Orbital Dermoids in Children

ABSTRACT Orbital dermoid cysts are benign congenital choristomas. They are common in pediatric population, developing adjacent to suture lines, most commonly located in antero-lateral fronto-zygomatic suture, and are slowly progressive. Complete surgical excision without rupture of cyst is the standard of care. Deep orbital cysts cause proptosis, require imaging, and may present a surgical challenge with a difficult approach. Rupture of the cyst leads to severe inflammatory reaction in surrounding tissues. Overall prognosis remains good with isolated reports of malignancy masquerading as dermoid cysts.

KEYWORDS

INTRODUCTION

Dermoid cysts in children are congenital developmental abnormalities resulting from inclusion of ectodermal elements during closure of the neural tube adjacent to fetal suture lines. They are the most common orbital tumors in children. They are usually found near the lateral canthus at the temporal fronto-zygomatic suture above the superior orbital margin. The second most common site is the superomedial orbital rim. The superficial cyst is small when first seen but may enlarge with age. Deep orbital dermoid cysts usually present later. They are most commonly found in teen or adults and present with proptosis. Inflammation occurs due to cyst leakage or rupture.

INCIDENCE

Dermoid cysts represent 3 to 9% of orbital tumors. The incidence of various orbital tumors in children varies in different studies (Table 1). Orbital dermoids are among the most common benign lesions, along with vascular tumors and inflammatory lesions. In the pediatric population (upto age 18), dermoid cystic lesions are seen in as low as 1.6% (Crawford et al., 1983) to as high as 52% (Shields et al., 1986) of diagnosed orbital tumors. Studies including both clinically diagnosed and biopsied cases showed lesser incidence of cystic lesions as compared to studies including only biopsied cases.

PATHOLOGY

Dermoid cysts are developmental abnormalities or choristomas. The dermoid cyst is lined by cutaneous epithelium that rests on dermal tissues containing skin appendages and may have pedicle attachments to the periorbita. Gross pathological appearance of an excised orbital dermoid shows hair follicles
TABLE 1 Comparison of Incidence of Orbital Dermoid Cysts Among Benign Orbital Tumors in Children

<table>
<thead>
<tr>
<th>Total benign orbital tumors</th>
<th>Iliff &amp; Green, 1978</th>
<th>Crawford et al., 1983</th>
<th>Shields et al., 1986</th>
<th>Rootman et al., 1988</th>
<th>Kodsi et al., 1994</th>
<th>Katowitz et al., 1996</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Dermoid Cysts</td>
<td>174</td>
<td>572</td>
<td>250</td>
<td>241</td>
<td>340</td>
<td>243</td>
</tr>
<tr>
<td>Percentage</td>
<td>29.8%</td>
<td>1.6%</td>
<td>46%</td>
<td>11.2%</td>
<td>19.1%</td>
<td>29.2%</td>
</tr>
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and sebaceous material lumped together. Keratin and lipid debris fill the lumen. Histologically, the cyst wall is lined by keratinizing, stratified squamous epithelium containing hair follicles, sebaceous and sweat glands and keratin. The cyst contents include white sebaceous-like material secreted by the epithelial lining, along with combination of hair, oily material, hemorrhagic fluid and cholestrin crystals.

Isolated case reports (Jones et al., 1935; Wright et al., 1977) show squamous cell carcinoma developing in orbital dermoid cysts. These warrant that a thorough study of the wall lining be done to rule out malignancy.

CLINICAL FEATURES

Dermoid cysts of the orbit are divided into superficial and deep types, with the superficial type presenting early at around 1–3 years of age. They are present at birth and grow slowly. They may be discovered accidentally by parents, during washing of the face of the child. However, a cyst may not be apparent until later in adult life.

Anterior orbital dermoid cysts have consistent anatomic location and physical findings that allows for their identification with a high degree of accuracy. They show adnexal structures as well as epidermal components. They present as a painless, smooth, firm, mobile mass in the upper eyelid anterior to the frontozygomatic suture. They do not displace the eye at the time of the initial presentation. Occasionally the cyst may extend into the orbit and creating a mass effect. Imaging studies are needed to confirm localization.

