

have well-described abnormalities in the ERG which define these conditions.³ Fenestrated sheen macular dystrophy has a characteristic clinical appearance, as does adult vitelliform macular dystrophy, with raised yellow subretinal circular deposits which leak on fluorescein angiography.^{4,5} Central areolar pigment epithelial dystrophy has a characteristic well-demarcated atrophy of the retinal pigment epithelium. None of these features are present in our case.

The finding of a normal ERG and subnormal EOG occurs in pattern dystrophy of the retinal pigment epithelium, but these are not associated with a bull's eye maculopathy and the fluorescein angiogram shows the characteristic pattern of the specific dystrophy. This was not present in our case.

Dyschromatopsia was not a predominant feature in our case whilst it was common in the series by Deutman¹ and also in that of Copetto and Ayazi.⁶ The defects were predominantly in the tritan axis and were detected using the Farnsworth–Munsell 100 Hue Test. We tested colour vision by the Ishihara test, which has limitations in detecting tritanopic defects. Even so, there was a high degree of variability in findings in the series by Copetto and Ayazi, highlighting the pleomorphic nature of this condition.

In summary, we have presented the case of a 49-year-old patient with normal visual acuity and bilateral bull's eye maculopathy. Fluorescein angiography revealed perifoveal circular dystrophy of the choriocapillaris and retinal pigment epithelium. The ERG was normal and the EOG was slightly subnormal.

We propose that this is a case of concentric annular macular dystrophy, which is a very rare disorder with a highly variable clinical picture and likely autosomal dominant mode of transmission. At present, this disorder is thought to be a dystrophy of the retinal pigment epithelium with a secondary effect on photoreceptor function. Progression may occur but this is an inconsistent finding.

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Sir,

An unusual cause of orbital apex syndrome

Orbital apex syndrome is a clinical entity consisting of a triad of ophthalmoplegia, visual loss and proptosis.¹ It can be caused by a variety of conditions located at the orbital apex such as infection, inflammation or neoplasm. We present the first reported case of orbital apex syndrome caused by a metastatic prostatic cancer to the pituitary gland.

Case report

A 74-year-old man presented to the ophthalmic casualty with a 2 week history of right painless loss of vision. Otherwise, he had no past medical history of note. The best-corrected visual acuity was 6/24 in the right eye and 6/6 in the left. There was a right relative afferent pupillary defect and funduscopy revealed a swollen optic disc. Left fundal examination was normal. The ESR was normal. An initial working diagnosis of right non-arteritic anterior ischaemic optic neuropathy was made.

When the patient was reviewed in the outpatient clinic 2 weeks later there was a marked deterioration in the right vision to counting fingers. In addition he had developed a right proptosis and restriction of eye movements. The measurements with Hertel's exophthalmometer were 24 mm in the right eye and 21 mm in the left. A brain and orbital MRI scan (Fig. 1) showed a mass arising from the pituitary gland extending into the right orbital apex. The patient was referred to the physician for further management.

Haematological and biochemical profiles revealed normochromic normocytic anaemia and a significantly raised prostate-specific antigen (PSA) level. On direct questioning the patient admitted to urinary symptoms consistent with prostatic disease. The rectal examination revealed a large craggy prostate gland. Subsequent bone scan revealed widespread bony metastases including the cranium. The oncologists were confident these lesions represented secondary prostatic malignancy and the patient was treated with systemic steroid, radiotherapy and hormonal manipulation. Following treatment the

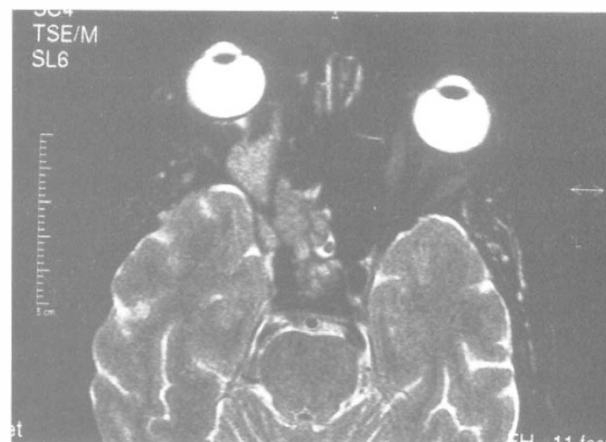


Fig. 1. Brain and orbital MRI scan showing a mass arising from the pituitary gland and extending into the right orbital apex.

