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# Primary Orbital Melanoma: A Case Study Highlighting the Critical Importance of Timely Diagnosis and Treatment

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**Abstract** **Case Report**

Primary orbital melanoma (POM) is a rare malignant tumor arising from the deep pigmented layer of the eye, potentially leading to tumor extension and metastases, particularly to the liver. Although there are fewer than 100 documented cases, diagnosis is often delayed, with an uncoded management approach and generally unfavorable prognosis. We present the case of a 57-year-old patient with no significant medical history who developed a pigmented lesion at the inner canthus of the right eye. An excision in 2023 revealed a nodular melanoma, but the patient was already lost to follow-up before the final diagnosis was obtained. In 2024, he presented for exophthalmos with signs of significant tumor extension, necessitating radical treatment including right exenteration, ipsilateral lymphadenectomy, and radio-chemotherapy. Orbital melanomas, which represent about 7% of melanomas, manifest as painless tumors and can be better assessed by MRI. Differential diagnosis includes choroidal angiomas and metastatic tumors. Treatment requires a radical surgical approach, while chemotherapy is often ineffective. Mortality is primarily due to hepatic metastases. At six months post-operation, the outcome was favorable, and the patient received an ocular prosthesis.

**Keywords:** Primary Orbital Melanoma, Exophthalmos, Delayed Diagnosis, Tumor Management, Surgical Intervention, Prognosis, Ocular Prosthesis, Imaging Techniques, Case Study.



## INTRODUCTION

Primary orbital melanoma (POM) is a rare malignant tumor composed of a mass of cells originating from the deep pigmented layer of the eye. This pathology can lead not only to local invasion but also to the extruding of the tumor beyond the eye, causing significant complications for the patient.

Furthermore, orbital melanomas have the potential to metastasize, with a preferential involvement of the liver, complicating the prognosis further.

According to the literature, fewer than 100 cases of POM have been reported, highlighting the rarity of this condition.

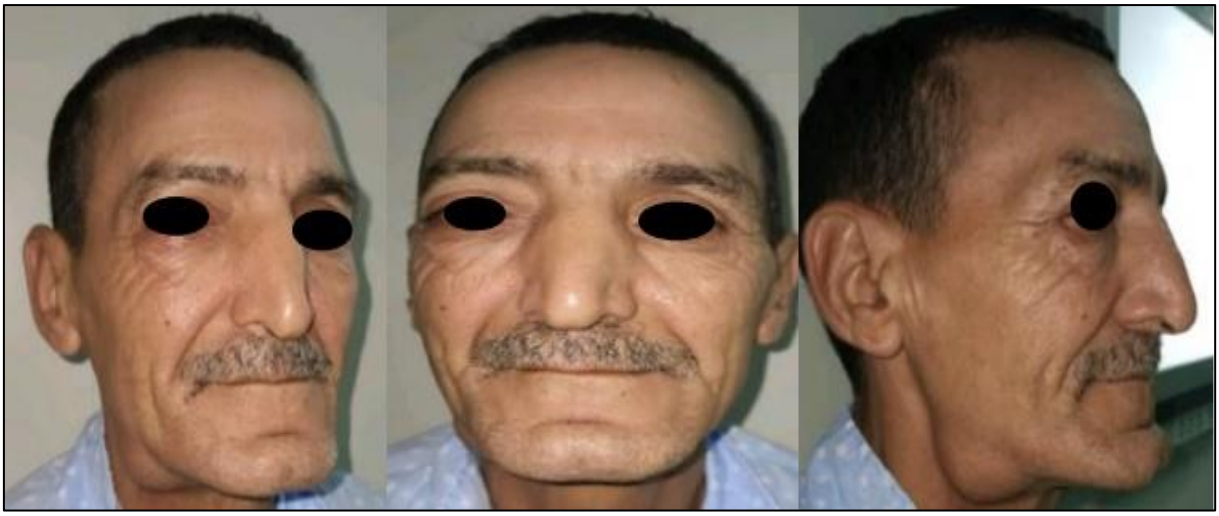
Diagnosis is often delayed due to the insidious clinical presentation of the disease, and management is generally uncoded, resulting in mutilating interventions. Due to these factors, the prognosis for patients with POM is frequently unfavorable.

## CASE REPORT

We present the case of a 57-year-old patient with no significant medical history who exhibited a pigmented exophytic lesion at the inner canthus of the right eye.

In 2023, he underwent an excision, which subsequently indicated a nodular melanoma based on immunohistochemical analysis. Following this procedure, the patient was lost to follow-up.