Deep orbital dermoid cysts may not be detected until teenage or later in adult life. They arise from the orbital bony suture in a similar manner to the anterior dermoid cysts. They present most commonly with proptosis and imaging studies are required for localizing the mass lesion. As the epidermal lining continues its secretory activity, the cyst enlarges and areas of bony erosion may appear and enlarge. The erosion may result in local deformity and may lead to generalized enlargement of the orbit. Occasionally, bony erosion may progress through the lateral orbital wall, with the dermoid taking an hourglass shape. Bone erosion may progress to extend intracranially and involve dural exposure.

Ultrasoundography is helpful for localizing anterior and mid-orbit disease and is a cost-effective screening tool (Char et al., 1982). Computed tomography (CT) has the ability to distinguish both normal and abnormal structures of various tissue densities. This has contributed to very low false positive and false negative rates. CT scanning may show internal calcification. Bony excavation, molding and intracranial extension may be seen on CT scan imaging.

Computerized tomography and magnetic resonance imaging help accurately measure dermoid cyst location, size, extension, differentiate cystic from solid lesions, identify cyst type and help in planning management approach for difficult cases. In a 20-year review of computed tomography of 160 cases of orbital dermoids, Chawda et al. (1999) reported that 69% (111/160) were found at lateral aspect of the orbit, 85% had adjacent bone changes and 14% had calcification.

DIFFERENTIAL DIAGNOSIS

A superficial dermoid cyst can be diagnosed clinically by the typical presentation of a non-tender, mobile mass at the outer orbital rim in a young child. However, an orbital mass with inflammation may be the presenting sign of a ruptured dermoid. Orbital cellulitis, abscess parasitic cysts and pseudotumor may present with similar orbital signs and need to be differentiated from orbital dermoids. There are reported cases of congenital frontotemporal dermoid cyst presenting as a cutaneous fistula (Scolozzi et al., 2005; Honig et al., 1998). Occasionally, these dermoid cysts may have intracranial extension (Niederhagen et al., 1998). Orbital imaging using computerized tomographic (CT) scanning (Song et al., 1990) and magnetic resonance imaging (MRI) (Song et al., 1994) help differentiate cases of orbital tumors with inflammation, including

R. Ahuja and N. F. Azar
TABLE 2 Differential Diagnosis of Orbital Dermoid Cyst

A. Orbital cysts:
1. Dermoid or epidermoid tumor
2. Pseudotumor
3. Mucocele
4. Lymphoma
5. Aneurysmal bone cyst
6. Lipodermoid
7. Epithelial lacrimal gland tumor
8. Dentigerous cyst
9. Teratomatous cyst
B. Inflammatory parasitic cysts
1. Echinococcal cyst
2. Cysticercosis
C. Noncystic orbital lesions with cystic component
1. Adenoid cystic carcinoma
2. Rhabdomyosarcoma
3. Lymphangioma
4. Squamous cell carcinoma
5. Orbital Abscess
6. Meningocele
7. Others

pseudotumor (Shields et al). CT scanning may show a soft tissue mass with a round to ovoid shape, with possible bony fossa formation, bony expansion and sometimes bone sclerosis. MRI can help delineate the full extent of the mass and may be quite useful for deep orbital cysts which present as difficult surgical cases (Song et al., 1994). It is important to remember that malignant tumors may masquerade as orbital dermoid cysts. Table 2 lists the various differential diagnoses for orbital dermoid cysts.

SURGICAL TREATMENT

Complete surgical excision without rupture is the treatment of choice and standard of care. Classically, general anesthesia is required for these surgeries. The anterior cysts can be approached by a curved incision to the skin, parallel to the orbital rim over the mass. The mass is found anterior to the orbital septum. Blunt dissection should be done to free the dermoid from surrounding adhesions, taking care not to rupture the cyst. Pedicle attached to periosteum should be cut. Complete removal of intact cyst is mandatory and curative. Any rupture or cut in wall of cyst may lead to extensive inflammation in the surrounding area. Cryoprobe use may help seal a small rupture and deliver the cyst intact. In case of inadvertent rupture, profuse saline irrigation is recommended. Hemostasis should be maintained at all times with generous use of cautery. Closure is done in layers with absorbable sutures, using 6-0 vicryl for deep closure and 4-0/5-0 silk for skin closure.

