CASE REPORT

Unilateral proptosis: an unusual presentation of prostatic carcinoma

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SUMMARY

A 68-year-old man presented acutely with periorbital pain and proptosis of the right eye, on a background of generalised pain and weight loss. Imaging showed bilateral signal abnormalities in the basal skull extending into the extraconal orbits with compression of the right optic nerve. His medical history revealed symptoms in keeping with benign prostatic hypertrophy. However, the prostate was irregular on rectal examination and prostate-specific antigen was markedly raised at 1880 ng/dl. A provisional diagnosis of metastatic prostatic carcinoma was made based on the clinical and radiological picture. This was later confirmed to be metastatic adenocarcinoma through means of tissue diagnosis.

BACKGROUND

Unilateral proptosis is uncommon, but can be the primary presentation of sinister pathology such as orbital metastasis. Up to 13% of orbital tumours are metastatic, and orbital involvement occurs in over 2% of cancer patients.¹ For as many as 25% of patients, orbital symptoms and signs such as proptosis, diplopia, and loss of visual acuity can present before a primary carcinoma.¹ Investigation of malignancy in the orbit is an analytical challenge, requiring scrutiny of the broad clinical picture. Imaging of a space-occupying lesion does not provide definitive diagnosis in up to 25% of cases, and anatomy of the orbit renders histological investigation difficult.¹

This case, an uncommon presentation of metastatic prostatic carcinoma, highlights the importance of a comprehensive history and clinical assessment in a man presenting with unilateral proptosis. Prostatic carcinoma accounts for 8.3% of orbital metastasis and is more likely to present prior to a diagnosis of carcinoma.² The prognosis of orbital metastasis is poor, with a median survival of just over a year.³ Nonetheless, accurate identification of the primary tumour can aid provision of tailored therapy for effective palliation and preservation of sight. This is particularly important in prostatic malignancy, 85% of which is responsive to hormonal manipulation.⁴

CASE PRESENTATION

A 68-year-old man presented with a 3-day history of periorbital pain, swelling and proptosis of the right eye. He reported a weight loss of 5–10 kg within 2 weeks, increased hesitancy on micturition and longstanding polyarthralgia. His medical history included prior diagnoses of type 2 diabetes

mellitus, hypertension and benign prostatic hypertrophy. The patient was fully independent and had previously been fit and well.

Assessment of the right eye revealed marked reduction of vision, with an acuity of 1/60 (finger counting at 1 m), loss of colour vision (Ishihara 0/17) and a relative afferent pupillary defect. Proptosis was evident on the right, with chemosis but minimal conjunctival injection. Function of the left optic nerve was intact with a visual acuity of 3/6 and full colour vision (Ishihara 17/17). Intraocular pressure in both eyes was within normal limits. There was no evidence of any extraocular neurological deficit. On examination, the patient was without fever, with no evidence of infection, masses or lymphadenopathy. On rectal examination, the prostate was enlarged and craggy, with distinct hard and irregular lumps at 11 and 1 o'clock.

INVESTIGATIONS

A CT of the head revealed skull base lesions with extension into the soft tissues, a picture consistent with bony metastatic deposits. An MRI scan of the brain further elucidated bilateral areas of expansile signal abnormality centred on the sphenotemporal buttresses and sphenoid triangles. The abnormalities showed heterogeneous gadolinium enhancement. Both lesions extended bilaterally into the lateral extraconal orbits, the anterior aspect of the middle cranial fossae and the temporal fossae. There was bilateral proptosis, more marked on the right, with crowding of the orbital apices and partial effacement of fat surrounding the right optic nerve sheath. The right-sided orbital abnormality extended further within the superior extraconal space and was confluent with a lesion in the sphenotemporal space.

Blood results revealed chronic normocytic normochromic anaemia (haemoglobin concentration 7.7 gm/dl), hyponatraemia and a raised alkaline phosphatase. Serum prostate-specific antigen (PSA) was grossly elevated at 1880 ng/dl. This result, supported by radiological findings and the clinical picture, was highly suggestive of metastatic prostatic carcinoma. Subsequent CT of the chest, abdomen and pelvis showed the presence of widespread bony metastatic disease.

A bone scan confirmed extensive osteoblastic disease in the skull, sternum, ribs, spine, pelvis, femurs, tibiae and humeri. To confirm prostate as the primary source of these metastases, a trans rectal ultrasound scan and 18-core biopsy of the prostate was carried out which demonstrated adenocarcinoma on histological analysis.

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Figure 1 Photo of patient's eyes clearly showing right-sided proptosis.

A CT-guided fine-needle aspiration cytology with immunocytochemistry was used to establish a tissue diagnosis. This confirmed the clinical and radiological suspicion of prostate primary.

