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CASE REPORT

Tension pneumocephalus in a patient with NF1 following ventriculoperitoneal shunt—deciphering the cause and proposed management strategy

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Abstract

Introduction Spontaneous pneumocephalus following ventriculoperitoneal shunting is a very unique complication, seen in a handful of patients. Small bony defects form as a result of chronically raised intracranial pressure, which can later lead to pneumocephalus once intracranial pressure decreases following ventriculoperitoneal shunting.

Case report Here, we present a case of a 15-year-old girl with NF1 who presented to us with pneumocephalus 10 months following shunting and our management strategy along with a literature review of this condition.

Conclusion NF1 & hydrocephalus can lead to skull base erosion, which needs to be looked up before proceeding with VP shunting to avoid delayed onset pneumocephalus. SOKHA with the opening of LT is a minimally invasive approach suitable to tackle both problems simultaneously.

Keywords Tension pneumocephalus · Ventriculoperitoneal shunt · Supraorbital keyhole approach · Skull base defect

Abbreviations

AS	Aqueductal stenosis
CT	Computed tomography
CSF	Cerebrospinal fluid
ETV	Endoscopic third ventriculostomy
ICP	Intracranial pressure
LT	Lamina terminalis
NCCT	Non-contrast computed tomography
NF	Neurofibromatosis
SOKHA	Supraorbital keyhole approach
VP	Ventriculoperitoneal

Introduction

Pneumocephalus following ventriculoperitoneal (VP) shunt surgery is a rare complication with few cases reported in the literature [1–9]. Small skull base defects, missed on initial scans, resulted in this complication in most reported cases. Both chronic hydrocephalus and neurofibromatosis (NF)1 can cause such defects independently. In this article, we have described our strategy for dealing with both hydrocephalus/pneumocephalus and encephalocele via a single surgical approach and the underlying pathophysiology.

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Case summary

A 15-year-old girl, a known case of Neurofibromatosis 1 (NF1), presented with headache and vomiting for 7 days and altered sensorium for 5 days. There was a history of trivial fall without head injury 2 weeks prior to the onset of symptoms. She underwent a VP shunt 10 months back, when she presented with headache, progressive diminution of vision, and urinary incontinence and on evaluation was found to have aqueductal stenosis (AS). On examination, there were no focal neurological deficits or signs of meningitis. A non-contrast computed tomographic (NCCT) scan done showed tension pneumocephalus with pneumoventricle and shunt in situ (Fig. 1a). The left frontal pneumocyst was seen communicating with the ventricle. There was no apparent bony defect in the calvaria. The shunt assembly was intact. It was only on a thin-cut CT head bone window, a small bony defect in the posterior wall of the left frontal sinus was made out (Fig. 1b, c). The left supraorbital keyhole approach (SOKHA) was planned to address the encephalocele. The VP shunt was removed.

