Case Report

Deep Orbital Dermoid Cyst Bulging into the Superior Orbital Fissure: Clinical Presentation and Management

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Abstract

Purpose: To present a case of deep orbital dermoid cyst with emphasis on clinical presentation, imaging spectrum, differential diagnosis and management.
Case Report: A 28-year-old female was referred to our hospital with chief complaint of drooping of right eyelid and progressive headache. Ocular motility, visual acuity and fundus examination were normal. computed tomography (CT) and magnetic resonance imaging (MRI) revealed a well-defined, intraosseous deep orbital dermoid cyst (5.9 mm × 12.5 mm) located near the apex of right orbit, extending from greater wing of sphenoid into the superior orbital fissure. Due to occulomotor nerve (superior and inferior divisions) compression which passes through the superior orbital fissure, ipsilateral headache and ptosis occurred. Complete surgical excision of cyst was performed using noninvasive extracranial lateral orbitotomy approach. After removal of the cyst, curette and cutting drill were used to thoroughly remove any residual cystic content. Histopathological analysis confirmed the diagnosis. The healing was uneventful postoperatively.
Conclusion: CT and MRI are easy, reliable, safe and effective imaging methods for establishing the diagnosis of orbital dermoid cyst. Size, location and manifestations are the most important determinants of the disease management. Complete surgical excision without rupture of the cyst is the treatment of choice.

Keywords: Choristoma; Dermoid Cyst; Orbit; Ptosis; Surgery

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INTRODUCTION

Dermoid cysts of the orbit are benign choristomas that originate from the aberrant primordial tissue trapped along bony suture lines or diploe of the orbital bones during embryologic development between the third and the fifth weeks of gestation and more rarely from posttraumatic implantation of the surface epithelium.[1,2] Orbital dermoid cyst is a distinct pathological entity constituting less than 5% of cases of intraorbital neoplasms.[3] The superior temporal quadrant at the frontozygomatic suture is the most common location, followed by nasoglabellar region at the frontoethmoidal suture.[4]

On imaging, the cyst may show fat, fluid or soft tissue signal and occasionally may show calcification. A computed tomography (CT) attenuation similar to fat is relatively frequent and considered as pathognomonic.[5] This case report is unusual as it is concerned with the description of a distinct

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disease entity and its radiological appearances for early preoperative diagnosis. Better understanding and management of the disease are discussed in the pertinent light of literature.

CASE REPORT

A 28-year-old woman presented with a one month history of progressive ptosis of her right eyelid associated with chronic headache. There was no history of vomiting, ocular pain, tearing, diminished vision, squint, proptosis, cervical pain, trauma or related surgical intervention. The patient was carefully examined for overall appearance, lid and globe positions, pupillary reaction, ocular balance and motility in the cardinal directions of gaze. Slit lamp and fundus examination findings were normal. There was no relative afferent pupillary defect. The visual acuity was 20/20 bilaterally and the intraocular pressure was 17.25 mmHg bilaterally. There was no palpable mass in the orbit. On imaging, CT revealed a well-defined, mixed density intraosseous abnormal lesion (5.9mm × 12.5mm) near the right orbital apex, extending through the greater wing of sphenoid and bulging into the superior orbital fissure. The lesion appeared predominantly isodense to brain with few areas of intralesional fat density [Figure 1a]. Magnetic resonance imaging (MRI) T1-weighted images revealed an iso- to hyperintense intraosseous lesion extending through the greater wing of sphenoid into the superior orbital fissure near the apex of the orbit [Figure 1b]. MRI STIR images showed suppression of the hyperintensity seen in T1 weighted images, confirming the characteristic fat component of the lesion [Figures 1c and d].

Surgical excision was performed and the cyst was removed completely, using a lateral crease incision to expose the lateral orbital rim via a non-invasive extracranial lateral orbitotomy approach. The cyst was dissected circumferentially with a blunt instrument from the periorbital soft tissues to avoid rupture of the cyst. The periosteum around the bony defect was incised to dissect the visible safety margin of the cyst extending into the superior orbital fissure and was elevated from the lateral orbital wall and temporalis fossa. Osteotomies were made with an oscillating saw just above the zygomatico-frontal suture and just above the zygomatic arch. After removal of the cyst, curette and cutting drill were used to thoroughly remove any residual cystic content. Histopathological analysis showed a cyst lined by keratinized stratified squamous epithelium with keratin, skin appendages and lipid debris in the lumen, consistent with a dermoid cyst. The healing was uneventful postoperatively and the patient's ptosis and headache resolved completely during a six-month follow-up period.

