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Metastatic paraganglioma presenting as ajunctional scotoma

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ABSTRACT

Purpose: To report a unique case of metastatic paraganglioma presenting as a junctional scotoma.
Observations: A 38-year-old Caucasian man with a history of abdominal paraganglioma presented with minimally blurred vision 20/25 visual acuity in the left eye. The patient was found to have a junctional scotoma upon visual field testing. Cranial MRI revealed a large peri-clival mass compressing the pre-chiasmal optic nerves and other loci of metastatic disease. Intracranial masses, including metastases, can present with a relatively intact central acuity and nonspecific visual symptoms.
Conclusions and Importance: To the best of our knowledge, this is the first report of metastatic paraganglioma causing a junctional scotoma. In cases with junctional scotoma, careful neuro-ophthalmic assessment and imaging are of paramount importance, even in patients with excellent visual acuity.

1. Introduction

A junctional scotoma is visual field defect that arises from damage to the junction of the optic nerve and the optic chiasm. Multiple etiologies may cause junctional scotoma with sellar masses being the most common cause of these visual field defects.[1,2]

We present a case of a 38-year-old man with mild blurred vision in the left eye and junctional scotoma on visual field testing. The identification of a junctional scotoma led to subsequent neuroimaging that revealed bilateral compressive lesions of the pre-chiasmal optic nerves by rare metastatic abdominal paragangliomas.

2. Case presentation

A 38-year-old white man noted painless, blurred vision in the left eye. He had a past medical history of lower back pain and paresthesia in his right ankle and left lower abdominal discomfort six years prior to presentation to the neuro-ophthalmology clinic. A CT scan of the abdomen and pelvis demonstrated a 17 × 12 cm pelvic mass displacing the bladder and causing bilateral hydronephrosis. No fever, sweats, weight loss, or hypertension was reported by the patient.

A biopsy of the mass, six years before his visual complaint, showed metastatic paraganglioma. The metastatic paraganglioma involved the urinary bladder with bony metastases along the spine, ribs, and hip. Urinary levels of vanillylmandelic acid were normal. Tumor embolization was performed with resection of the pelvic mass and placement of bilateral ureteral stents. This was followed by post-surgical radiotherapy and chemotherapy. The patient subsequently failed radiation therapy and began external beam radiation therapy to the neck for palliation of cervical disease. The patient became debilitated and required a wheelchair for mobility. Afterwards, Thalidomide was begun as an experimental chemotherapy.

On ophthalmic examination, the best-corrected visual acuity was 20/20 in the right eye and 20/25 in the left eye. Intraocular pressures were 13 mm Hg and 12 mm Hg in the right and left eye, respectively. Extraocular motility was full in both eyes. No relative afferent pupillary defect was observed. Anterior segment and fundus exams were normal in both eyes, except for mild bitemporal optic nerve head pallor. The pattern deviation of a Humphrey visual field exam demonstrated a superior-temporal defect in the right eye and a central scotoma in the left eye compatible with a junctional scotoma (Fig. 1). The patient's visual complaints had been largely ignored due to preservation of good central visual acuity (20/25 in the worse eye) and vague complaints (slight blurry vision).

An MRI was obtained to evaluate for the possibility of an intracranial mass and revealed a large skull-based mass in the clivus extending

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anteriorly involving both optic canals (Figs. 2 and 3). Given these findings, along with the previous history of metastatic paraganglioma, it was concluded that the mass impinging on the optic nerves was secondary to metastatic disease spread. As the patient had already failed chemotherapy, and with no further chemotherapeutic options, palliative radiation (30 Gy) was performed on the new cranial metastatic lesions. The patient was referred for hospice care after completion of radiotherapy.

3. Discussion

Junctional scotomas are defined as a unilateral central scotoma and a contralateral temporal visual field defect, commonly in the superior-temporal quadrant.[3] Junctional scotomas have been previously thought to be related to the damage of Wilbrand's knee, a loop of decussating fibers that detours into the contralateral optic nerve before entering the optic tract, but this has been challenged by other studies that postulate that Wilbrand's knee is an artifact of monocular enucleation, and Wilbrand fibers form only after an adequate length of time after enucleation.[4,5] Junctional scotomas can be caused by intracranial masses, inflammatory lesions, or vascular lesions with pituitary tumors the most common cause.[6–8] MRI is recommended for any patient with a junctional scotoma. The MRI in our case showed the optic chiasm was fairly clear from compression, thus the visual field defects were presumed to be due to compressive forces against both pre-chiasmal optic nerves near the involved optic canals.