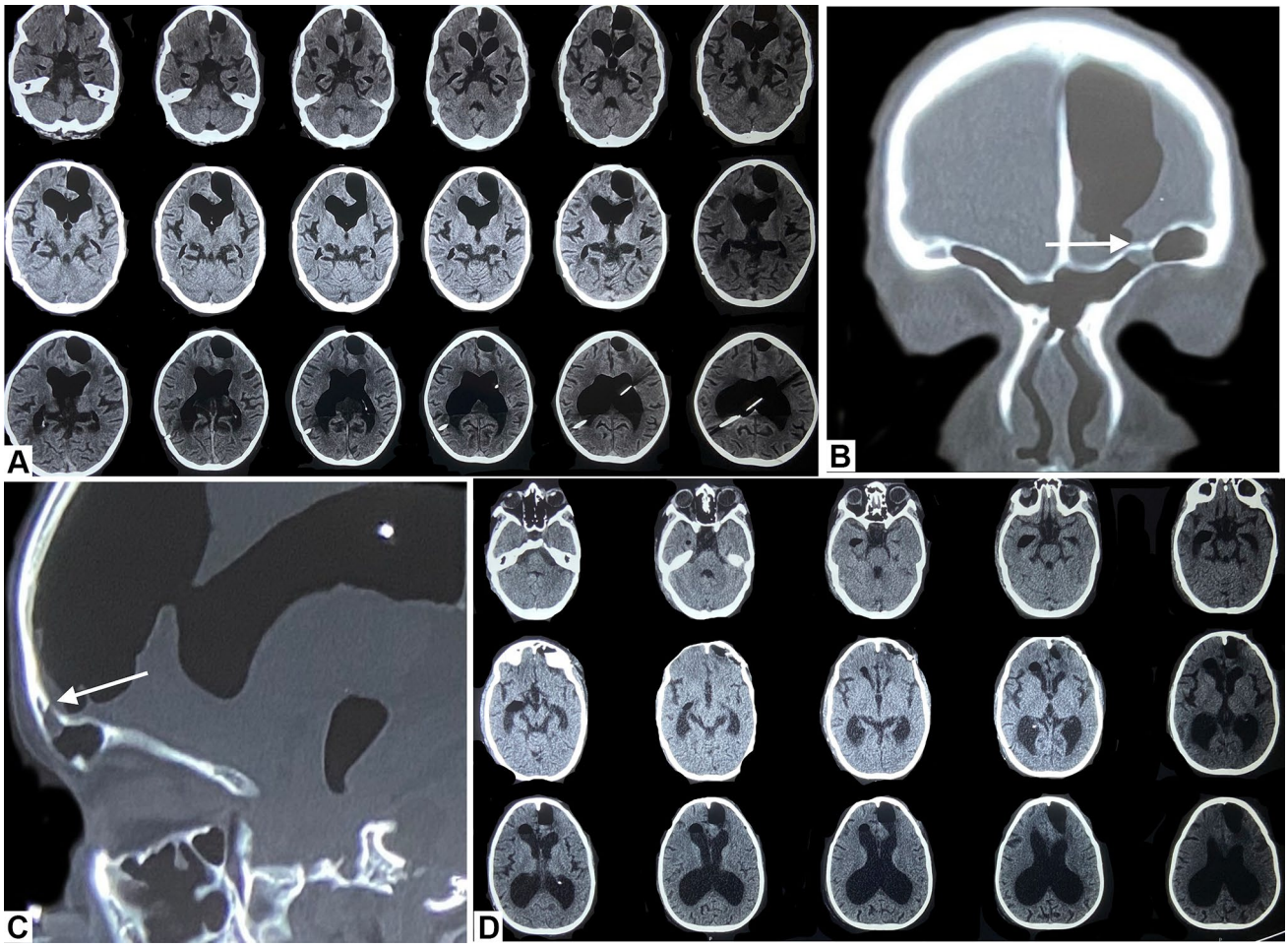

Fig. 1 **A** Preoperative NCCT showing the presence of pneumoventricle with a shunt in situ. An air pocket present in the left frontal area can be seen clearly communicating with the lateral ventricle. **B** CT head—coronal section showing a defect (marked by white arrow) in the posterior wall of the frontal sinus. **C** CT head—sagittal section—clearly shows a defect (marked by a white arrow). **D** Post-operative scan demonstrating partial resolution of hydrocephalus and pneumoventricle

Extra-dural exposure of the dural and adjacent bony defects was done (Fig. 2a). The herniating gliotic brain was resected. The frontal sinus was completely exteriorised and packed with fat harvested from the abdomen. After dural opening, the bulging lamina terminalis was opened, thus relieving the pneumoventricle and simultaneous internal cerebrospinal fluid (CSF) diversion (Fig. 2b). Dural defects were covered with artificial dura and reinforced with a vascularized pericranial graft (onlay technique). The patient made an uneventful recovery and post-operative imaging on day 7 revealed partial resolution of pneumocephalus along with the resolution of hydrocephalus (Fig. 1d). The patient was asymptomatic at 3 months of follow-up (Fig. 3).

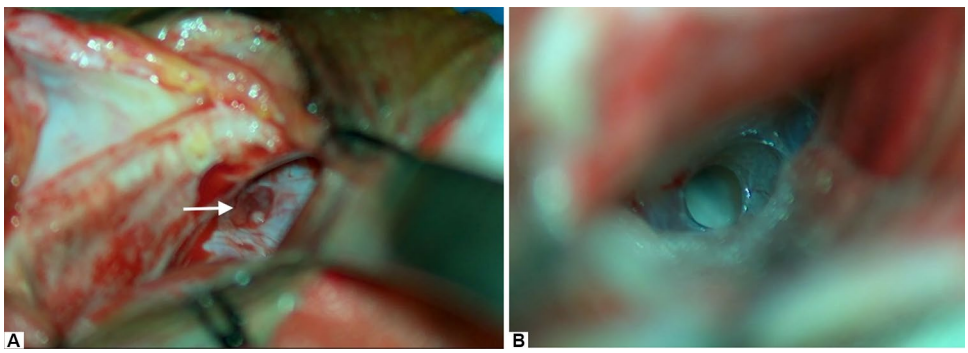

Fig. 2 **A** Intraoperative photograph showing a well-defined dural defect (marked by a white arrow). **B** Intraoperative photograph after the opening of the lamina terminalis