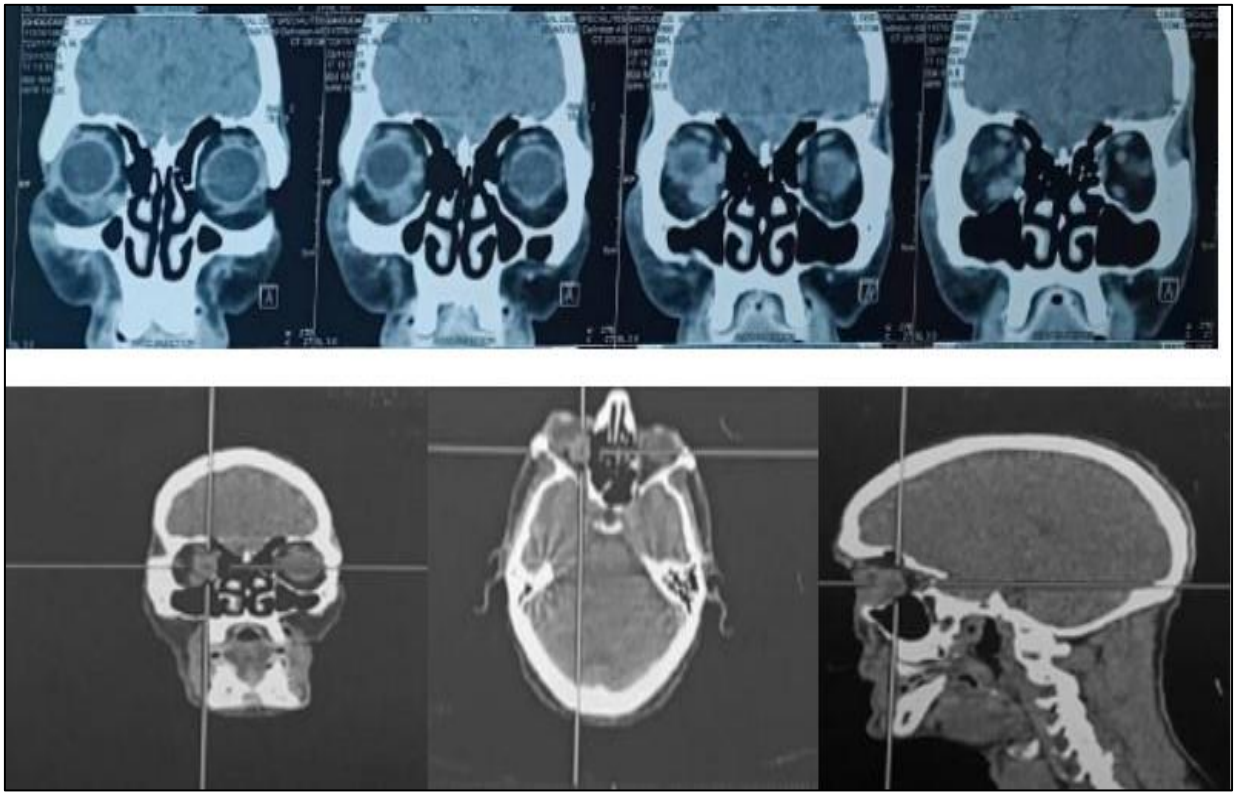
In 2024, he returned for evaluation due to exophthalmos of the right eye, accompanied by swelling of the lower eyelid and a palpable lesion on the lower conjunctiva of the right eye. Cervical examination revealed a mobile, painless lymphadenopathy under the right angle of the mandible, measuring approximately 3 cm in its longest dimension (Figures 1, 2, 3).



**Figures 1, 2 and 3: Pre-operative photos of the patient showing right exophthalmos due to right orbital melanoma**

A computed tomography (CT) scan of the orbit revealed an intra-conal tissue mass on the right side, displacing the optic nerve and infiltrating the eyeball, accompanied by homolateral cervical lymphadenopathy. A CT angiography of the orbit found the intra-orbital process on the right without any other suspicious lesions.

Bone scintigraphy and a PET-CT scan were performed, both showing no evidence of secondary localization and confirming the primary site (Figures 4, 5).



**Figures 4, 5: Axial, coronal, and sagittal CT scans of the facial mass in bone windows showing the right intra-orbital tumor**

The patient underwent right exenteration with ipsilateral lymphadenectomy in our facility, followed by radio-chemotherapy (33 sessions). The postoperative outcome was deemed satisfactory at a six-month follow-up. (Figures 6).