Paragangliomas are rare extra-adrenal neuroendocrine tumors derived from the chromaffin cells commonly located in the abdomen.[9] Intra-abdominal tumors secrete catecholamines more frequently, although this was not observed in our patient. Presenting manifestations of paragangliomas are usually due to mass effects related to the site of presentation but can include secreted catecholamine-associated signs and symptoms include hypertension, anxiety, flushing, sweating, weight loss, and cardiac arrhythmias.[9] Additionally, these tumors can metastasize to many locations. Brain metastasis can produce a myriad of neurologic symptoms, including sensory deficits and motor deficits – which were present in our patient.[10]

In suspected cases of paraganglioma, it is recommended to perform biochemical tests before imaging studies. For catecholamine-secreting paragangliomas, catecholamine metabolites (vanillylmandelic acid,

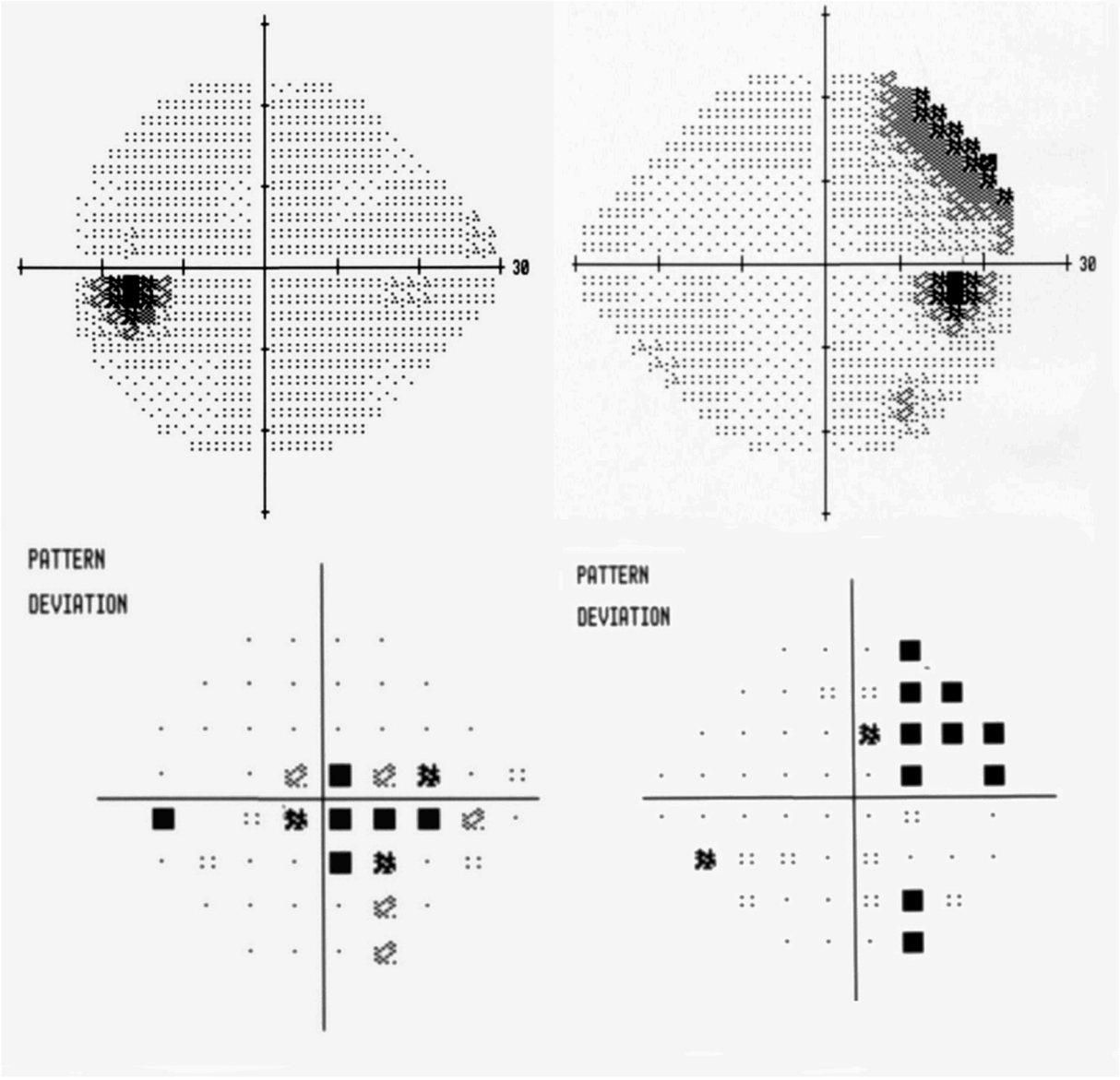

Fig. 1. Humphrey 24-2 Swedish Interactive Testing Algorithm fast visual fields showing a junctional scotoma, superior-temporal defect in the right eye and central scotoma in the left eye.