DISCUSSION

Dermoid cyst is a benign heterotopic inclusion cyst which is not common in the orbital and intracranial cavities; however, no organ of the body is immune. They should be considered in the differential diagnosis of orbital cystic lesions such as teratoma, choristoma (epidermoid and dermolipoma), colobomatous cyst, and the congenital cystic eye.

Deep orbital dermoid cyst is a diagnostic and therapeutic challenge due to its highly variable clinical presentation and non-specific symptoms and should be considered in the differential diagnosis of benign bone-lytic lesions affecting the orbit. The clinical presentation depends on the location, size, rate of growth, intracranial extension and correlation to adjacent structures.[6] Varied clinical signs range from a mass with lid swelling, ptosis, globe displacement, proptosis to ocular protrusion, ocular dissimilarity, limitation of ocular motility and optic nerve compression syndrome.[1-3] Ruszkowski et al suggested that deep seated dermoid cysts could be associated with pain due to pressure induced stretch of a related sensory nerve.[7] Our patient also suffered from headache and ptosis without any signs of orbital inflammation.

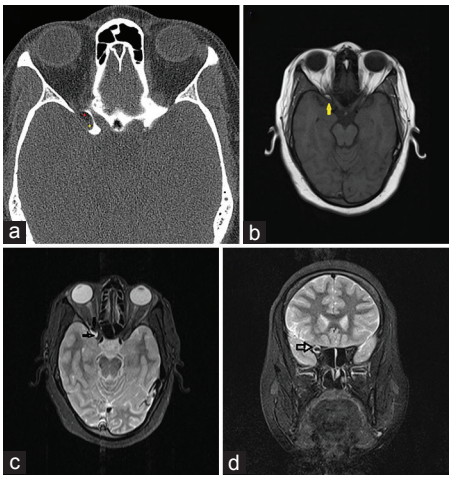

Figure 1. (a) Axial non-contrast computed tomography shows a well-defined, mixed density intraosseous abnormal lesion near the right orbital apex, extending through greater wing of sphenoid and bulging into the superior orbital fissure. The lesion appears predominantly isodense to brain (+26 Hounsfield units) with few areas of intralesion fat density (−71 Hounsfield units). (b) MRI T1-weighted image showing well defined, iso to hyper intense intraosseous abnormal lesion arrowhead) near the right orbital apex, extending through greater wing of sphenoid and bulging into the superior orbital fissure. (c and d) MRI STIR axial and coronal images exhibiting area of fat suppression (arrowhead) in the abnormal intraosseous lesion near the right orbital apex, extending through greater wing of sphenoid and bulging into the superior orbital fissure, again confirming the fat component of the lesion, thus signalling towards dermoid cyst.

Periocular and orbital dermoid cysts are generally categorized into superficial and deep lesions, with superficial lesions presenting early in life.[1] Pryor et al reviewed 49 cases of pediatric dermoid cysts and found that dermoid cyst commonly occurs in the periorbital region (61%), anterior to the frontozygomatic suture line followed by midline nasal and forehead dermoids (16%).[8] Deeper orbital dermoids grow indolently and remain clinically occult until adolescence or adulthood.[1,8] Our patient is a rare case of a deep orbital dermoid extending into the superior orbital fissure, causing ophthalmic and neurologic symptoms due to a probable third nerve compression, a finding that has not been previously described in the literature. Although the vision was prefect and the evidences of the optic nerve compression was not apparent yet, the authors decided to surgically remove the lesion regarding the rapidly progressive nature of the disease and proximity of the lesion to the optic nerve canal. An upper eyelid incision provides adequate exposure of most orbital lesions.[4] However, in our case, complete surgical excision of the cyst was performed using non-invasive extracranial lateral orbitotomy approach. Dermoid cysts should be differentiated from the epidermoid cysts although both are usually grouped together; however, they are different entities with various clinical behaviours. A careful histopathologic examination suggests dermoid cyst characterization by the presence of mesodermal elements such as hair follicles, sebaceous, and sweat glands. Within the cyst, keratin, hair, smooth muscle, and lipid debris may be found.[2] In our case, histopathological analysis showed skin appendages in the cyst wall, consistent with a dermoid cyst.