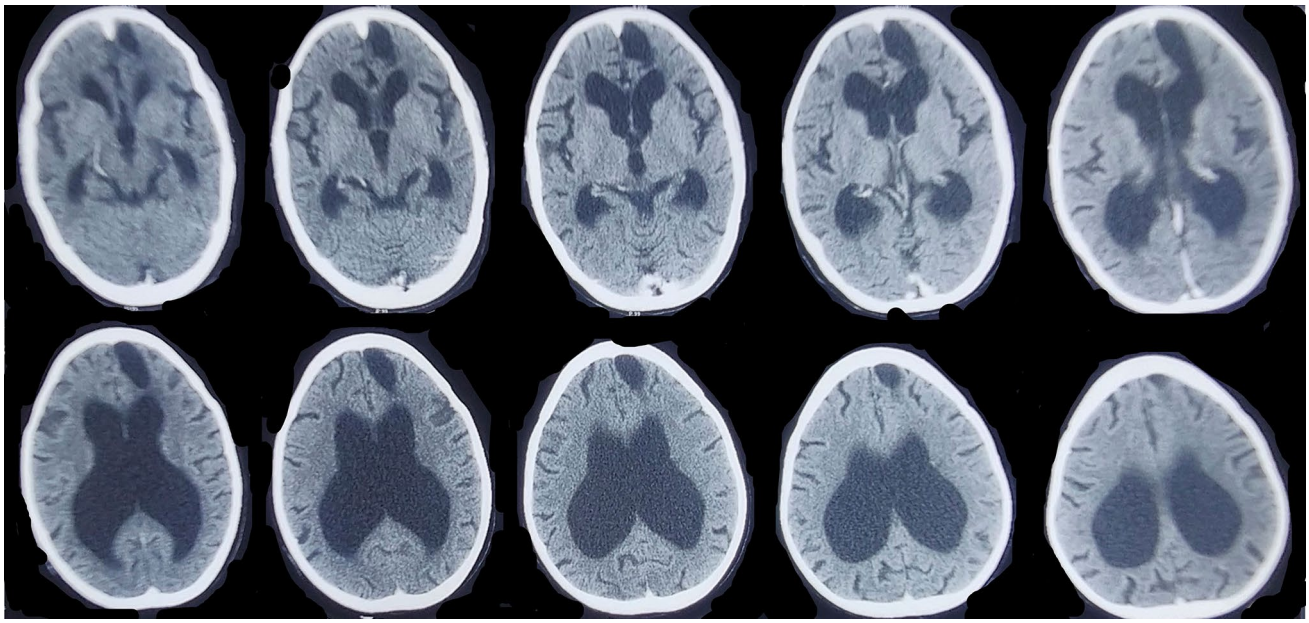

Fig. 3 Follow-up CT at 3 months shows complete resolution of pneumocephalus with no change in ventricle size compared to post-op scan

Discussion

First described by Kessler and Stern in 1962 [1], pneumocephalus following ventriculoperitoneal shunting is a rare problem with the literature limited to a few case reports only [2–9]. Owing to its rarity, decision-making is often individualized and dependent on the neurosurgeon's experience. In the following sections, we have tried to shed some light on the underlying pathophysiology, surgical approach, and management options for this unique problem.

