

# Iris Lymphocytic Lesion Mimicking Amelanotic Melanoma: A Clinicopathologic Case Report.

## Abstract

Lymphoproliferative disorders of the iris are rare, they can be benign or malignant and only few cases have been reported in the literature. We have encountered a 70 year-old male patient who developed an iris mass in his left eye over a period of 1 month. He was referred to our tertiary eye care center with the provisional diagnosis of melanoma. His clinical findings raised the suspicious of amelanotic melanoma because of the high intraocular pressure in the involved eye, the noted vascularization of the mass, and the anterior segment findings. The histopathological diagnosis of a T-cell rich lymphocytic lesion was not suspected. The clinicopathologic findings of this case are described with brief specific review of the literature on the topic of lymphomatous lesions of the iris.

**Abbreviations:** KP'S: Keratic Precipitates; UBM: Ultrasound Biomicroscopy; OCT: Optical Coherence Tomography

## Introduction

The differential diagnosis of an iris mass includes variety of benign and malignant primary lesions such as iris cyst, nevus, juvenile xanthogranuloma, Lisch nodules, inflammatory conditions (such as a granuloma) and several tumors including leiomyoma/leiomyosarcoma, schwannoma and most importantly melanoma [1]. Lymphoproliferative disorders of the iris are rare, they can be benign or malignant and only few cases have been reported in the literature [2-7]. We are presenting our own experience with a 70 year-old male patient who developed an iris mass and was referred to our tertiary eye care center with the provisional diagnosis of amelanotic melanoma. The histopathological findings of this case are described with brief specific review of the literature on the topic of lymphocytic lesions of the iris.

## The Case

A 70 year-old Saudi male patient presented with headache and left eye pain of 1month duration. The patient was seen by a local general practitioner, diagnosed with a left iris mass and referred to us with the presumed diagnosis of amelanotic melanoma for further management. On examination, his best-corrected vision was 20/20 in the right eye and 20/40 in the left eye. The intraocular pressure was 22 mmHg in the right eye and 26 mmHg in the left. Slit lamp examination of the left eye revealed a non-pigmented vascularized raised iris mass infero-nasally, extending from 3 to 6 o'clock, involving the pupillary margin and causing ectropion uvea. (Figure 1A) Keratic precipitates (KP'S) were noticed; however the anterior chamber was deep and quite. (Figure 1B) Gonioscopy showed peripheral anterior synechiae and possible sectorial neovascularization; however the mass did not involve the angle. The lens showed early nuclear sclerosis. Dilated fundus examination of the right eye was within normal limits, while the left eye showed irrelevant macular changes.

## Case Report

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Ultrasound Biomicroscopy (UBM) and Optical Coherence Tomography (OCT) of the left eye confirmed a localized Iris mass of 1.43 mm in thickness and ruled out any ciliary body involvement. (Figure 1C) The patient systemically, was known to have non-insulin dependent diabetes for 3 years on treatment, and he denied any history of trauma, weight loss or close contact with any Tuberculosis patient.

At this point the differential diagnosis included: Amelanotic melanoma as the most likely diagnosis followed by metastasis, and an inflammatory lesion. An excisional biopsy was performed. The specimen consisted of a pigmented piece of tissue measuring 6mm x 3mm. The histologic sections showed that the iris tissue was diffusely infiltrated by mature lymphocytes, the majority of which showed positive staining for CD3, while few only were positive with CD20. (Figure 1D-F) Numerous large pale cells with positive staining for CD 68 were noted. (Figure 1G) Other immunohistochemical and special stains were performed to rule out infectious etiology, melanoma and other malignancies. (Figure 1H) The final diagnosis was benign T- cell rich lymphoid infiltrate of the iris.

