Case study

Internal angular dermoid indenting on the globe

Running Title: Internal angular dermoid

UNDER PEER REVIEW

Abstract

Angular dermoid cysts are common periorbital tumours in children. They are tumours of embryonic origin that arise along bony sutures as a result of abnormal ectodermal sequestration during development. Angular dermoid cysts usually present in early childhood, are characteristically small benign and slow growing lesion. External angular dermoid present in the superotemporal quadrant is more common compared to internal angular dermoid in the superonasal quadrant. Early surgical excision is recommended and performed in the majority of cases, particularly to restore facial cosmesis.

Here we report an unusual case of a large internal angular dermoid cyst indenting the globe in a 3-year-old girl presented with left upper eyelid mass at medial angle since one year of age. The cyst was excised completely through a small superior lid crease incision.

Key words

Dermoid cyst, angular dermoid cyst, internal angular dermoid, superior eyelid crease incision.

1. INTRODUCTION

Dermoid cyst is a benign choristoma of the orbit, consist of keratinized epithelium with dermal appendages and adnexal structures such as hair follicles, sweat glands, and sebaceous glands, fat, smooth muscle and elastic fibres [1]. Angular dermoid cysts usually present in early childhood, are characteristically small benign and slow growing lesion. External angular dermoid is more common, present in the superotemporal quadrant; Internal angular dermoid in superonasal quadrant is relatively rare. Deeper orbital cysts are more insidious and may not be palpable or partially palpable, usually present later in older children or adults.

Angular demoid cyst usually present as a painless, subcutaneous, smooth, well-circumscribed, small round or oval masses in the periorbital region. However a large cyst may cause mechanical ptosis, but usually do not cause globe displacement. Here we report an unusual case of a large internal angular dermoid cyst indenting the globe in a 3-year-old girl presented with left upper eyelid mass at medial angle since one year of age.

2. CASE PRESENTATION

A 3-year-old girl presented with left upper eyelid mass at internal angle (Figure 1a). Her mother noticed a small swelling medially in the upper eyelid of left eye when the patient was one year old. It was painless and slowly progressively increasing in size. They consulted a local doctor and advised a CAT scan of orbit. A differential diagnosis of epidermid, dermoid and dacryocystocele was suggested by the radiologist. The patient was referred to this institute for further management. The patient was born at full term by normal delivery. Antepartal and intrapartal history was uneventful. General pediatric examination revealed a normally developed healthy child.

Ocular examination revealed, soft non-tender subcutaneous mass 1.6cm x 1.2 cm in size in left upper eyelid medially. The lesion was appear to be fixed to the underlying bone. There was no overlying discoloration, oedema, or erythema (Figure 1b). The globe was not displaced and extraocular motility was full. There was mild narrowing of palpebral aperture particularly medially in left eye. The lesion was present above the lacrimal sac area. Visual acuity was 20/20 in both the eyes. Anterior segment examination and fundus examination was within normal limits in both eyes.

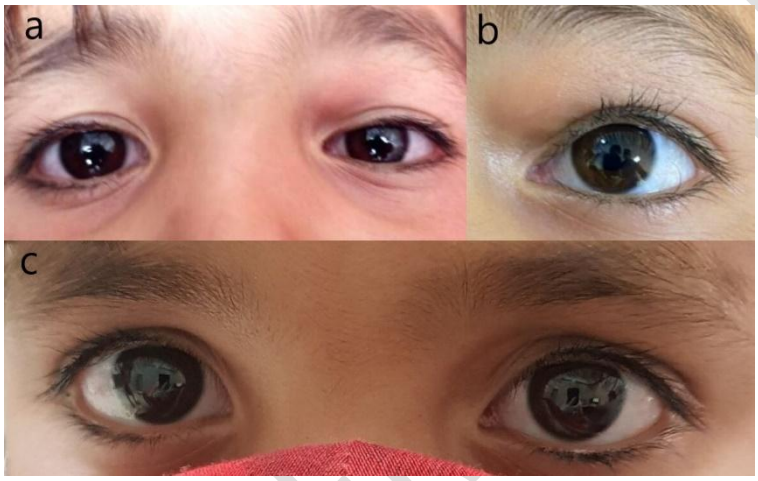

Figure 1: clinical photograph showing (a) Internal angular dermoid in the superomedial aspect of left upper eyelid with mild ptosis (b) Close-up view (c) postoperative appearance 2 month after surgery.