Neurofibromatosis and encephalocele—the "two-hit hypothesis"

Both NF1 and chronic-raised intracranial pressure can result in spontaneous encephalocele formation by causing skull base defects. The effect of NF1 on bones developing by endochondral ossification like sphenoid bone and occipital is very well documented in the form of multiple dysplasias. However, its effect on calvarial bones like frontal bone, which develop by membranous ossification, is less clear. Chronically raised ICP can independently lead to multiple dural defects causing frontal intra-diploic encephalocele formation [10]. Most of these defects are limited to the anterior cranial fossa [3, 5, 8, 9], followed by the middle cranial fossa [4]. Only 13.3% ($n=4$, including this case) of the reported cases of tension pneumocephalus following VP shunt had NF1 along with chronic hydrocephalus (Table 1). Thus, raised ICP may be an independent factor in most cases for the occurrence of spontaneous encephalocele. Interestingly, most of these patients had undergone VP shunt for a late onset aqueductal stenosis [2, 5, 6, 8, 9].

Pathophysiology and evaluation

Following the ventriculoperitoneal shunt, air can enter the skull via either the distal catheter (shunt breakage or ruptured viscus) or through a defect in the skull base [1, 6]. However, the most common reason attributed to this is the underlying skull base defect. After shunting as ICP decreases, along with the siphoning effect of the shunt, air can get sucked in through these potentially weak areas. These defects act as a ball-valve action, and hence the air pocket will keep on increasing with time. A dip in the intracranial pressure along with underlying bony and dural defects is what initiates this irreversible series of events.

Evaluation of patients presenting with suspected shunt malfunction includes an NCCT head to look for the status of the ventricles and shunt. A thin-cut bone window CT scan is shown to have good resolution for the delineation of the defect in the bone, as was used in our case also.

Surgical strategy

Once the dural defect is identified on imaging, the next step is to repair it. The location of the defect is perhaps the most important factor when considering a surgical repair of these defects. Among open approaches, bifrontal or unifrontal craniotomy with coverage of defect using vascularized pericranial graft is the most commonly employed surgical strategy [2, 5]. The endoscopic approach is sometimes limited to

Table 1 Reported cases of spontaneous tension pneumocephalus following VP shunt

S No	Author/reported year	Age/gender	NF1	Indication of VP shunt	Duration b/w shunt and pneumocephalus	Site of bony defect	Management
1	Pitts et al. 1975 [11]	21/M	No	CH	1 year	Tegmen tympani (petrous bone)	Mastoidectomy
2	Little and Mac-Carty 1976 [9]	21/M	No	AS	2 months	Frontal sinus	Craniotomy and repair
3	Ikeda et al. 1978 [12]	22/F	No	AS	3 months	Frontal sinus	Craniotomy and repair
4	Steinberger et al. 1979 [13]	29/F	No	AS	1 week	Frontal sinus	Craniotomy and repair
5	Findler et al. 1980 [14]	18/M	No	AS	8 years	Frontal sinus	Aspiration and anti-siphon device
6	Jooma and Grant 1983 [15]	12/M	No	AS	6 months	Cribriform plate	Craniotomy and repair
7	Sunada et al. 1984 [16]	15/M	Yes	AS	2 months	Skull base (exact site not known)	Shunt revision
8	Komisar et al. 1985 [17]	50/M	No	CH	14 months	Middle cranial fossa base	Craniotomy and repair
9	Ruge et al. 1985 [6]	28/M	No	AS	9 years	Ethmoid sinus	Shunt externalization
10	Tanaka et al. 1986 [7]	47/M	No	VS	2 weeks	Mastoid air cells	Craniotomy and repair
11	Tanaka et al. 1986 [7]	39/M	No	VS	8 months	Mastoid air cells	Craniotomy and repair
12	Mylonas 1991 [3]	77/F	No	CH	6 weeks	Sellar floor	Craniotomy and repair
13	Mylonas 1991 [3]	24/F	Yes	CH	2 years	Crista galli	Craniotomy and repair
14	Cantisani et al. 1999 [18]	33/M	No	AS	2 years	Not known	Not known
15	Kuba et al. 2000 [19]	29/F	No	AS	7 years	Ethmoid sinus	ETV and shunt
16	Ugarriza et al. 2001 [4]	15/F	Yes	IV tumor	8 months	Frontal sinus	Anti-siphon device
17	Ugarriza et al. 2001 [4]	62/F	No	Clinoidal meningioma	5 years	Not known	Anti-siphon device
18	Mineo et al. 2004 [20]	14/M	No	PFT	na	Petrous bone	Craniotomy and repair
19	Sankhla et al. 2004 [5]	19/F	No	AS	6 months	Frontal sinus	Frontal craniotomy and repair
20	Czepko et al. 2005 [21]	38/F	No	AS	2 months	Sellar floor	Frontal craniotomy and repair
21	Novak and Molnar 2005 [22]	33/F	No	CH	5 years	Anterior skull base and tegmen tympani	Craniotomy and repair
22	Honeybul and Bala 2006 [2]	19/M	No	AS	2 months	Frontal sinus	Craniotomy and repair
23	Najera Aguilar et al. 2014 [23]	65/M	No	CH (Post SAH)	1 year	Not known	Aspiration and anti-siphon device
24	Najera et al. 2014 [23]	64/M	No	CH (Post meningitis)	5 years	Frontal sinus	Craniotomy and repair with anti-siphon device
25	Salem-Memou et al. 2016 [24]	60/M	No	IV tumor	2 months	Petrous bone	Craniotomy and repair

