

An Overview of Paediatric Oculoplastics Part III: Orbital

Introduction

Orbital disease in a child poses one of the most challenging problems for an ophthalmologist and is the cause of great anxiety in the patient and family. Identifying whether the pathological processes are confined to the orbital spaces or have originated or extended outside of the orbit is fundamental to the assessment of orbital disease. Congenital craniofacial disorders frequently affect the eye and ocular adnexae, therefore approaches towards paediatric orbital disorders can broadly be divided into two major categories: diseases intrinsic to the orbit and those that are part of craniofacial syndromes. The latter are usually managed with a multidisciplinary approach with the ophthalmic/oculoplastic surgeon as part of the craniofacial team.

Embryology of the orbits

The structural development of the orbit occurs during the first trimester, and is mainly derived from ectodermal neural crest cells. The paired segments of pharyngeal arch one begin to grow early in week four and fusion of the maxillary and mandibular prominences form the bulk of the orbital walls. The normal formation and further development of the optic cup and subsequent migration of the neural crest cells around this structure ultimately give rise to the orbital walls. Mesenchymal neural crest cells from the skull base also contribute to the orbit forming the sphenoid and ethmoid bones. During the third month the orbital bones become differentiated and with time the angle between the two globes begins to decrease (from 105° at three months, to 71° at birth and eventually 68° as an adult).

Evaluation of the orbits

A detailed history from the family or carer is vital. Acquired neoplasias rarely occur in the first six months of life and are more likely to be identified when the child is one-year-old. Congenital lesions are, by definition, present at birth but may be slow growing and only become apparent later in life, such as a dermoid cyst. The duration and speed of growth are important factors in defining the lesion. Generally, lesions that grow slowly are benign and intermittent growth patterns are more likely to suggest neoplasia. Rapid growth patterns can be malignant or benign, such as orbital cellulitis, and associated pain implies inflammation, neural invasion or rapid expansion. Any history of birth trauma, prior surgery, evidence of other cutaneous lesions, recent travel or family history (eg. neurofibromatosis) should be sought. Assessment should include a thorough visual assessment and ocular examination, including evaluation of ocular motility. Position of the lids and canthi should be noted as well as degree of exophthalmos, although anatomical variations in orbital rims in craniofacial disorders may give rise to difficulty in measurement. Nasolacrimal outflow abnormalities should also be noted.

Neuroimaging

Magnetic Resonance Imaging (MRI) is superior to Computerised Tomography (CT) in resolving soft tissue

detail while CT provides better bone resolution. CT utilises ionising radiation, but has the advantage that it is a rapid examination, particularly with the advent of spiral CT. This may reduce the need for sedation which may otherwise be necessary in the paediatric age group.

Intrinsic disorders of the orbit

1) Preseptal and orbital cellulitis

Both conditions have a peak incidence in childhood.^{1,2} Preseptal cellulitis is limited to the skin and subcutaneous tissues anterior to the septum, with the septum acting as a barrier to infection spread. Most common predisposing factors include trauma, skin infection and Upper Respiratory Tract Infection, URTI. *Streptococcus* is now the most common pathogen³ along with *Staphylococcus aureus*, *Haemophilus Influenzae* and *S. epidermidis*. Treatment for children under the age of two years and possibly between ages two and five should be as an inpatient with intravenous antibiotics. In mild cases in older children oral antibiotics should provide good control.

Orbital cellulitis results most commonly from spread of chronic sinusitis (75-85% of cases).⁴ Early signs include eyelid erythema and oedema, proptosis, restriction of extraocular movements, pupillary defects and alteration of vision. CT scans will show post-septal soft tissue swelling. Only when the infection progresses to form subperiosteal abscess or orbital abscess is surgical intervention necessary. Posterior spread can lead to cavernous sinus thrombosis which may present with contralateral lid swelling, altered mental status or signs of meningeal irritation. Other complications include subdural abscess, meningitis and even death. All cases should be admitted, treated with intravenous antibiotics and scanned as a matter of urgency.

2) Non-specific orbital inflammation

Idiopathic Orbital Pseudotumour

Up to a sixth of idiopathic orbital pseudotumour occurs in patients under 20 years of age though it has not been reported in children under three years of age. It is classified by the anatomic location: anterior, diffuse, diffuse sclerosing, apical, myositic and lacrimal. Presentation is most frequently with swelling of the lids and conjunctiva, proptosis, motility disorders and/or pain. Different anatomic locations will lead to different presentations.

CT imaging shows an irregular margin adjacent to the focus of inflammation which enhances with contrast. There are no signs of bone destruction or hypertrophy. Systemic inflammatory diseases should be excluded and first line treatment is with systemic steroids.

3) Benign orbital tumours

Tumours of the orbit are rare in children and most are congenital hamartomas or choristomas. Those that develop after birth are nearly always benign.

Dermoid / epidermoid cysts

Epidermoid cysts (epidermal components only) and dermoid cysts (adnexal structures plus epidermal components) make up to 50% of all the benign orbital



Mr Robin Hamilton
MBBS, MRCOphth,

is a Specialist Registrar on the North Thames rotation. He is currently working at Central Middlesex Hospital where he has developed an interest in Oculoplastics, Medical and Vitreoretinal specialities.



