Solitary iris plasmacytoma with anterior chamber crystalline deposits

Andrew W. Stacey MD[1,2,5], Alenka Lavric MD[1,2], Caroline Thaung FRCPath DPhil[1,3], Samreen Siddiq FRCPath[4], Mandeep S. Sagoo FRCS (Ed) PhD[1,2,3]

1. Moorfields Eye Hospital, London, UK
2. St. Bartholomew's Hospital, London, UK
3. University College London Institute of Ophthalmology, London, UK
4. Department of Haematology, North Bristol NHS Trust UK
5. Department of Ophthalmology, University of Washington, Seattle, Washington, USA

Corresponding Author
Mandeep S. Sagoo
Moorfields Eye Hospital
162 City Road
EC1V 2PD
London, UK
mandeep.sagoo@moorfields.nhs.uk
+44 0207 253 3411

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ABSTRACT (250 words)

Purpose: To report a case of solitary iris plasmacytoma successfully treated with ruthenium plaque radiotherapy

Methods: A 44-year-old Caucasian female presented with right eye pain and raised intraocular pressure. Her past medical history included breast cancer treated 11 years earlier with lumpectomy, lymph node clearance, chemotherapy, and radiotherapy. On examination, the right iris had a tan coloured mass with crystalline deposits visible on the mass surface and the corneal endothelium. The fundus and left eye were normal. Anterior segment ultrasound scan showed a mass with mixed internal echogenicity and internal blood flow.

Results: An iris biopsy showed an infiltrate of plasma cells. Immunohistochemistry for kappa and lambda light chains demonstrated lambda light chain restriction. Systemic hematologic investigations, including complete blood count, serum paraproteins, bone marrow biopsy, and full body magnetic resonance image were normal. The monoclonal plasma cell infiltrate was consistent with a solitary iris plasmacytoma. The mass was treated with ruthenium plaque radiotherapy. After four years of follow up, the mass remained regressed and no systemic myeloma has developed.

Conclusion: Iris plasmacytoma is rare and should prompt systemic evaluation to rule out multiple myeloma. Solitary iris plasmacytoma can be successfully treated with plaque radiotherapy.

INTRODUCTION

Plasmacytoma is a neoplasm composed of monoclonal plasma cells and can occur as a solitary lesion or as a part of systemic multiple myeloma, a malignant plasma cell neoplasm. Solitary plasmacytomas are divided into solitary extramedullary or solitary bone plasmacytomas. Solitary extramedullary plasmacytomas most often develop in the head and neck, followed by gastrointestinal tract or lymph nodes.[1]

Ocular plasmacytoma is rare. It has been reported in the orbit and eyelid[2], conjunctiva[2], and uvea[2–6]. The majority of these cases, including those with iris plasmacytomas, were found in patients with systemic multiple myeloma. Shakin et al. reported a case with multiple myeloma and infiltration of the iris that simulated a nongranulomatous uveitis.[5] Adkins et al.[2] reported a 67-year-old patient with a history of multiple myeloma with raised intraocular pressure and iris thickening. You et al.[6] reported a case of an anterior uveal tract solitary plasmacytoma diagnosed and treated with iridociliary resection. Herein we describe a case of a solitary iris plasmocytoma diagnosed with an iris biopsy and treated with plaque radiotherapy.

CASE REPORT

A 44-year-old caucasian female presented with right frontal headache and painful right eye. Past medical history included breast cancer treated 11 years prior with lumpectomy, lymph node clearance, chemotherapy (including goserelin, tamoxifen, and adriamycin), radiotherapy, and oophorectomy as the cancer was oestrogen dependant.

On examination, visual acuity was 6/4 in each eye. Her intraocular pressure (IOP) was raised in the right eye (51 mmHg) and normal in the left eye. In the right iris, a tan coloured lesion from 4 to 10 o'clock position was discovered (Figure 1A), with an episcleral sentinel blood vessel in the same quadrant. The lesion demonstrated overlying crystalline deposits which were also dispersed on the endothelial surface of the cornea (Figure 1B). Gonioscopy demonstrated a mass with surface crystals that

obscured the angle from 3 to 7 o'clock (Figure 1B). The left anterior segment was entirely normal. Dilated fundus examination of both eyes was normal. Transpupillary transillumination did not cast an abnormal shadow in the ciliary body of the right eye. Anterior segment ultrasound revealed a temporal and inferior elevated iris lesion that involved the inferior ciliary body. The lesion had mixed internal echogenicity and there was internal blood flow through the lesion with velocity in one vessel 15 cm/s. The lesion was 3.6 mm in elevation and had a base of 10.3 mm (transverse) by 4.2 mm (longitudinal) (Figure 1C).