Table 1 (continued)

S No	Author/reported year	Age/gender	NF1	Indication of VP shunt	Duration b/w shunt and pneumocephalus	Site of bony defect	Management
26	Martinez-Perez et al. 2017 [25]	75/M	No	NPH	2 months	Petrous temporal bone	Craniotomy and repair
27	Verhaeghe et al. 2018 [26]	81/M	No	NPH	10 months	Petrous bone	Craniotomy and repair
28	Rodriguez et al. 2020 [27]	81/M	No	NPH	2 months	Petrous bone	Craniotomy and repair
29	Orlando et al. 2022 [28]	28/M	No	Cerebellar AVM	8 years	Anterior skull base	Endoscopic repair
30	Present case	21/F	Yes	AS	10 months	Frontal sinus	SOKHA- encephalocele repair, opening LT, removal of VP shunt
Summary		Age = M: F = 1.7:1	Percent of cases with NF1 = 4/30 (13.3%)	AS = 13 CH = 7 NPH = 3 Mass lesions (VS, IV tumors, AVM, PFT) = 7	Mean duration of presentation post CSF diversion = 2.06 years	Frontal sinus = 9 Cribriform plate = 2 Ethmoid sinus = 2 Petrous bone/ mastoid air cells = 9 Sellar = 2 Not specific = 6	

F female, *M* male, *AS* aqueductal stenosis, *SAH* subarachnoid haemorrhage, *PFT* posterior fossa tumor, *na* not available, *VS* vestibular schwannoma, *IV* intraventricular, *AVM* arterio-venous malformation, *CH* communicating hydrocephalus, *SAH* subarachnoid hemorrhage, *NPH* Normal pressure hydrocephalus

poor visualization of these defects as they are not associated with CSF leaks [2]. To the best of our knowledge, this is the first report of repair of frontal encephalocele via a minimally invasive SOKHA. This approach has the advantage of simultaneously tackling both dural defect and hydrocephalus via a single small craniotomy. One limitation of this approach is the inability to do a water-tight primary dural closure. However, a proper delineation of dural defect, coagulation of herniated brain matter, cranialization of the frontal sinus, obliteration of dead space by fat, and a vascularized pericranial graft are the most important steps while dealing with such defects.

What to do after repair of dural defect?

How to deal with the shunt once pneumocephalus sets in is a debatable question. Methods that have been used in past cases include:

1. Direct repair of dural defect with no manipulation of the shunt [5]
2. Ligation of the shunt at the neck with the repair of the dural defect and insertion of an anti-siphon device after a period of 2–3 weeks (2)
3. Repair of defect with endoscopic third ventriculostomy with shunt removal and re-insertion in case ETV fails
4. Repair of defect with the opening of lamina terminalis with shunt removal (current case)

Conclusion

NF1 can result in early erosion of the skull base in the presence of chronic hydrocephalus. Such erosions should be looked for in preoperative imaging before ventriculoperitoneal shunt to avoid tension pneumocephalus. SOKHA with the opening of LT is an effective minimally invasive surgical strategy.

Author contribution All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by Kavindra Singh and Ashutosh Kumar. The first draft of the manuscript was written by Kavindra Singh and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Data availability All data supporting the findings of this study are available within the paper.

Declarations

Consent to participate Informed consent was taken from the patient and her parents for this publication.

Consent for publication The participant and her parents have consented to the submission of the case report to the journal.

Conflict of interest The authors have no competing interests to declare that are relevant to the content of this article.

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