Vickie Lee MA,
FRCOphth,

is a Consultant Ophthalmic & Oculoplastic Surgeon at the Central Middlesex Hospital, London. Her special interests include adnexal trauma and paediatric oculoplastics.

Correspondence:

Miss Vickie Lee,
Central Eye Service,
Central Middlesex Hospital,
Acton Lane,
Park Royal,
London,
NW10 7NS,
UK.

Email: vickielee@mac.com

tumours in children and can be divided into superficial and deep varieties. Superficial dermoids often appear in the first year of life as a painless, firm, sometimes mobile mass laterally in the upper lid though this will change if the cyst ruptures into an acute inflammatory lesion. Deep dermoids remain hidden and appear later in life or with slow onset of proptosis and globe dystopia. Imaging is particularly important to confirm diagnosis and to assess the posterior extent of the orbital component. CT scans generally show a well-circumscribed mass with a uniform appearance and only minor indentation. Increased age and size leads to moulding and eventual bony destruction. Clinical, radiological and pathological examination of periocular dermoid cysts show evidence of inflammation from an early age⁵ and the recommended treatment is surgical excision en masse.

Dermolipomas

Dermolipomas are an ectopia of skin and adnexal structures within connective tissue and may be associated with Goldenhars and Treacher Collins syndrome. They often present as an incidental finding as a yellowish-white superotemporal epibulbar mass, which is soft, smooth, mobile and non-inflamed. They need not be excised unless they cause clinical symptoms such as corneal dellen, lid vaulting, foreign body sensation from hairs, or anterior extrusion. Surgical excision should be conservative and aimed only at removal of the symptomatic portion.

Microphthalmos with cyst

This cystic orbital lesion represents the arrested closure of the foetal fissure at approximately four weeks of foetal life. The cystic portion and degree of microphthalmos vary in size. Treatment depends on the size of the cyst – observation, repeated aspiration or excision. Concurrent use of socket expanders can also be used to encourage growth of the palpebral fissure and orbit.

Mucocoele

Mucocoeles are rare in children and are most commonly from the ethmoidal sinuses. CT imaging is invaluable in the diagnosis by showing the extent of the mucocoele. Surgical correction with re-establishment of drainage into the nasal cavity is required to prevent further damage to non-sinus structures.

Orbital teratoma

This is a neoplastic growth of tissue containing all three germ cell layers. They vary in size though most are small well-circumscribed lesions that appear adjacent to the orbit inferiorly. They have a yellowish-white appearance when small, becoming orange-red when larger. Teratomas often grow rapidly and may appear cystic or solid. CT is important in eval-



Child with left preseptal cellulites with incipient abscess forming on lower lid. This resolved without surgical intervention, treated intravenously and followed by oral antibiotics.

uating these features and in showing the extent of the lesion prior to surgical resection.

Orbital vascular lesions

Orbital vascular malformations are classified as no flow, venous flow, and arterial flow lesions based on clinical and imaging criteria.⁶

Capillary haemangioma

Haik et al found that 33% of capillary haemangiomas had superficial involvement, 7% had deep (behind the orbital septum) involvement, and 60% had both. Ninety-five percent present before six months of age.⁷ The decision to treat periocular haemangiomas is dependent on the location and extent of the haemangioma, and the risk of amblyopia. Most capillary haemangiomas resolve spontaneously. Intra-lesional and oral steroids can

be used. Surgical excision is generally reserved for quiescent residual lesions.

Lymphangioma

These present usually in the first five years of life. Deep lesions often present with sudden haemorrhage into the cystic spaces producing sudden onset of proptosis, pain, decreased vision, limited motility and periocular ecchymosis (chocolate cysts). Conservative care should be advised unless significant visual dysfunction, proptosis with exposure, pain, or optic nerve compromise is identified.

Neuronal tumours

Optic nerve glioma

Gliomas are the commonest intrinsic optic nerve lesion in children, accounting for 5.1% of all childhood orbital lesions. More common in females (2:1 ratio) they often present in the



Treacher Collins Syndrome (Courtesy of Mr Richard Collin). Mandibulofacial Dysostosis is a syndrome with clefts 6, 7 & 8 of the Tessier system and involve both bone and soft tissue. The bony deformities manifest as poorly developed inferior and lateral orbital rims with hypoplastic zygomas. Ocular adnexal soft tissue deformities include the anti-mongoloid slant of the palpebral fissures, inferior displacement of the lateral canthi and malar midface soft tissue deformity.

first decade. Approximately 29% have neurofibromatosis type 1 (NF-1). Although up to 15% of NF-1 patients have anterior visual pathway optic nerve gliomas, routine screening by neuro-imaging is not recommended if the patient is asymptomatic. Extensive investigations should be done to assess progression of the lesion before considering surgical excision, radiation or chemotherapy.

Optic nerve sheath meningioma

These benign tumours are less common than gliomas but 9% also have NF-1. They present differently in children with proptosis, restricted ocular motility, a relative afferent papillary defect as well as the classic triad of visual loss, optic atrophy and opticociliary shunt vessels. The natural course is progressive enlargement and invasion of the optic nerve. There is an estimated 42% recurrence rate after treatment with a 20% intracranial extension rate.