proptosis and ocular movements improved but the vision remained counting fingers. He later underwent a transurethral resection of prostate for his prostatic symptoms. Histological examination showed poorly differentiated adenocarcinoma of the prostate.

Comment

Pituitary metastasis with extension into the orbital apex is a rare condition and, to our knowledge, this has not previously been described as a presenting feature of prostatic cancer.

Metastatic tumours of the pituitary gland are uncommon and are usually observed in widespread metastatic disease or at autopsy. Rarely do such tumours represent the onset symptoms of an underlying malignancy, as in our patient. The most common primary cancers causing pituitary metastases are breast and lung.

Radiologically, it is difficult to differentiate metastatic pituitary tumours from primary tumours.² However, the differentiation is important, as the treatment and prognosis are significantly different. Clinically, there are certain signs suggestive of metastatic tumour. Diabetes insipidus is an important sequela of metastatic disease, which is found in up to 20% of patients.^{3,4} This is because the posterior pituitary lobe receives direct blood flow from the systemic circulation and is therefore more commonly involved than the anterior pituitary lobe, which is secondarily supplied by the pituitary portal vasculature. An ophthalmoplegia is unusual with pituitary adenomas as they rarely invade the cavernous sinus. The presence of diabetes insipidus and ophthalmoplegia should suggest a malignancy such as metastatic tumour.⁵

Prostatic carcinoma is common in elderly men and typically metastasises to bone causing osteoblastic lesions. The incidence of pituitary metastases has been reported to be between 1% and 6% amongst patients with prostatic carcinoma, but they are rarely detected in life.^{6,7}

The optic neuropathies can be caused by metastatic prostate cancer to the optic canal. The mechanism is thought to be optic nerve compression resulting from diffuse bony changes. Imaging is useful in showing bone hypertrophy and deformity.⁸ However, this feature was not seen in our patient, suggesting that his optic neuropathy may have resulted from direct tumour compression. Treatment with a combination of steroid and radiotherapy can improve the vision if given early.

Our patient is unusual in many respects: the site of the lesion and the absence of an underlying malignancy at presentation. In retrospect, the rapid progression of the ophthalmoplegia and visual loss should have raised the suspicion of a malignancy. Our case shows that metastatic disease should be included in the differential diagnosis of pituitary lesions.

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Sir,

Orbital apex syndrome as a presenting sign of maxillary sinus carcinoma

Orbital apex syndrome is a rare symptomatological complex characterised by proptosis and paralysis of extraocular muscles, associated with involvement of the first division of the trigeminal nerve and visual loss of various degrees. Only two cases of maxillary sinus neoplasms presenting as orbital apex syndrome have been reported in the literature.^{1,2} We report, to the best of our knowledge, the first case of poorly differentiated squamous cell carcinoma of the maxillary sinus presenting unusually as orbital apex syndrome. Immunohistochemistry was used to confirm the epithelial nature of the tumour cells.

Case report

A 66-year-old man was referred to the eye clinic with an 8 week history of swelling, pain and discharge from his right eye. The patient was a chronic smoker and had no previous history of sinusitis or nasal block. The visual acuities were 6/36 NIP in the right eye and 6/6 unaided in the left eye. Examination of the right eye showed periorbital swelling, non-axial proptosis, ptosis and chemosis (Fig. 1, above). There was an afferent pupillary defect with impairment of colour vision. Corneal hypoesthesia with decreased sensation along the first division of the fifth cranial nerve distribution was noted. The intraocular pressures were normal. Fundus examination showed disc oedema with normal retinal