Imaging modalities such as B-scan ultrasonography, CT scan, and MRI of the orbital dermoid cysts are valuable in the early preoperative diagnosis, to demonstrate their intraorbital and intracranial extension, thereby determining the surgical intervention strategy.[9-12] Chawda and Moseley reviewed CT images of histologically proven orbital dermoid cysts and found lateral canthus was the most common site with male predilection. They suggested orbital dermoids are tumors of childhood. Bone sparing, calcification, fluid level, no visible wall and no abnormal soft tissue outside the cyst are infrequent. A CT attenuation similar to fat is relatively frequent.[5] In the current study, the CT image of our adult female patient revealed a well-defined, mixed density intraosseous lesion which appeared predominantly isodense to the brain (+26 Hounsfield units) with few areas of intralesion fat density (–71 hounsfield units) [Figure 1a]. MRI T1-weighted images exhibited an iso- to hyperintense lesion near the orbital apex in the superior orbital fissure [Figure 1b], which showed reduced intensity with fat suppression images, the latter again confirming the characteristic fat density of the lesion [Figures 1c and d]. Both CT and MRI findings were consistent with dermoid cyst.

In conclusion, the present case is an exceedingly rare occurrence of unilateral ptosis and ipsilateral headache resulting from deep bony orbital dermoid cyst primarily located near the apex of the right orbit and bulging into the superior orbital fissure. CT and MRI are easy, reliable, safe, and effective imaging methods for establishing the diagnosis. The size, location and manifestations are the most important determinants of the disease management. Complete surgical excision without rupture of the cyst is the treatment of choice.

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Nil.

Conflicts of Interest

There are no conflict of interest.

REFERENCES

1. Ahuja R, Azar NF. Orbital dermoids in children. *Semin Ophthalmol* 2006;21:207-211.
2. Shields JA, Shields CL. Orbital cysts of childhood- Classification, clinical features and management. *Surv Ophthalmol* 2004;49:281-299.
3. Grove AS. In: Oculoplastic Surgery. McChord CDJ, editor. Orbital disorders: Diagnosis and management. New York: Raven Press; 1981. pp. 274-277.
4. Bartlett SP, Lin KY, Grossman R, Katowitz J. The surgical management of orbitofacial dermoids in the pediatric patient. *Plast Reconstr Surg* 1993;91:1208-1215.
5. Chawda SJ, Moseley IF. Computed tomography of orbital dermoids: A 20-year review. *Clin Radiol* 1999;54:821-825.
6. Veselinović D, Krasić D, Stefanović I, Veselinović A, Radovanović Z, Kostić A, *et al*. Orbital dermoid and epidermoid cysts: case study. Srpski Arhiv za Celokupno Lekarstvo 2010; 138:755-759.
7. Ruszkowski A, Caouette-Laberge L, Bortoluzzi P, Egerszegi EP. Superior eyelid incision: An alternative approach for frontozygomatic dermoid cyst excision. *Ann Plast Surg* 2000;44:591-595.
8. Pryor SG, Lewis JE, Weaver AL, Orvidas LJ. Pediatric dermoid cysts of the head and neck. *Otolaryngol Head Neck Surg* 2005;132:938-942.
9. Kaufman LM, Villablanca JP, Mafee MF. The Radiological Clinics of North America. Philadelphia: W B Saunders Company; 1998. pp. 1149-1163.
10. Stark DD, Bradley WG. Magnetic resonance imaging. 2nd ed. St. Louis: Mosby; pp. 801-2. pp. 1016.
11. Osborn AG. Diagnostic Neuroradiology. Mosby; 1997. pp. 66-67.
12. Nugent RA, Lapointe JS, Rootman J, Robertson WD, Graeb DA. Orbital dermoids: Features on CT. *Radiology* 1987;165:475-458.