4) Malignant orbital tumours

Primary orbital malignancies **Rhabdomyosarcoma**

Rhabdomyosarcoma accounts for nearly 4% of all childhood malignancies. Arising from pluripotential mesenchymal cells, 75% occur within the first decade of life with 22% in the second. Four major categories of histopatho-

logical appearance are used to classify orbital rhabdomyosarcoma:

- embryonal, the most common making up approximately 65%
- alveolar
- pleomorphic
- botryoid.

Many authors suggest a correlation between cell type and prognosis. Botryoid tumours have the best prognosis and alveolar the worst. The prognosis has improved with radiotherapy and chemotherapy with now up to 82% two-year survival. Tumour diagnosis and staging of the disease guides treatment. Other soft tissue sarcomas including alveolar soft tissue sarcoma and mesenchymal chondrosarcoma. They are rare and do not respond to radiotherapy. Surgical excision is the treatment of choice.

Adenoid cystic carcinoma

Typically a primary epithelial lacrimal tumour in adults, several cases have been reported in children. It is the most common epithelial malignancy in the lacrimal gland in children and presents with slowly progressive proptosis, pain and globe displacement. Biopsy and surgical excision is the treatment of choice though 15 year survival rate remains poor at 22%.

Metastatic orbital malignancies

Neuroblastoma

Neuroblastoma is a malignant tumour of primitive neuroblasts with 90% of cases presenting in the first five years of life. Eight percent present with ophthalmic signs and up to 20% will eventually have some ophthalmic involvement. The classic presentation is with sudden onset proptosis, ptosis and periorbital swelling with ecchymosis. Horner's syndrome may be the presenting feature and has a better prognosis.

Other childhood metastatic orbital malignancies include leukaemia (8% in acute myeloid leukaemia) and acute lymphoma (3%), Langerhans cell histiocytosis, Ewing sarcoma and extraocular extension of retinoblastoma.

Orbital disease associated with Craniofacial syndromes

These fall into two main categories:

- **Craniosynostoses**, which result from premature closure of one or more of the cranial sutures. The most common craniosynostoses include Crouzon and Apert syndromes where shallow orbits and secondary proptosis result in severe corneal exposure.
- **Clefting syndromes** are believed to result from either failure of fusion between the facial processes (true clefts) or failure of mesodermal migration (pseudo-clefts).

The regional topographic classification of craniofacial clefts by Tessier⁸ is most widely used and, depending on the location, can manifest as hypertelorism, medial canthal dystopia with nasolacrimal abnormalities, lateral canthal dystopia and eyelid colobomas. Common ophthalmic clefting syndromes include Treacher-Collins, Goldenhar and amniotic band syndromes as well as hemifacial microsomia. The management of children with craniofacial syndromes requires a multidisciplinary approach, tailoring the management for each patient and often involves multi-stage sequential surgery involving both the cranium and the facial skeleton. The ophthalmologist plays a pivotal role in the monitoring of ocular structures, particularly corneal and optic nerve function which may impact on the timing of major intervention.

Conclusion

Orbital disease in children can be very difficult to assess. If the clinical suspicion of disease is high and the examination difficult, early non-invasive imaging should be utilised. Good rapport and communication with child and family should never be underestimated, and a good relationship with the paediatricians is imperative. It is important to remember that orbital disease in children can progress rapidly, and may not only threaten the child's vision but also their life.

Recommended Further Reading
Katowitz JA. Paediatric Oculoplastic Surgery.
Springer-Verlag; New York, USA; 2002.

References

1. Jackson K, Baker SR. Clinical implications of orbital cellulitis. *Laryngoscope* 1986;**96**:568-74.
2. Jackson K, Baker SR. Periorbital cellulitis. *Head Neck Surg* 1987;**9**:227-34.
3. Donahue SP, Schwartz G. Preseptal and orbital cellulitis in childhood. A changing microbiologic spectrum. *Ophthalmology* 1998;**105**:1902-5.
4. Schramm VL Jr, Curtin HD, Kennerdell JS. Evaluation of orbital cellulitis and results of treatment. *Laryngoscope* 1982;**92**:732-8.
5. Abou-Rayyah Y, Rose GE, Konrad H, Chawla SJ, Moseley IF. Clinical, radiological and pathological examination of periorbital dermoid cysts: evidence of inflammation from an early age. *Eye* 2002;**16**:507-12.
6. Harris GJ. Orbital vascular malformations: a consensus statement on terminology and its clinical implications. Orbital Society. *Am J Ophthalmol* 1999;**127**:453-5.
7. Haik BG, Jakobiec FA, Ellsworth RM, Jones IS. Capillary hemangioma of the lids and orbit: an analysis of the clinical features and therapeutic results in 101 cases. *Ophthalmology* 1979;**86**:760-92.
8. Tessier P. Anatomical classification facial, cranio-facial and latero-facial clefts. *J Maxillofac Surg* 1976;**4**:69-92.