

# An orbital masquerade syndrome: A case report

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## Summary

This report presents the case of a 20-year-old female who presented with features of right panophthalmitis with secondary orbital cellulitis masquerading as an orbital tumour. This presented a diagnostic difficulty to several ophthalmologists. An orbital ultrasound scan revealed an underlying orbital mass, which on histology was discovered to be a well differentiated invasive large cell keratinizing squamous cell carcinoma. This report further emphasizes the value of ultrasound scan in detecting orbital tumours.

**Key-words:** *Masquerade, Orbital, Syndrome.*

## Résumé

Ce rapport présente le cas d'une femme âgée de 20 ans qui s'est présentée avec des symptômes de la panophtalmie du côté droit avec une cellulite orbitaire secondaire qui essaie de se faire passer pour une tumeur orbitaire. Ce phénomène produit une difficulté diagnostique pour plusieurs ophtalmogistes. Echographie orbitaire avait montré une masse orbitaire sous-jacent, dont l'histologie avait démontré d'être une grande Kératinisation cellule kératinisation squameuse cellule carcinome nettement différenciée. Ce rapport attire l'attention de plus sur la valeur d'échographie afin de détecter des tumeurs orbitaires.

## Introduction

The evaluation of neoplastic orbital lesions is an interdisciplinary concept and includes modern imaging techniques as well as rhinosurgical approaches to the orbit.<sup>1</sup> It is expected that orbital tumours, being space occupying lesions, will present with displacement of the eyeball or proptosis. However, several cases of unusual presentations have been reported. Ocular tumours have been shown to masquerade as amaurosis fugax,<sup>2</sup> orbital cellulitis,<sup>3</sup> an apparent subperiosteal abscess,<sup>4</sup> as an orbital apex syndrome,<sup>5</sup> or even as an eyeball in a patient whose eyeball was previously enucleated.<sup>6</sup>

These cases result in diagnostic difficulties and may cause delay in starting appropriate management. However, if modern imaging techniques such as three-dimensional ultrasound imaging,<sup>7,8</sup> computed tomography,<sup>1,6,9</sup> and magnetic resonance imaging,<sup>1,5</sup> are used, a lot of these diagnostic problems can be overcome.

In this report, a case of orbital squamous cell carcinoma masquerading as panophthalmitis with secondary orbital cellulitis is presented.

## Case report

The patient is a 20-year-old female who presented to the eye clinic of the University of Benin Teaching Hospital, Benin-city on 3<sup>rd</sup> of April, 2004. She complained of redness and pain in the right eye of one year duration. There was associated tearing, slight discharge and photophobia. She developed

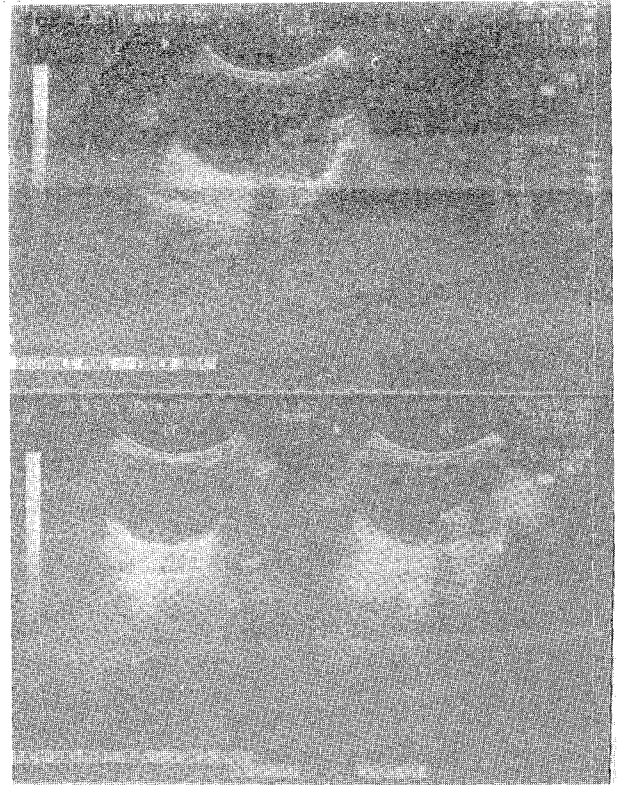


Fig. 1 Orbital scan showing a right sided intra-orbital, extra-ocular mass.

