evidence of hypopituitarism in 81% at the time of diagnosis of nonfunctional pituitary adenoma. Peripheral temporal defects were noted in all cases, but it is noteworthy that in three of their patients direct confrontation failed to reveal defects detected on perimetry; this is somewhat reminiscent of the left eye in our second patient.

It is apparent, therefore, that visual symptoms and signs alone are often not specific for either pituitary adenoma or giant cell arteritis. However, the gradual onset of a limited, upper-temporal, unilateral, or bilateral visual field defect without papilledema, hemorrhages, or exudates is more consistent with a pituitary tumor, whereas ischemic optic neuritis is more characteristic of giant cell arteritis. The beneficial effect of corticosteroids on failing vision as observed in these cases does not favor a diagnosis of either disease, because, although uncommon in pituitary adenoma, this effect has been described in both conditions.^{24,25}

The present two cases illustrate the importance of a careful neuro-ophthalmologic examination and roentgenograms of the head in patients with giant cell arteritis who have visual field loss. The initial findings of fever, headache, and visual symptoms in the first patient, and headache, scalp tenderness, and visual deficit in the second patient fit well with a clinical diagnosis of giant cell arteritis. However, when atypical findings were pursued in these two patients, it was found that

the visual loss was related to a coincidentally present pituitary tumor rather than to the arteritis as initially thought. The arteriogram was helpful in supporting the diagnosis of giant cell arteritis in the first case and in choosing a segment of temporal artery to biopsy.⁹

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