## Discussion

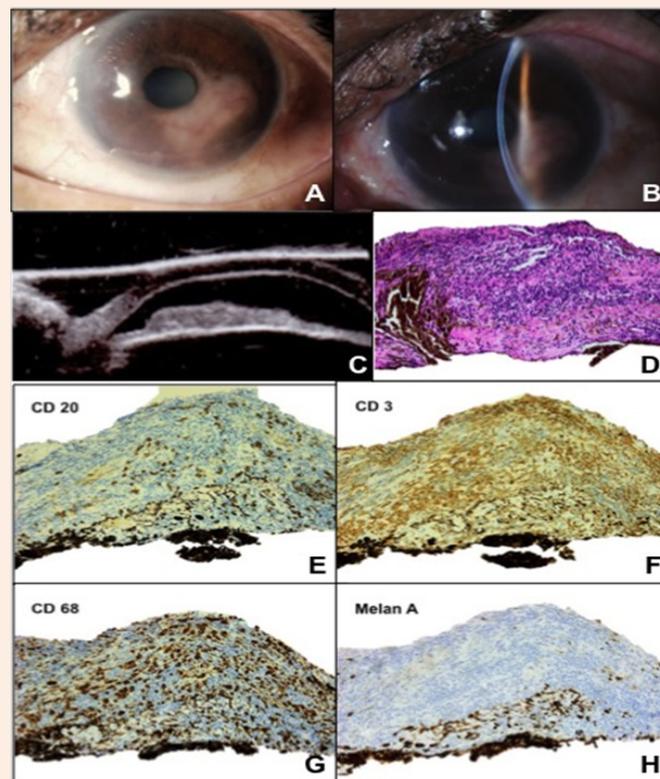
Lymphoma of the iris is uncommon; but unlike choroidal lymphoma it is mostly of a high grade [1-8]. On the other hand, benign lymphocytic Infiltrate of the Iris is extremely rare. Two cases have been reported in the literature. Shields and co-authors published the first case in 1981 [9]. They described a 32 year-old lady who was systemically free, and presented with a dark mass of her right iris slowly increasing in size over a one year period of follow up. Provisional diagnosis was iris melanoma and

accordingly the patient had excisional biopsy. The histopathology showed an iris tissue infiltrated by mature lymphocytes, few plasma cells and clusters of histiocytes. In 2002, Sharma and his group reported a case of a 49 year-old man referred to the oncology service at Wills Eye Institute with an iris mass suspected to be an iris melanoma [10]. Similar to our case, the histopathology revealed a focal, intensely cellular infiltrate of lymphocytes and histiocytes within the iris stroma. Most of the lymphocytes were immunohistochemically reactive for T-cell marker. A lesser number of B-cell lymphocytes were also detected. Melanoma, epithelial cell lineage tumor, infectious etiologies were all excluded.

In 2013, Mashayekhi and his co-authors reviewed 13 patients (14 eyes) who presented with lymphomatous involvement of the iris and were managed at the Ocular Oncology Service at Wills Eye Institute between 1978 and 2010 [2]. Seven out of these 13 patients had previous or concomitant systemic lymphoma. All of these seven patients had received prior chemotherapy before the onset of their ocular symptoms. The most common symptom in his review was blurred vision, followed by eye redness and eye pain [2].

Uveitis with variable degrees of anterior chamber reaction and KPs have been reported as the first presenting manifestations in iris or intraocular lymphoma [2,4,6,7]. Rothova has shown that 48% of all uveitic masquerade syndrome cases had intraocular malignancy, 70% of which was intraocular lymphoma [11]. Diffuse and ill-defined thickening of iris or visible iris tumor was the commonest clinical presentation. Presence of abnormal iris vessels and hyphema were helpful to differentiate lymphoma from anterior uveitis.

Most symptoms associated with primary iris lymphoma (blurred vision, red eye, and eye pain) as well as the clinical findings of secondary eye involvement due to systemic lymphoma like anterior chamber reaction, KPs, posterior synechiae, ciliary injection, and pseudo-hypopyon might overlap with those of idiopathic anterior uveitis. This makes it difficult to make the proper definite diagnosis based on the clinical findings alone. Tissue diagnosis should be considered in middle-aged and elderly patients with atypical and corticosteroid-resistant anterior uveitis [12].



**Figure 1A:** The clinical appearance of the left iris non-pigmented vascularized iris mass.

**Figure 1B:** Slit lamp photo of the left anterior chamber with some keratic precipitates.

**Figure 1C:** UBM showing the left iris mass measuring 1.43mm in thickness.

**Figure 1D:** The histopathological appearance of the iris stroma showing heavy lymphocytic infiltrate (original magnification X25 Hematoxylin & Eosin).

**Figure 1E:** The B-lymphocytes showing positive staining with CD20 (original magnification X25, CD20).

**Figure 1F:** The majority of the cellular infiltrate consists of T-lymphocytes showing positive staining with CD3 (original magnification X25, CD3).

**Figure 1G:** The infiltrate includes few macrophages (original magnification X25, CD68).

**Figure 1H:** The diagnosis of melanoma is totally ruled out by the lesion appearance and the negative IHC stain (original magnification X25, Melan-A).

## Conclusion

This case is presented to alert ophthalmologists regarding the possibility of lymphomatous lesions of the iris, which might not be always malignant. Lymphocytic infiltrate of the iris should be considered in the differential diagnosis of a non-pigmented iris mass and also in masquerade syndromes.

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