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The orbits in cancer imaging

V F H Chong

*Department of Diagnostic Radiology, National University Hospital, Yong Loo Lin School of Medicine,
National University of Singapore, Singapore, Republic of Singapore*

*Corresponding address: Dr Vincent Chong, Department of Diagnostic Radiology, National University Hospital,
5 Lower Kent Ridge Road, Singapore 119074, Republic of Singapore. E-mail: dnrcfhv@nus.edu.sg*

Abstract

Primary malignant lesions in the orbit are relatively uncommon. However, the orbits are frequently involved in haematogeneous metastasis or by direct extension from malignancies originating from the adjacent nasal cavity or paranasal sinuses. This paper focuses on the more commonly encountered primary orbital malignancies and the mapping of tumour spread into the orbits.

Keywords: *Orbital metastasis; periorbita; lacrimal tumours; lymphoma.*

Introduction

The approach to the accurate diagnosis and delineation of primary or secondary malignancies requires a working knowledge of orbital anatomy. For descriptive purposes, the orbit has been divided into the following parts: globe, optic nerve and sheath complex, conal–intraconal and extraconal spaces. As each space has unique contents, the diagnostic possibilities can be predicted accordingly.

Primary malignant lesions in the orbit are relatively uncommon. In clinical practice, most cancer imaging studies are carried out to confirm the presence or the absence of orbital involvement from malignancies originating elsewhere. The delineation of disease extent is therefore crucial as the selection of treatment options depends heavily on the extent of malignant involvement.

Normal anatomy

Each orbit has a roof, medial and lateral walls, and an apex. The roof comprises the orbital plate of the frontal bone and most parts of the lesser wing of the sphenoid bone. The frontal sinus is located anteromedially, and anterolaterally there is a deep hollow (lacrimal fossa). At the posterior end is the orbital apex. The medial wall is very thin and the lacrimal groove for the lacrimal

sac is located anteriorly and communicates with the nasal cavity through the nasolacrimal canal. The floor comprises the orbital part of the maxilla, the orbital process of the zygoma and at the posterior end, the orbital process of the palatine bone. Posteriorly the medial and lateral walls are separated by inferior orbital fissure. The medial lip of the fissure is notched by the infraorbital groove which continues anteriorly as the infraorbital canal, transmitting the infraorbital nerve.

The superior orbital fissure and the optic canal form the osseous orbital apex. The optic canal is surrounded entirely by the sphenoid bone. It is bounded by the superior root of the lesser wing of the sphenoid bone superiorly, the optic strut inferolaterally and the body of the sphenoid bone medially. The canal contains the optic nerve and the ophthalmic artery.

Inferolateral to the optic canal, and just separated by the optic strut, is the superior orbital fissure. This fissure is formed by the lesser wing of the sphenoid superiorly and medially; the greater wing of the sphenoid bone laterally; and the orbital process of the palatine bone inferiorly. The superior orbital fissure contains the superior ophthalmic vein, cranial nerves III, IV, VI and the ophthalmic division of the trigeminal nerve. Posterior to the superior orbital fissure is the cavernous sinus. Orbital fat can be seen extending through the superior orbital fissure abutting the anterior aspect of the cavernous sinus.