Topical antihypertensive medications were commenced and a biopsy was taken. The iris biopsy showed an intense infiltrate of plasma cells and small numbers of lymphocytes (Figure 2). The plasma cells were massively expanded by immunoglobulin, with numerous Russell and Dutcher bodies. There was little nuclear atypia. Immunohistochemistry for kappa and lambda demonstrated lambda light chain restriction. The monoclonal plasma cell infiltrate was consistent with iris plasmacytoma. The patient underwent additional hematological investigations, including complete blood count, renal function and serum calcium levels which were all within the normal range. There was no serum monoclonal paraprotein detected and the serum free light chain ratio was normal. A diffusion weighted magnetic resonance image of the whole body showed no lytic lesions or further plasmacytomas and there was no increase in clonal plasma cells on bone marrow biopsy. The urine was positive for Bence Jones protein. This can occur in solitary extramedulary plasmacytoma without indicating multiple myeloma[7].

The diagnosis of solitary extramedullary plasmacytoma of the iris was made. A ruthenium-106 plaque applicator was used to deliver a radiation dose 40 Gy to the apex. This was placed overlying the cornea, centered on the inferior iris mass. Following treatment, the mass thickness regressed to ~1.0mm and the intraocular pressure normalized. After four years of follow-up, the patient remains free from local or systemic recurrence. There has been no residual cornea edema or decompensation. Interestingly, two years after the iris plasmacytoma was treated, she developed an unrelated oligodendroglioma (Grade III) for which she underwent local resection, chemotherapy, and radiotherapy.

DISCUSSION

This case demonstrates a patient who presented with a solitary iris plasmacytoma diagnosed by iris biopsy and successfully treated with plaque radiotherapy. In addition, although this patient was not diagnosed with multiple myeloma, immunoglobulin crystals were found overlying the iris mass and the corneal endothelium. While similar crystal deposits are well recognized in multiple myeloma[8], they have not been described in a solitary intraocular plasmacytoma.

Radiotherapy has been the gold standard treatment for solitary plasmacytoma throughout the body.[1] This is also true in ocular disease. Shields et al. described bilateral solitary extramedullary plasmacytoma of the ciliary body successfully treated with plaque radiotherapy[4] as well as a solitary choroidal plasmacytoma successfully treated with external beam radiotherapy[3]. In this case, low-dose plaque radiotherapy led to a reduction in the iris mass and eventual total regression of the tumor.

The differential diagnosis of pale iris masses includes inflammatory granuloma, metastatic carcinoma, lymphoma, amelanotic melanoma, or as seen in this case a plasmacytoma. A biopsy should be undertaken in cases where plasmacytoma is a plausible diagnosis. If a diagnosis of iris plasmacytoma is made, prompt systemic evaluation should follow. While solitary bone plasmacytomas often progress to multiple myeloma (>75% of the time), solitary extramedullary plasmacytoma as diagnosed in this patient, has been reported to progress in less than 25% of patients.[1] Patients found to have multiple myeloma may require systemic treatment as well as local radiotherapy, emphasizing the need for thorough systemic workup. Long term follow-up, including systemic evaluation, is required as some patients with solitary ocular plasmacytoma can develop multiple myeloma. At 4 year's follow-up, the present case remains in remission for the eye tumour and has not developed any other plasmacytoma or multiple myeloma.

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FIGURES

Figure 1

A: A tan colored mass in the inferior aspect of the right iris. B: Gonioscopy reveals the lesion with crystals overlying the iris lesion as well as on the corneal endothelial surface. The associated angle is closed. C: An ultrasound shows the iris lesion involving the anterior ciliary body prior to radiotherapy with a thickness of 3.6mm.

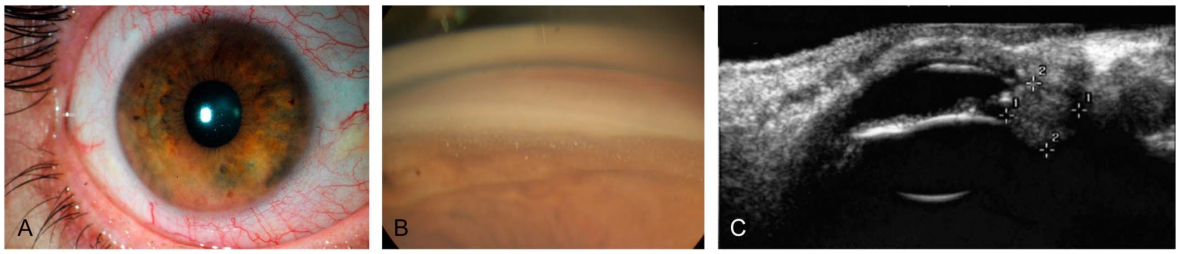

Figure 2

A low-magnification view of the iris biopsy shows plasma cell infiltration with small numbers of lymphocytes.