Guerrissi et al. (2004) have described endoscope assisted surgery in three steps: hair line incision, exposure of the cyst and cyst removal. Using local anaesthesia, 18 patients were operated using endoscopic technique with minimal occult scar and excellent post-operative outcome. Out of these 18 patients, 90% remained without recurrence at 2 years of follow-up.

Deep orbital cysts are difficult to approach and imaging studies should be used to prepare a surgical plan. The surgical goal remains complete excision and removal without rupturing the cyst, as also avoiding injury to delicate orbital contents, including the ocular tissues, vessels and nerves. They may present a surgical challenge to the surgeon. For deep orbital cysts, the approach can be supero-nasal orbitotomy or more commonly lateral orbitotomy. A large majority of posterior orbital cysts are located in the upper temporal orbit and hence, postero-lateral orbitotomy is the recommended approach (Carta et al., 1998). They report 8 cases with posterior intraconal space orbital tumors using postero-lateral approach through a small opening on the orbital and temporal portions of the greater wing of the sphenoid. The lesser sphenoidal wing, the orbital plate of the frontal bone and the lateral rim of the orbit being maintained intact. This technique allowed adequate exposure of the orbital apex and successful removal of the tumors. On histological examination these tumors included four cavernomas, one dermoid cyst, a lymphoma, a hemangiopericytoma and a metastatic melanoma.

Intracranial extension of dermoid cyst requires a multi-disciplinary approach with head and neck surgeons and neurosurgeons. Gabibov et al. (1989) report transcranial approach on 12 cases with dermoid cysts of the orbit spreading into the cranial cavity. Orbital roof defect with extension to the lateral wall was present in these 8 patients and cyst capsule adhesion with the dura mater and orbital periosteum were present. They recommend use of an operative microscope and microsurgical instruments for enhanced visualization and control over surgery. In their series, the dermoid cysts were removed without injury to the dura mater and orbital periosteum with good functional and cosmetic results (Gabibov et al., 1989).
COMPlications

Orbital dermoid cyst may cause local complications due to mass effect. Giant orbital cysts may cause local deformity of the bone and ocular motility problems (Reim et al., 1975). Perinatally ruptured dermoid cyst has been reported as presenting with congenital oculomotor palsy (Coëvoet et al., 2000). Orbital dermoid cyst may be located within lateral rectus muscle (Howard et al., 1994) and can clinically mimic Duane’s syndrome type II (Gatziolis et al., 2002). Proptosis may be seen with slowly developing deep orbital dermoids. Cutaneous fistula due to dermoid cyst can present as a discharging sinus (Honig et al., 1998; Scolozzi et al., 2005; Wang et al., 1983) and may become infected (Wells et al., 2004). Lacrimal drainage pathway may be involved by orbital dermoid tumor (Hurwitz et al., 1982).

Complications may occur during surgical removal of the cyst. Rupture of dermoid cyst is the main complication if adequate care is not taken for localization and freeing up the cyst from its attachments to adjacent structures. Injury to adjacent orbital tissues during surgery can produce functional defects. Patient may show transient weakness of lateral rectus muscle due to surgical manipulation with lateral orbitotomy, which may subside in few months. Post-surgical hemorrhage may require re-surgery to control bleeding. Optic nerve damage may be inadvertent and a check on pupillary reaction helps avoid undue traction on globe and optic nerve manipulation in deep orbital tumors. Other rare complications include superior oblique palsy, which may occur with dermoid cyst impacting the region of the trochlea (Atilla et al., 2000). The orbital cysts may become infected (Ayache et al., 2003) and present with periorbital cellulitis in young child, requiring a craniofacial approach (Posnick et al., 1994).

SUMMARY AND CONCLUSION

Superficial orbital dermoid cysts are most common orbital tumors in children and usually have a typical presentation. Surgical removal of intact cyst has an excellent visual and cosmetic prognosis in such cases. Deep orbital dermoid cysts present later in life and are more complex to diagnose and treat. They require radiological imaging for planning surgical approach and may be difficult to remove. Higher risk of injury to orbital and ocular tissues exists with their surgery.

R. Ahuja and N. F. Azar

REFERENCES


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