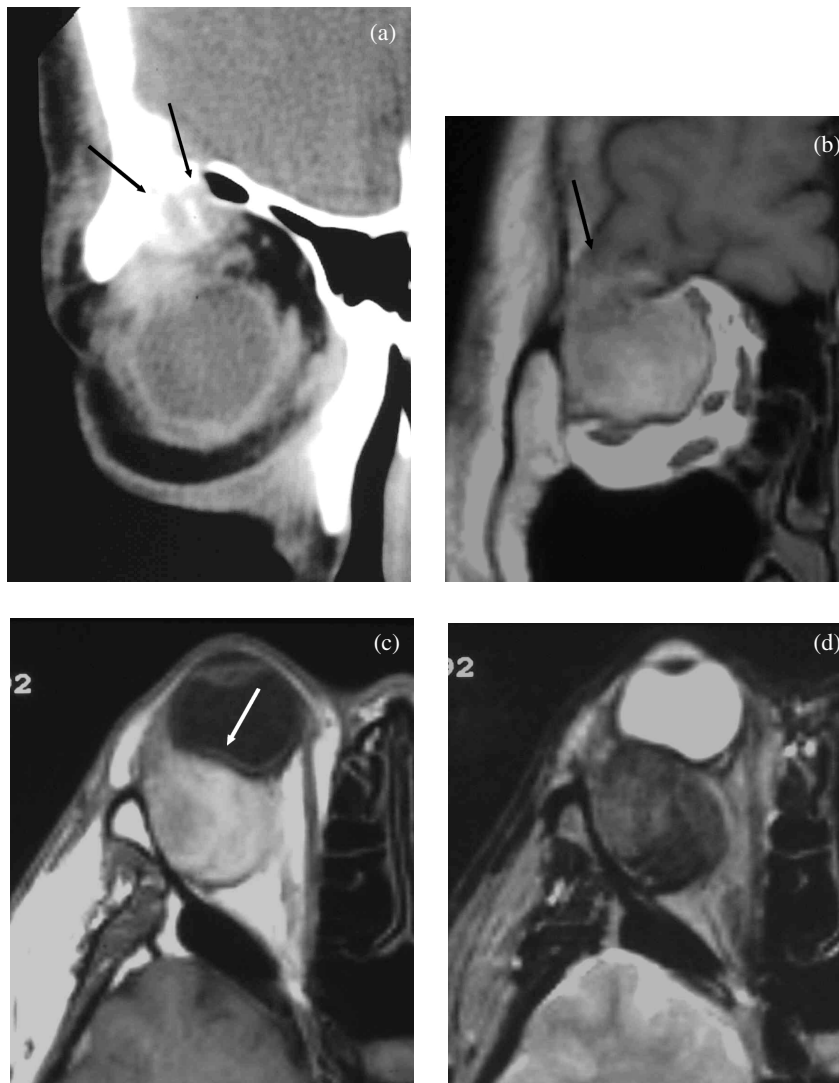


Figure 1 Right lacrimal mucoepidermoid carcinoma. (a) Coronal enhanced CT shows large lacrimal tumour with erosion of superior orbital margin (arrows). (b) Coronal contrast-enhanced MRI shows early tumour invasion into floor of anterior cranial fossa (arrow). (c) Axial contrast enhanced MRI shows enhanced lacrimal tumour indenting right globe (arrow). (d) Axial T2-weighted MRI shows tumour exhibiting low signal intensity.

Lacrimal tumours

In general, epithelial tumours represent 50% of the masses involving the lacrimal gland^[1]. The remaining lesions are due to the lympho-inflammatory group of diseases. Approximately half of all epithelial tumours encountered in the lacrimal gland are benign neoplasms and pleomorphic adenoma is by far the most commonly diagnosed tumour. Adenoid cystic carcinoma and mucoepidermoid carcinoma form most of the malignant lesions originating in the lacrimal gland.

Pleomorphic adenomas are well-defined tumours. They show variable degrees of enhancement on computed tomography (CT) and typically do not cause any bony erosion. On T1-weighted magnetic resonance imaging (MRI), pleomorphic adenomas show intermediate signal intensity and demonstrate medium to high contrast

enhancement. On T2-weighted MRI, they show high signal intensity. Although pleomorphic adenomas are usually classified as benign lesions, they have a propensity to recur. Furthermore, they can undergo malignant transformation especially in patients who have a history of repeated recurrences. It is estimated that such lesions have a 10–15% cumulative risk of malignant transformation (carcinoma ex-pleomorphic adenoma) after 10 years.

The most common malignant lacrimal neoplasm is adenoid cystic carcinoma, followed by mucoepidermoid carcinoma. Squamous cell carcinoma, adenocarcinoma and undifferentiated carcinomas are rare. These lesions typically show an infiltrative pattern of growth. On CT, these lesions typically show bone erosion and contrast enhancement. On MRI, they exhibit strong contrast enhancement. In many patients, the T2-weighted images

show low heterogeneous signal intensity (Fig. 1). It is thought that the low signal intensity is probably related to the high cellularity of these tumours.