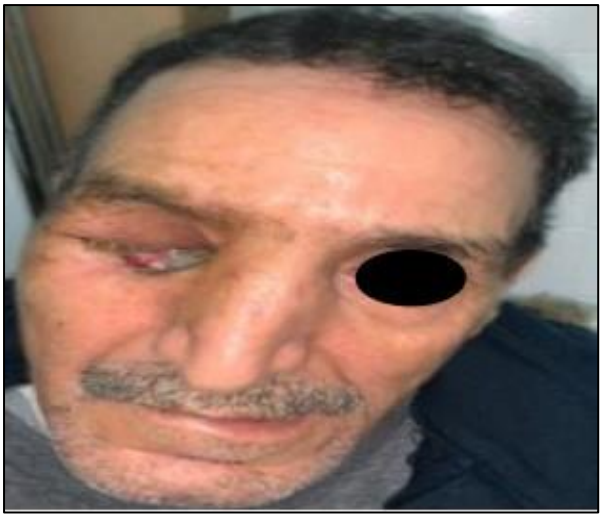


**Figure 6: Post-operative image following right eye exenteration**

The patient subsequently received an ocular prosthesis for aesthetic purposes, which was significant for his psychological well-being.

## DISCUSSION

Orbital melanomas account for approximately 7% of all melanoma cases and are classified among the most malignant tumors, often leading to serious clinical implications [3].

The primary location for these tumors is typically cutaneous, making intra-orbital occurrences relatively rare. In our patient, the initial clinical manifestation of a pigmented lesion at the inner canthus of the right eye emphasizes the critical need for prompt evaluation to prevent potential complications and metastasis.

Clinically, orbital melanomas often present as painless, dark lesions, which may easily be overlooked in differential diagnoses.

Magnetic resonance imaging (MRI) is particularly valuable, being more specific than computed tomography (CT) due to the characteristic hyperintensity in T1-weighted images and hypointensity in T2-weighted images associated with melanin [4, 5]. The application of MRI in our patient's case could have facilitated an earlier diagnosis, allowing differentiation from other conditions such as choroidal angiomas or metastatic tumors from the breast or lung, as well as rhabdomyosarcomas.

Early immunohistochemical confirmation is essential for accurate diagnosis and treatment planning [6, 10].

The prognosis for orbital melanomas is often bleak, with survival rates significantly impacted by the timing of diagnosis and the tumor's inherent aggressiveness.

Studies show that hepatic metastases, which commonly arise due to hematogenous spread linked to the lack of lymphatic drainage in the uvea [6, 11], are a leading cause of mortality.

Delays in diagnosis can lead to more advanced stages of disease, further complicating treatment options and leading to poorer outcomes, as supported by several studies documenting the correlation between early intervention and improved survival rates [9, 12].

Surgical treatment remains the cornerstone of management and must be radical, often involving complete exenteration to ensure comprehensive removal of malignant tissue. In less aggressive cases, achieving clear surgical margins is critical to minimize the risk of local recurrence. In our patient's scenario, the decision for exenteration was necessitated by the reevaluation of his condition, characterized by exophthalmos and aggressive tumor extension.

This underscores the importance of timely surgical intervention in potentially improving patient outcomes. It is noteworthy that chemotherapy typically shows limited efficacy in these cases [7, 13], reinforcing the reliance on surgical and radiotherapeutic methods.

Furthermore, the integration of an ocular prosthesis is crucial in the post-operative setting. Beyond aesthetic restoration, prostheses are vital for psychological support, enhancing the overall well-being of the patient post-surgery [8, 14].

Addressing the psychological impact of disfigurement is a significant component of holistic patient management, and future research should focus on incorporating psychological assessments into treatment protocols.

In summary, this case highlights the aggressive nature of primary orbital melanoma and the urgent need

for vigilance in monitoring atypical ocular lesions. Early diagnosis and intervention are paramount in optimizing patient outcomes, and adopting a multidisciplinary approach that includes psychological support may significantly enhance the quality of life for affected patients. Future research should continue to explore innovative treatment options and strategies to improve early detection and management of this rare but devastating condition.

## CONCLUSION

This case study highlights the critical challenges in diagnosing and managing primary orbital melanoma (POM), a rare yet aggressive tumor. The patient's journey—from the initial pigmented lesion to the later development of exophthalmos—emphasizes the importance of early evaluation and intervention. Delayed diagnosis can lead to significant complications, making timely imaging and immunohistochemical analysis vital for effective management.

Surgical excision remains the cornerstone of treatment, complemented by psychological support through the use of ocular prostheses, which enhance the patient's quality of life post-surgery. This case serves as a reminder of the need for increased awareness and education regarding orbital melanomas, advocating for a multidisciplinary approach to improve patient outcomes and overall well-being.

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