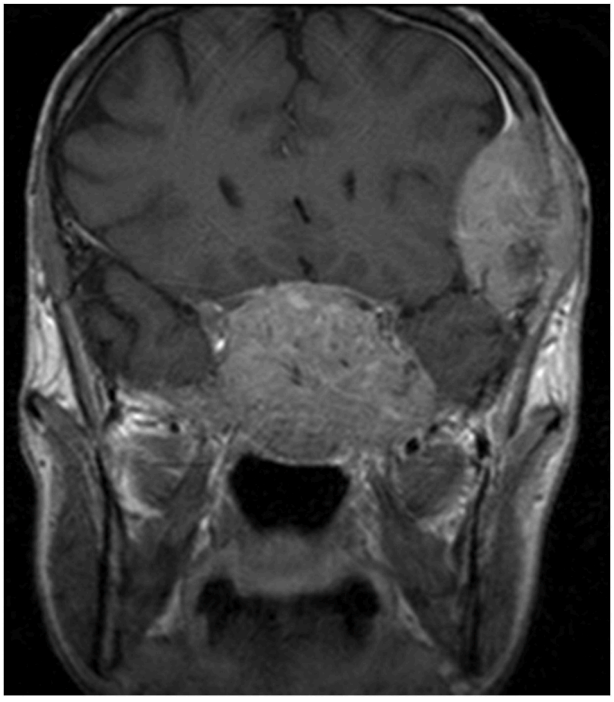

Fig. 2. Coronal post contrast T1-weighted MRI scan showing large skull base mass compressing both prechiasmal optic nerves. Note the left sided skull metastatic lesion as well.

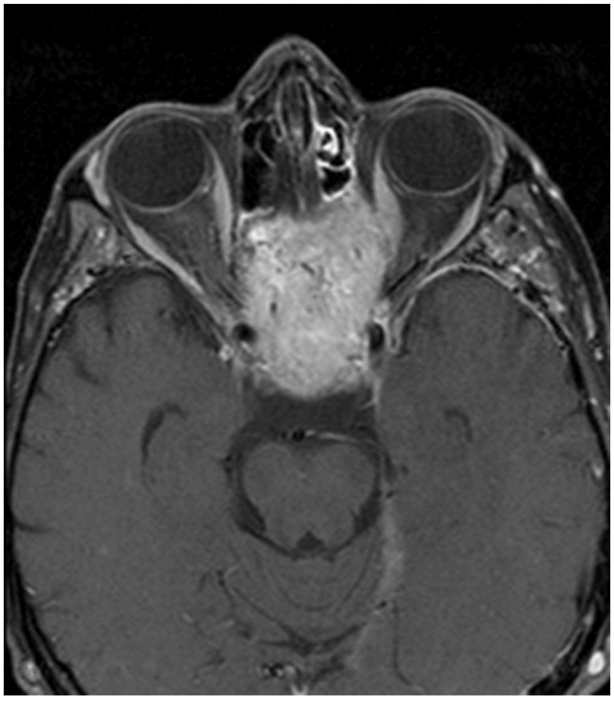

Fig. 3. Axial post contrast T1-weighted MRI revealing a large mass within the ethmoids and skull base with displacement of the left medial orbital wall and obliteraion of both optic canals.

homovanillic acid, epinephrine, and norepinephrine) can be detected in the blood or urine. Imaging techniques including computerized tomography (CT) and MRI have high sensitivity in detecting paraganglioma. Functional imaging, using agents as iodobenzyl hydrazine (MIBG), octreotides, and radionuclides, might be helpful in detecting multifocal and metastatic lesions. Definitive diagnosis of a paraganglioma can be made by identification of the characteristic histology via biopsy. The ocular associations of paragangliomas are uncommon and mostly occur as a result of metastasis to the brain or the orbits.[11]

Treatment of paragangliomas requires a multidisciplinary approach. Patients and family members are recommended to receive genetic testing and counseling. These tumors can be associated with inherited syndromes such as Von Hippel Lindau, multiple endocrine neoplasia type 2, or neurofibromatosis type 1.[9] Surgical resection is still the mainstay of treatment, although stereotactic radiotherapy and systemic tyrosine inhibitors as sunitinib are also options for the treatment.[12]

4. Conclusions

In summary, to our knowledge, this is the first report of a junctional scotoma caused by a metastatic paraganglioma to the skull base. Although the lesion was large by the time of diagnosis, the clinical finding of optic nerve head pallor and junctional scotoma were very subtle underscoring the importance of clinical visual field testing in patients with unexplained visual complaints. Metastatic disease should be considered in patients with visual complaints in association with a known primary paraganglioma even if central visual acuity is intact and emphasizes the utility of visual field testing in mild visual disturbances.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors have no relevant conflicts of interest to disclose with this manuscript.

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