Figure 2 Lacrimal and conjunctival lymphoma. Axial contrast-enhanced CT shows tumour involving the left lacrimal gland and the preseptal soft tissues.

Lymphomas

Lymphoid tissues are found in two locations within the orbits, namely, the lacrimal gland and the conjunctiva. It is therefore not surprising that early orbital lymphomas characteristically involve the lacrimal gland and the eyelid (Fig. 2). When lymphomas grow they tend to spread over the surface of the globe and subsequently involve other compartments of the orbit. Lymphomas constitute approximately 10–15% of orbital masses^[2]. Of all patients with orbital lymphomas, 75% have or will have systemic lymphomas. These lesions usually have sharp margins and show intermediate density on CT and moderate degree of contrast enhancement. On MRI, they also tend to enhance moderately and on T2-weighted images show low to intermediate signal intensity. Lymphomas, unlike carcinomas tend not to show bony infiltration until late in the disease process.

Orbital cranial nerves

The ophthalmic nerve (first division of the trigeminal nerve) provides sensory innervation to the forehead. It courses through the superior portion of the orbit and traverses the superior orbital fissure, the cavernous sinus before joining the Gasserian ganglion. Skin malignancies in the forehead may show perineural infiltration of this nerve and spread retrogradely into the intracranial cavity.

The infraorbital nerve (second division of the trigeminal nerve) provides sensory innervation to the cheek. Skin malignancies can similarly spread along this nerve located within the floor of the orbit (infraorbital groove), traverse the pterygopalatine fossa and enter the middle cranial fossa through the foramen rotundum.

The preferred modality for demonstrating perineural spread is MRI. Although CT may be adequate in demonstrating tumour spread within the orbit and pterygopalatine fossa, intracranial spread is more clearly

delineated by MRI^[3]. On MRI, the involved nerves show strong contrast enhancement along the expected course of these nerves. It is worthwhile noting that tumour spread in the pterygopalatine fossa is often more clearly identified on T1-weighted images. The pterygopalatine fossa contains high signal intensity fat, the obliteration of fat strongly suggests tumour involvement.

Optic nerve and sheath complex

Optic nerve meningioma

These tumours are usually seen in middle-aged women. Childhood meningiomas are much more aggressive than in adults. Radiation therapy stabilizes or improves vision in 82% of patients^[4]. Bilateral optic nerve meningiomas may occur in patients with or without neurofibromatosis. However, bilateral optic nerve gliomas are associated with neurofibromatosis type I.

On CT contrast enhanced meningiomas appear as a tubular thickening in two-thirds of cases. Optic nerve meningiomas are also commonly seen as a localised eccentric expansion in the orbital apex. The tramline sign, originally described in meningiomas may also be seen in pseudotumours. On MRI optic meningiomas are seen as uniform or localised enlargement of the optic nerve. These tumours retain the same signal intensity as brain tissue on most pulse sequences. These tumours enhance intensely following contrast injection.

Optic nerve glioma

These tumours are usually seen in childhood, and the average presenting age is 7 years^[5]. Optic nerve glioma in childhood is benign and slow growing (juvenile pilocytic glioma). Bilateral optic nerve gliomas are characteristic of neurofibromatosis. The tumour eventually causes optic nerve atrophy because of pressure effects on the nerve fibres as well as the nutrient arteries. Malignant optic nerve glioma is a rare adult disease with a fatal outcome (glioblastoma multiforme). CT or MRI shows fusiform thickening of the optic nerve. Tumours may show uniform or heterogeneous enhancement. On T1-weighted images, the tumour is isointense with white matter but the T2 signals are more variable.