NCCT image revealed a well-defined rounded low attenuation cystic lesion of approximately 15x14x19 (AP x AT x CC) mm in the medial canthus of left orbit in extraconal region, indenting on globe. No obvious fat or calcification seen (Figure 2).

Figure 2: NCCT showing a well-defined rounded low attenuation cystic lesion (arrow) indenting on left eye.

UNDER PEER REVIEW

Excisional biopsy under general anaesthesia was planned. Surgical excision was done by a medial superior lid crease incision. The skin and pretarsal orbicularis muscle were incised. A sub muscular blunt dissection was performed to free the cyst circumferentially. Adhesions from deeper bone is dissected with periosteal elevator. The dissection was completed with westcotts scissor and cyst was removed intact (Figure 3). Incision was closed with polygalactin 6/0 sutures, and the skin was closed with black silk 6/0 sutures. Postoperative period was uneventful, no signs of inflammation noted. Cosmetic outcome after surgery was excellent with equal palpebral aperture height in both eyes (Figure 1c). The histopathological evaluation confirmed the diagnosis of dermoid cyst.

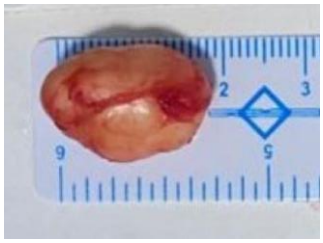

Figure 3: Gross appearance of excised dermoid cyst.

UNDER PEER REVIEW

3. DISCUSSION

Dermoid cyst is one of the most common space occupying orbital lesion; about 3-9 % of all orbital masses [2]. The majority are superficial or anterior periorbital lesion and present in early childhood. Deep orbital dermoid cysts are infrequent, difficult to diagnose, present late and need more extensive surgery [3].
The anterior periorbital dermoid cyst is most commonly present as external angular dermoid at the superolateral aspect of the orbit at the frontozygomatic suture. Internal angular dermoid is less common, present at the medial angle at frontoethmoidal or frontolacrimal sutures [4, 5]. Angular dermoid typically present in the first few years of life as painless, subcutaneous, smooth, well-circumscribed, round or oval masses in the periorbital region. In Most of cases the cyst is small around 1 cm in size, however a large cyst may cause mechanical ptosis, but usually do not cause globe displacement [2]. In this case there was mild ptosis and CAT scan showed a large cyst indenting on the globe. Deep orbital cysts often developing at the sphenozygomatic or sphenoethmoidal suture, are more insidious, and may present in late adolescence or adulthood with painless, progressive proptosis, motility deficits or diplopia due to mass effect on the surrounding structures [6].

External angular dermoid cysts may be easily diagnosed clinically without imaging if the posterior aspect can be palpated. Internal angular dermoid should be differentiated form other medial lesions like mucoceles, encephalocele, with the help of imaging. Mucoceles can be distinguished by their relationship to the sinuses, evidence of focal destruction of the bones, and associated opacification of the sinuses. In encephalocele a focal defect continuous with the cranial cavity may be noted, usually at the root of the nose. CT imaging is useful for detection of the orbital bone defect. But the intracranial connection and extent of cerebral tissue in an encephalocele is better defined with MR imaging [7]. Any of the solid tumours of

the orbit should be included in the differential diagnosis, especially if there is a focal bony defect [2].
Imaging is also useful in cases of periorbital dermoid to assess the extent of the lesion, and to rule out any intra orbital or intra cranial extension [8, 9]. On CT image, a dermoid typically has a hyper-dense wall and a hypo-dense cavity, which remains non-enhancing with contrast. On MRI The lesion is best visualized with fat-suppression techniques and appears as relatively hypointense with respect to orbital fat on T1-weighted images and relatively hyperintense on T2-weighted images and do not enhance with contrast.[5] MRI is particularly useful in the children as it does not expose the patient to radiation.

The definitive treatment of dermoid cyst is surgical excision with goal to remove the cyst intact without disrupting the cyst wall. An iatrogenic rupture and leakage of the cystic contents into the orbit can result in inflammation and recurrence. If the cyst is too large to excise intact, can be aspirated to facilitate removal. Small asymptomatic orbital dermoid cyst may be observed regularly [4]. However surgical excision of dermoid cysts is recommended to prevent the possibility of spontaneous rupture, cutaneous fistula formation, continued growth and pressure-related bony erosion which may affect facial symmetry and aesthetic appearance [2, 4].

