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# Congenital sphenoid wing defect with lateral angular dermoid presenting with CSF leak after orbital surgery

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## ABSTRACT

Lateral angular dermoid may have associated bony defects which must be recognized before surgery. High-resolution 3-D CT scans and MRIs define these lesions very clearly. Total excision with attention to the bony defect is usually curative. CSF leak from the operative wound at the surgery or in the post-operative period signifies the intracranial connection or dural adhesion unnoticed by the Ophthalmologist before surgery.

## INTRODUCTION

Dermoid cysts are congenital benign tumours that originate from an embryonic ectodermal cell. Angular dermoid present at the outer angle of eye, beneath or in close proximity to the eyebrow. Various types of bony defects are associated with long standing dermoid in this location. Congenital sphenoid wing dysplasia (SWD) is a rare occurrence. It has a strong association with Neurofibromatosis type I (NF-1)<sup>1</sup>, although it can occur alone. We present a case of a 3 years old male child presenting with orbital cellulitis, cerebrospinal fluid (CSF) leak after orbital surgery for lateral angular dermoid with congenital sphenoid wing dysplasia, without any features of NF-1.

## CASE REPORT

A 3 years old male child presented with CSF leak from wound near lateral canthus of right eye with suture breakdown and features of meningitis (Fig 1A). He had history of recurrent right orbital swelling on crying since birth, and fowl smelling discharge from the swelling before surgery. Excision biopsy of the swelling was reported to be a dermoid cyst. Three-dimensional CT scan with CT-cisternogram showed a bony defect in the right sphenoid wing forming lateral wall of orbit (Fig 1B) and CSF pooling at lateral angle of orbit (Fig 2A). MRI revealed evidence of soft tissue thickening and fluid collection in extraconal compartment,

## Keywords

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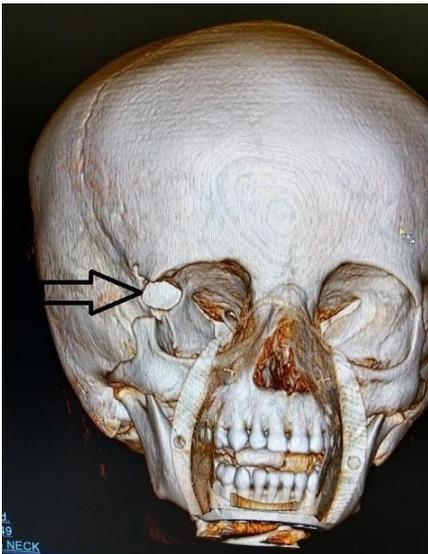


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with orbital and para-orbital cellulitis. We performed a right pterional craniotomy, excision of infected residual dermoid, wound debridement and CSF fistula repair with the help of a rotated pedicled temporalis muscle graft and Tisseel (Fibrin) glue. In the immediate post-operative period, there was no CSF leak (Fig 2B).



**Figure 1A.** CSF leak from operative wounds with breakdown of stitches (white solid arrows)



**Figure 1B.** 3D-CT of face showing a large bony defect in the greater sphenoid wing, involving the orbital rim and zygomatic arch on the right side (black arrow)



**Figure 2A.** CT-cisternogram showing pooling and leakage of contrast at the defect site



**Figure 2B.** Post-operative image

## DISCUSSION

Dermoid cysts are congenital benign tumours that originate from an embryonic ectodermal cell. These ectodermal cells are involved in neurulation during neural tube closure resulting in abnormal fusion. Common sites of dermoid cysts are periorbital (zygomatico-frontal suture), nasal (fronto-nasal suture and rhinion), intraoral (floor of mouth), scalp (anterior fontanelle and cranial sutures) and postauricular<sup>2</sup>. Angular dermoid occurs at the outer angle of eye, beneath or in close proximity to the eyebrows<sup>3</sup>. These tumours are ovoid, soft to firm, may be lobulated, containing a lipomatous element having a cystic nature.

SWD is a characteristic feature and one of the diagnostic criteria of NF-1. It occurs in 5-10% of the cases. Features of NF-1 are six or more café-au-lait macules, 2 or more cutaneous/ subcutaneous neurofibromas or plexiform neurofibromas, axillary or groin freckling, optic pathway glioma, two or more Lisch nodules in iris, bony dysplasia (SWD, bowing or long bone +/- pseudoarthrosis) and first degree relative of NF-1. SWD can be an isolated finding also, like in our case, which is extremely rare.

Zygomatico-frontal suture defect has a strong association with orbital dermoid cyst<sup>4</sup>.

N. Sathanathan et.al reviewed CT scan of 70 patients (43 male and 27 female) who had a lesion palpable at outer canthus, and histologically proven dermoid cyst<sup>5</sup>. Mean age of patients was 29 years. The lesion was always unilateral and 34 were on the left. The bone of the lateral wall and supero-temporal angle of orbit showed following abnormalities, often in combination: pressure erosion in 61 cases, abnormal developmental shape

in 55, the dermoid cyst entering a tunnel or canal through the lateral wall in 24, a blind pit or crater in 15, and a cleft in 20.

There are reports of unusual and interesting presentations of angular dermoids<sup>6,7,8</sup>. Proptosis of eye with mastication was reported in a woman because of orbital mass communicating with the temporal fossa through a bony defect in lateral orbital wall; pressure by the temporalis muscle with mastication, on the dumb-bell shaped cyst caused the proptosis<sup>6</sup>. Frontal headache and chewing related oscillopsia is reported in 60 years old man<sup>7</sup>. Illusion of movement was due to mechanical displacement of retro-orbital cyst and eye by contraction of the temporalis muscle; removal of the cyst leads to complete remission of oscillopsia. History of blurring of vision of an eye with chewing is reported in a 29 years old lady with a dumb-bell dermoid cyst<sup>8</sup>.

In this endoscopic era, direct surgical excision is an excellent approach, with a low complication rate and a very high aesthetic success rate, when performed through supra-eye brow or infra-eyebrow approach<sup>9,10</sup>.

Proper anatomical delineation of the lesion and the bony defect with High resolution CT and MRI imaging is extremely important as many of the angular dermoid have intracranial extension. Involvement of a neurosurgeon in such cases is thus imperative and combined transcranial approach is done where indicated.

## CONCLUSION

Lateral angular dermoid is associated with superolateral bony abnormality of orbital wall. Congenital sphenoid wing dysplasia is a spectrum of bony defect. We present a case of congenital sphenoid wing dysplasia in a case of lateral angular dermoid presenting in post-orbital surgery CSF leak. Proper anatomical delineation of angular dermoid is extremely important as there are high chances of intracranial extension of the angular dermoid, and transcranial approach for complete excision for possible intracranial extension have to be kept in mind while dealing with angular dermoid.

**Conflicts of interest:** None declared.

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