DIFFERENTIAL DIAGNOSIS

On initial evaluation of unilateral proptosis, differential diagnoses to consider include thyroid ophthalmopathy, vascular lesions (such as a carotid-cavernous fistula), infection, orbital inflammatory syndrome and trauma. However, the patient was without fever, euthyroid and had no history of trauma.

Histological examination is the most effective way to deduce the primary source of the lesion, with definitive diagnosis from almost 90% of fine-needle biopsy samples.³ However, the tight anatomy of the orbit renders this dangerous and difficult. Adenocarcinoma is the most common cause of orbital metastasis with primary sources of breast, lung and prostate carcinoma and melanoma being the most frequent. Prostatic carcinoma is frequently cited as the third most common cause.¹ Renal cell carcinoma accounts for about 3% of cases and should also be considered in the case of atypical response to treatment for prostatic carcinoma (figures 1–5).⁵

TREATMENT

Treatment of orbital metastases is directed at preserving vision and also palliation of symptoms as overall prognosis is usually poor. In this patient it was felt that orbital debulking would be complex and unlikely to improve clinical outcome. The patient was started on a trial of dexamethasone 8 mg daily, with the

Figure 2 Initial CT section showing soft tissue lesion on posterior wall of the right orbit. Also demonstrates right-sided proptosis.

Figure 3 MRI which more clearly shows the soft tissue mass in the right orbit as well as a smaller lesion on the left.

aim to reduce optic nerve compression. Treatment induced marginal reduction in right-sided proptosis and left optic nerve function remained stable. However, the patient experienced erratic blood glucose levels secondary to the steroids requiring starting of insulin and dexamethasone was weaned down. The patient was deemed suitable for retrobulbar external beam radiotherapy aimed at shrinking the tumour size.

Once the diagnosis of prostatic metastasis was established, the patient was started on the antiandrogen hormonal treatment bicalutamide.

OUTCOME AND FOLLOW-UP

The patient was referred to the pain team for regular review and to provide adequate analgesia for increasing back pain. He was discharged home after an inpatient stay of 15 days, with referral to the community palliative care team. He required close coordination between ophthalmology, oncology and urology for ongoing management. External beam radiotherapy to the orbit achieved good results with a decrease in proptosis, eye discomfort and a mild improvement in visual acuity.

DISCUSSION

In a review of the literature, Lefresne *et al* found 48 published cases (between 1975 and 2012) of prostatic carcinoma metastasis in the orbit and eye. For 38% of cases orbital involvement was the initial presentation of the carcinoma.⁵ The most common symptoms are diplopia, pain and visual loss, and around 63% have signs of proptosis.⁶ In the case of orbital

Figure 4 MRI sagittal section showing lesion in the right orbit.

Figure 5 Nuclear medicine isotope bone scan image confirming extensive osteoblastic disease in the skull, sternum, ribs, spine, pelvis, femurs, tibiae and humeri in what is known as a Superscan.

metastasis proptosis is usually unilateral, with a rapid onset and progressive course.

In elucidation of a prostatic primary neuroimaging has an important role. Ocular metastasis to bone is relatively uncommon occurring in only one-eighth of cases whereas prostatic carcinoma is 4.4 times more likely to metastasise to bone than soft tissue.^{3–7} Radiological findings of bony metastases from a prostatic primary can mimic Paget's disease of the bone. Prostate metastases are usually blastic in nature; however, lytic lesions can occur, resulting in pathological fractures. The sensitivity of CT scan in detecting bone metastasis has been validated as vastly superior to plain radiography.^{8–9} Furthermore, skeletal surveys may have diagnostic value as 70% of cases of prostatic carcinoma will metastasise to bone, but orbital involvement is often late or rare.¹⁰

PSA is a proteolytic enzyme produced in the epithelial cells of the prostate, and can be demonstrated in biopsy samples or other histological specimens using immunohistochemistry. A small amount of serum PSA is present in men with healthy prostates. However, as levels increase, so does the risk of this disease. When the PSA is 1 ng/ml, cancer can be detected in about 8% on biopsy. With a PSA level of 4–10 ng/ml, the likelihood of finding prostate cancer is about 25%; with a level above 10 ng/ml the likelihood is much higher.^{11–12}

Hormonal manipulation of prostatic carcinoma is effective in roughly 85% of cases, with a median time of 18–24 months before the carcinoma develops resistance to treatment.⁴ A high index of suspicion and use of urological history and

investigation in any male presenting with unilateral proptosis is paramount to aid early diagnosis and treatment of a rapidly progressive disease.

Learning points

- ▶ Prostate cancer is a rare cause of unilateral proptosis.
- ▶ Prostatic cancer is the third most common cause of orbital metastasis.
- ▶ Prostate cancer has a strong predilection to metastasise to bone and form osteoblastic lesions.
- ▶ Prostate cancer may undergo a combination of lymphatic, haematogenous or contiguous local spread.

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