visual loss in the right eye 6 months before presentation. About 4 months before presentation she started having recurrent bouts of fever associated with chills and rigors. A week before presentation she started having severe right sided throbbing headache and started talking irrationally. The patient had received several treatments without success. General examination revealed no obvious abnormality. The visual acuity was no light perception in the right eye and 6/6 in the left eye. There was slight proptosis and ptosis in the right eye, limitation of ocular motility, periorbital swelling, ulceration of the lower lid margin, and purulent conjunctival discharge in the right eye. The conjunctiva was chemosed, the cornea was hazy and there was hypopyon inferiorly. The other structures could not be visualized in the right eye. The left eye was normal. An initial impression of panophthalmitis and secondary orbital cellulitis in the right eye was made. She was started on I.V. ciprofloxacin 200mg 12hourly, I.M gentamicin 80mg 8hrly, I.V metronidazole 500mg 8hrly, gutt okacin 6hrly RE, gutt atropine 1% b.d. RE, oc chloramphenicol nocte RE, tabs cataflam 50mg b.d. and subconjunctival injections of gentamicin daily for 5 days.

There was no significant improvement after 9 days. The cornea had developed a stromal abscess. By the tenth day, there was corneal perforation at the superonasal quadrant. It was decided to eviscerate the eye. An orbital ultrasound scan was done. This revealed an extrinsic indentation of the posterolateral margin of the right globe by a mixed echogenic

## Correspondence

mass measuring 2.3 X 1.4cm (fig 1). The intraocular structures appeared normal. An impression of a right sided intraorbital mass was made. Enucleation of the right eye was then performed with excision biopsy of the mass. Findings at operation were necrosed and friable conjunctiva, a tense eyeball and an inferior intraorbital mass.

The histology revealed a malignant neoplastic lesion composed of clumps of epithelial cells with indistinct cell borders, containing pleomorphic hyperchromatic nuclei with prominent nucleoli. They were invading the loose myxocollagenous stroma in nests and cords. There were heavy infiltrates of chronic inflammatory cells especially plasma cells and macrophages which had ingested melanin pigments. Keratin pearls were present. A diagnosis of a well differentiated invasive large cell keratinizing squamous cell carcinoma was made.

The patient was discharged after 5 weeks on admission and to come to the eye clinic daily for dressing of the right eye. On one of those visits she complained of severe diarrhoea. She was then sent to the accident and emergency unit for management. Unfortunately she died about 24 hours after admission into the emergency unit. Autopsy could not be carried out because no consent was given.

#### Discussion

This patient presented a diagnostic and therapeutic dilemma to all the clinicians that managed her case. She was initially managed as a case of panophthalmitis and secondary orbital cellulitis. Her condition gradually got worse. Because of the unusual appearance of the conjunctiva and the general clinical picture, an orbital ultrasound scan was ordered which revealed the underlying tumour and changed the management plan. She was initially booked for an evisceration, which would have made biopsy impossible. This had to be changed to an enucleation. This report further emphasizes the value of the ultrasound scan in the diagnosis of underlying orbital tumours in suspicious cases.

It has been reported that orbital malignancies can present as acute infections.<sup>3</sup> The tumour itself could become infected or it may stimulate an inflammatory reaction especially when it undergoes necrosis.<sup>3</sup> This may be by the release of tumour necrosis factor (TNF). Neoplastic cells may even cause fever by the release of pyrogenic cytokines including interleukin-1, interleukin-6 and TNF.<sup>10</sup> Furthermore, antigens that elicit an immune response have been demonstrated in many human cancers.<sup>11</sup> These are broadly classified into tumour-specific-antigens, which are present only on tumour cells and tumour-associated antigens, which are present on tumour cells and also on some normal cells. These are able to stimulate both cell-mediated and humoral immunity, which have antitumour activity.

It is possible that there may have been an associated infection in this case. Culture samples from the enucleated globe did not reveal any growth. On the other hand, histology showed that the tumour had invaded the globe. The intraocular involvement could be responsible for the intraocular inflammation, which mimicked features of panophthalmitis while the orbital portion mimicked the features of orbital cellulitis.

The use of the B scan ultrasonography in this case greatly aided the diagnosis of an orbital tumour. The use of ultrasonography in the accurate diagnosis of ocular diseases

has been previously reported.<sup>12-14</sup> Its use is becoming essential in developing countries like Nigeria where three-dimensional ultrasound scan, CT scanning and MRI are not readily available and are very expensive.<sup>14</sup>

Since early suspicion, diagnosis and treatment of intraorbital and intraocular malignancies offer the best chance for survival, ophthalmologists should consider the possibility of an orbital or ocular malignancy when managing unresolving cases of intraorbital or intraocular inflammation. Orbital imaging techniques are of great value in doubtful cases.

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