Haematogeneous metastatic disease

Haematogeneous metastasis to the eye and orbits account for 10% of orbital tumours^[6]. Bilateral orbital metastasis is rare but bilateral ocular metastasis is not uncommon. Metastatic disease, as elsewhere in the body, usually shows non-specific features on CT and MRI (Fig. 3). However, the diagnosis of orbital metastatic disease is usually not difficult as most patients already have a

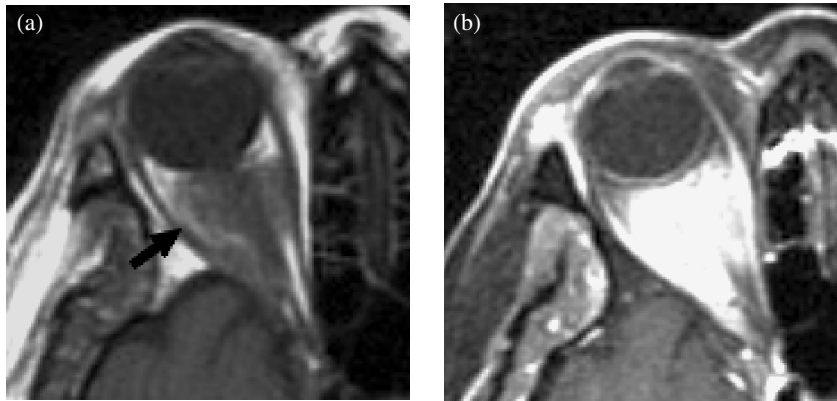


Figure 3 Breast carcinoma with intraconal metastasis. (a) Axial T1-weighted MRI shows ill-defined lesion in right intraconal space (arrow) replacing normal high signal intensity fat. (b) Axial contrast-enhanced MRI shows diffuse intraconal tumour enhancement.

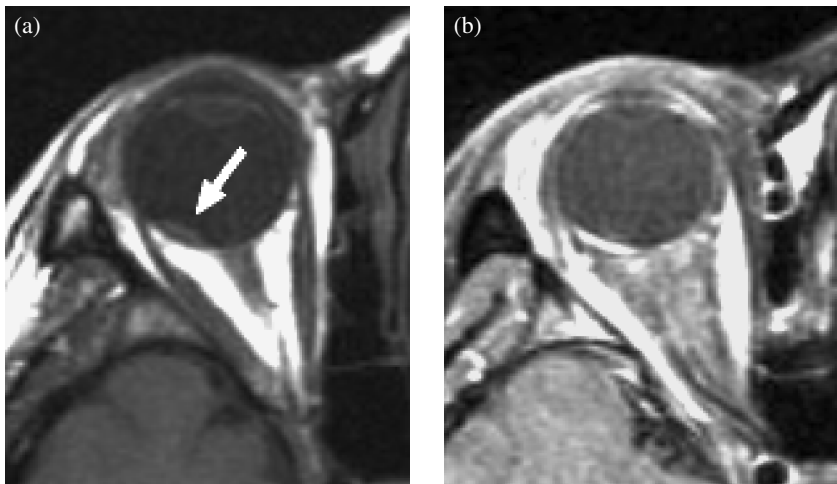


Figure 4 Lung carcinoma with ocular metastasis. (a) Axial T1-weighted MRI shows intermediate signal intensity lesion in right globe (arrow). (b) Axial contrast-enhanced MRI shows focal choroidal enhancement.

history of malignant neoplasm. The common primary sites are lung and breast. Metastatic disease can involve multiple sites. In the orbits, these deposits can be seen in the globe, optic nerve, intraconal, conal and extraconal spaces.

Metastasis in the globe is more commonly seen in adults. Malignant cells gain access to the globe usually through the short posterior ciliary arteries which explains why the majority of lesions are located in the posterior half of the eye. These deposits are most commonly located in the choroid because of the rich vascular network. They are often identifiable on ophthalmoscopy or ocular ultrasound. In the identification of metastatic disease to the eye, MRI is more sensitive than CT because of superior contrast resolution. MRI typically demonstrates strongly enhancing plaque-like lesions in the choroid (Fig. 4). Some lesions may protrude into the vitreous humour. This may or may not be associated with choroidal or retinal detachment.

