A Challenging Case of Paediatric Clival Encephalocele

Arshad Zubair, MRCS(ENT)[1,2]; Samuel Leong, FRCSEd(ORL-HNS)[2]; Ajay Sinha, FRCS(SN)[1]; Grace Khong, FRCSEd(ORL-HNS)[1]

[1]Alder Hey Children's NHS Foundation Trust, Liverpool UK, [2]Liverpool University Hospitals NHS Foundation Trust, Liverpool, UK

Introduction

- Paediatric clival encephalocele is an exceedingly rare pathology, with handful of cases reported in literature[1-3]
- Basi-occipital clival defects are believed to be caused by persistent notochordal remnants
- In adults, clival encephaloceles are commonly repaired via transnasal endoscopic approach
- We report a paediatric clival encephalocele repaired via transnasal endoscopic approach and reflect on challenges encountered in this case perioperatively

Background

6 year old

Overseas patient

Cyclical vomiting syndrome

Recurrent meningitis

Previous transoral repair of clival encephalocele at age of 3

Surgical Management

Primary surgery

Transnasal endoscopic approach repair with multilayer reconstruction using fat, haemopatch onlay graft, middle turbinate(MT) graft, and nasoseptal flap (NSF)

Foley catheter for reinforcement and lumbar drainage (LD) in situ for 5 days

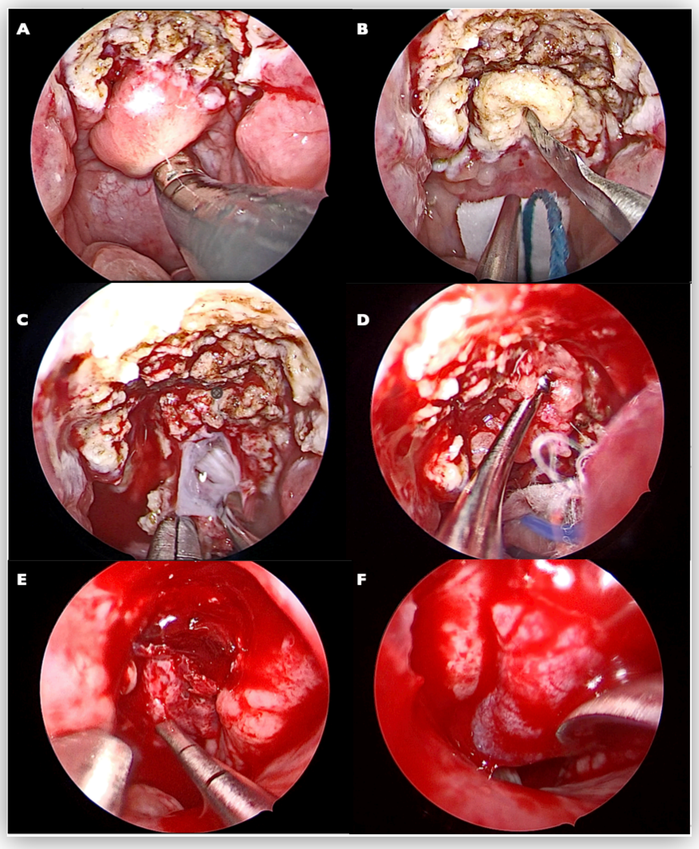

Figure 1A-F. Intraoperative endoscopic images. (A) Reduction of encephalocele following adenoidectomy for exposure. (B) Incision of overlying mucosa. (C) Division of arachnoid-mucosal adhesions. (D-F) Multilayer reconstruction with fat graft, middle turbinate graft and nasoseptal flap respectively.

Postoperative course

POD-2 Developed stress-induced vomiting

POD-9 Developed meningitis and commenced on IV antibiotics

POD-13 Minor recurrent nasal discharge - positive for B2 transferrin

Revision surgery

POD-15 Return to theatre.

NSF found to have migrated. MT graft removed. Repair reinforced with Tachosil, Bioglue and LD re-insertion

POD-30 LD removed

POD-55 Discharged home

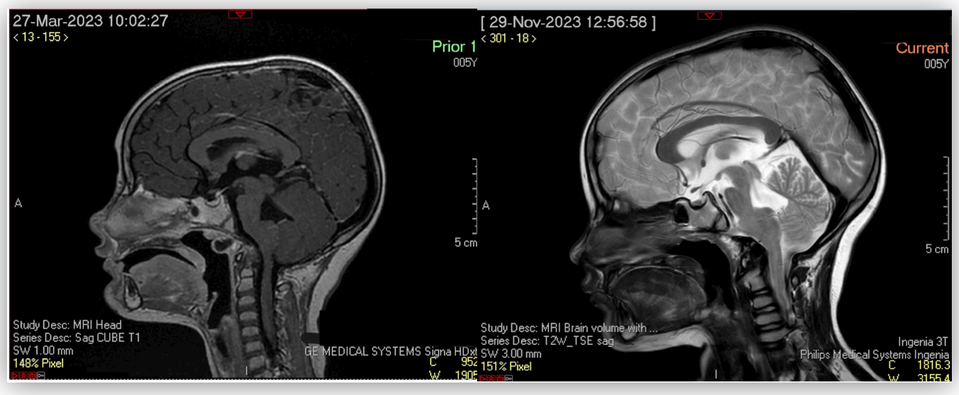

Figure 2. MRI Head scan showing preoperative (left) and postoperative (right) appearances.

Discussion

Case Challenges

Known difficulties in repair of clival skull base defects

Technicalities of using NSF in children

Tolerability of Foley catheter in children

Raised ICP due to stress-induced vomiting

Overseas patient & associated considerations for discharge

- Clival defects are among the most difficult reconstructions due to their dependant location with high CSF flow[4]
- These often need a wider flap due to horizontal orientation of the NSF when it is transposed[4]
- Special considerations for NSF in children include nasal septum length as well as length of anterior skull base[5]
- Early experiences of endoscopic repair of high-flow CSF leaks suggest that paediatric patients are considered a high-risk group for postoperative repair failure, though more recent evidence suggests otherwise[6,7]
- Tachosil and BioGlue have been demonstrated to be safe in anterior skull base surgery[8,9]
- Post-operative CSF diversion via lumbar drainage minimises post-operative CSF leaks (prospective RCT)[10]

Conclusions

- Transnasal endoscopic repair is an effective management option for paediatric clival encephalocele
- Optimal positioning and multilayer reinforcement of nasoseptal flap for clival defects is challenging
- Placement of prophylactic lumbar drain is strongly recommended

Contact

Arshad Zubair
Liverpool University Hospitals Foundation Trust
Department of ENT, Aintree Lodge, Aintree University Hospital, Liverpool L9 7AL
arshad.zubair@nhs.net

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