The various surgical approaches have been evolved to remove dermoid cysts are direct approach over the mass; above, below, or through the brow and parallel to the superior orbital rim; superomedially with a Lynch incision and in the upper eyelid crease. [10, 11] The direct approach, is a simple and straightforward approach, however leaves a visible scar. The superior eyelid crease incision has the advantage of being a straightforward, with minimal complications and aesthetic outcome. A prospective study by Cozzi et al., demonstrated remarkably better scar cosmesis in the group that underwent superior eyelid crease incision compare to the trans-eyebrow incision technique [12]. The superior eyelid crease incision, provides sufficient access to the medial angular dermoid cysts and incision can be extended according to need, also allows excision of angular dermoid cysts some distance away from the incision. In addition, highly elastic skin facilitates easy traction and allow the incision to be hidden in the natural fold of the upper eyelid, making it barely perceptible.
Now the endoscopic technique for orbitofacial dermoid cyst excision have demonstrated with satisfactory cosmetic outcome, however complications like infection and transient frontal nerve paresis have been reported [13]. Furthermore, endoscopy require technical expertise and expensive equipment.

4. CONCLUSION

Angular dermoid cysts are common benign periorbital lesions. External angular dermoid cysts may be diagnosed easily however internal angular dermoid should be differentiated form other medial lesions like mucoceles, encephalocele, dacryocystcele. A large angular dermoid cyst may cause mechanical ptosis and further extension may rarely have compressive effects on the globe, therefore require an early surgical excision. CT and MRI are effective methods for diagnosis and determination of their site and extent, and helpful in deciding the surgical approach. The definitive treatment is surgical excision of the cyst without disruption of the cyst wall.

CONSENT

Written informed consent was obtained from the patient’s parents.

UNDER PEER REVIEW

ETHICAL APPROVAL

Not applicable.

REFERENCES

1. Pfeiffer RL, Nicholl RJ. Dermoid and epidermoid tumors of the orbit. Arch Ophthalmol 1948; 40:639-64.
2. Sherman RP, Rootman J, Lapointe JS. Orbital dermoids: clinical presentation and management. Br J Ophthalmol. 1984;68: 642-652.
3. Pollard ZF, Harley RD, Colhoun J: Dermoid cysts in children. Pediatrics 1976; 57: 379-82.
4. Shields J, Kaden I H, Eagle RC, Shields CL. Orbital Dermoid Cysts: Clinicopathologic Correlations, Classification, and Management, the 1997 Josephine E. Schueler Lecture. *Ophthal Plast Reconstr Surg.* 1997;13(4): 265-276.
5. Shields J and Shields C. Orbital Cysts of Childhood Classification, Clinical Features and Management. *Surv Ophthalmol.* 2004;49(3):281-299.
6. Henderson J and Garrity J. "Cysts and Celes" in *Henderson's Orbital Tumors*. Mayo Foundation for Medical Education and Research, 2007; pp. 33-39.
7. Morón, F. E., Morriss, M. C., Jones, J. J., & Hunter, J. V. (2004). Lumps and Bumps on the Head in Children: Use of CT and MR Imaging in Solving the Clinical Diagnostic Dilemma. RadioGraphics, 24(6), 1655–1674.
8. Pham N S, Dublin AB, Strong E W. Dermoid Cyst of the Orbit and Frontal Sinus: A Case Report. Skull Base 2010; 20 (4), 275-278
9. Chen L, Guo-Xiang Song G X, Tian WF, Hsu HT. Twenty-six cases of dumbbell-shaped orbital dermoids: preoperative diagnosis and management.Orbit.16 (4)1996; pp 233-240.

UNDER PEER REVIEW

10. Grove AS, McCord CD Jr: Orbital disorders: Diagnosis and management, in McCord C Jr,Tanenbaum M (eds): Oculoplastic Surgery. NewYork, Raven Press, 1987, pp 223-256.
11. Leone CR Jr: Surgical approaches to the orbit. Ophthalmology 1979;86:930-941.
12. Cozzi DA, Mele E, D'Ambrosio G, Totonelli G, Frediani S, Spagnol L. The eyelid crease approach to angular dermoid cysts in pediatric general surgery. J Pediatr Surg 2008;43(8):1502e6.
13. Huang, M. H. S., Cohen, S. R., Burstein, F. D., & Simms, C. A. (1997). Endoscopic Pediatric Plastic Surgery. Annals of Plastic Surgery, 38(1), 1–8.

UNDER PEER REVIEW