Malignant orbital infiltration

Most secondary involvement of the orbit originates from malignancies in the nasal cavity or the paranasal sinuses (Fig. 5). Nasoethmoidal or anterior skull base tumours can readily penetrate the lamina papyracea while maxillary sinus tumours can spread superiorly through the floor of the orbit. Anterior nasal cavity neoplasms can extend superiorly through the nasolacrimal duct into the nasolacrimal sac and anterior orbit. This finding has poor prognostic significance and alters surgical planning^[7]. In addition, tumours originating from the nasopharynx or posterior nasal cavity can infiltrate the pterygopalatine fossa and spread to the orbital apex along the inferior orbital fissure^[8]. Malignant neoplasms after gaining access to the orbits can infiltrate the superior orbital fissure to involve the intracranial cavity.

The periosteum of the orbit is called the periorbita. Posteriorly it is fused with the dura in the superior

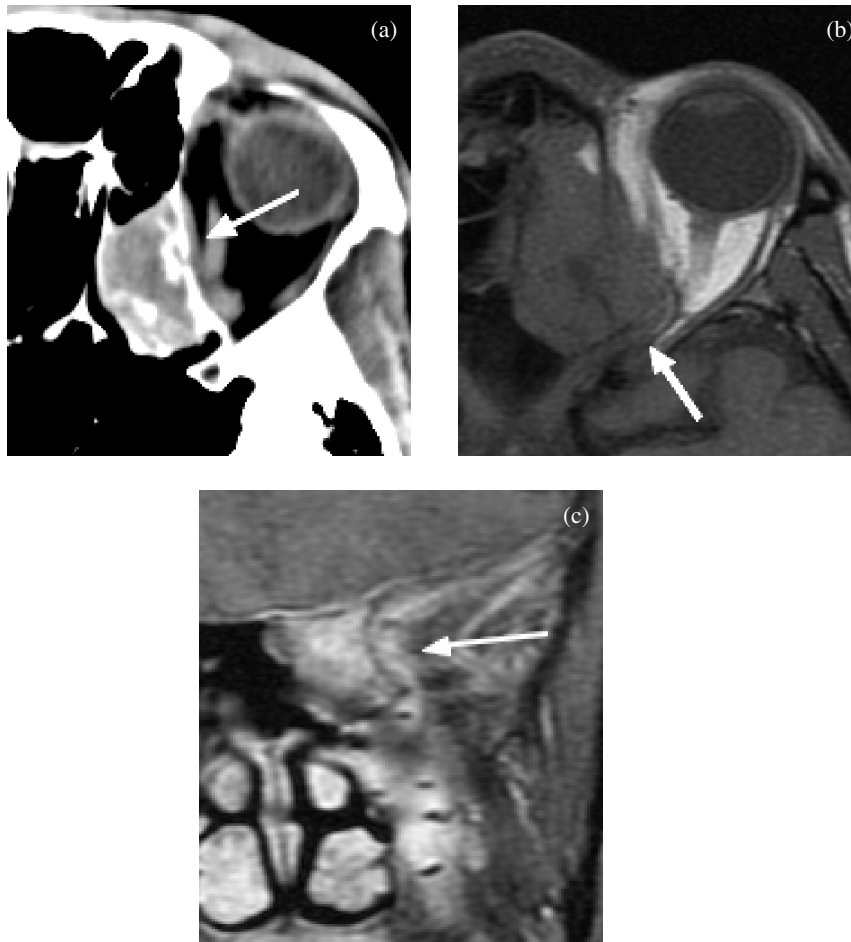


Figure 5 Left ethmoid sinus carcinoma. (a) Axial contrast-enhanced CT shows left ethmoid mass with early orbital extension (arrow). (b) Axial T1-weighted MRI shows very rapidly growing mass invading the orbit and displacing optic nerve (arrow). (c) Coronal contrast-enhanced MRI shows enhanced tumour infiltration in left orbit (arrow).

orbital fissure. It is also continuous with the dura of the optic nerve in the orbital apex and the optic canal. The unique anatomical relationship between the dura and the orbital periosteum has direct clinical implications. First, malignant invasion of the orbital apex that requires bone excision often requires the sacrifice of the optic nerve. This is because the optic nerve cannot be separated from the orbital periosteal lining. Second, meningiomas originating from the middle cranial fossa can extend unimpeded into the orbit, and third, meningiomas can arise in the orbit as a primary tumour.

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