First Annual Cedars-Sinai Intracranial Hypotension Symposium - October 14, 2017

Serious Complications of Intracranial Hypotension



WOUTER I. SCHIEVINK, M.D. Professor of Neurosurgery Department of Neurosurgery, Cedars-Sinai Medical Center Los Angeles, California

CSF leak program, Cedars-Sinai Medical Center, Los Angeles, CA, USA

Neurosurgery 8

Wouter I. Schievink, M.D.

Neuroradiology

M. Marcel Maya, M.D.

Franklin G. Moser, M.D., M.M.M.

Headache Medicine \odot

Steven Graff-Radford, D.D.S.

Ronald Andiman, M.D

Anesthesiology 8

> Charles Louy, M.D., Ph.D. Howard Rosner, M.D.















Clinical Manifestations

- 90 100% Headache
- 50 90% Neck Pain

Hearing abnormalities

- 25 50%
 - Nausea / Vomiting
 - Light/noise sensitivity
- 1 25%
 - Diplopia (CN VI or III)
 - Cognitive decline
 - Behavioral variant fronto-temporal dementia
 - Myelopathy/radiculopathy
 - Tremors/Parkinsonism/ataxia
 - Coma

Serious complications of spontaneous intracranial hypotension

Coma

Se Frontotemporal Dementia

- Superficial Siderosis
- Bibrachial amyotrophy
- Idiopathic Spinal Cord Herniation
- Diffuse Non-aneurysmal Subarachnoid Hemorrhage

Serious complications of spontaneous intracranial hypotension

Coma:

Any CSF leak type

Se Frontotemporal Dementia:

No CSF leak identifiable

- Superficial Siderosis
- Bibrachial amyotrophy
- Idiopathic Spinal Cord Herniation
- Diffuse Non-aneurysmal SAH

Type 1a ventral CSF leak

Coma in Spontaneous Intracranial Hypotension

- Service Frequency: 1.5%
- ✤ N=16
- Age: 56.2 years (34-72 years)
- More common: Men

Older

Residents of Los Angeles/California

Require surgery



COMA and brain sagging



COMA and Duret's hemorrhage and brain stem edema

4 Pathways to Coma in Spontaneous Intracranial Hypotension

A: Coma develops after onset of SIH prior to treatment

B: Coma develops after onset of SIH following treatment

 C: Coma develops <24-48 hours of evacuation of subdural hematomas due to SIH

D: Coma develops <24-48 hours of craniotomy for unrelated pathology

Don't let this happen to your patient















Post-op









Outcome of coma due to SIH

15 patients

Glasgow coma scale score:	8	n=5
	7	n=3
	6	n=5
	5	n=2
Glasgow outcome scale score:	5	n=13
	4	n=1
	3	n=1*

Behavioral-Variant Frontotemporal Dementia in Spontaneous Intracranial Hypotension

Older / Men

- Service Frequency: 3.5%
- ✤ N=33
- 10 women/23 men
- Age: 52.9 years (37-65 years)
- More common:

Brain sagging Non-Residents of Los Angeles Absence of extra-dural CSF* Require surgery

Behavioral variant Frontotemporal Dementia

- Second most common cause of dementia in those <60 years</p>
- Genetically determined
- Link with ALS
- Progressive deterioration in social functioning and personality
- Untreatable

Behavioral variant Frontotemporal Dementia in SIH

- A: Behavioral disinhibition
- B: Apathy or inertia
- C: Loss of sympathy or empathy
- D: Compulsive/ritualistic behavior
- E: Hyperorality
- F: Neuropsychological profile of executive deficits



bvFTD in SIH





Frontotemporal dementia in spontaneous intracranial hypotension



Symptoms of bvFTD in SIH





bvFTD and brain sagging I



bvFTD and brain sagging II



Temporal lobe herniation



Cause of intracranial hypotension is

• Spinal CSF leak

Pooling of CSF

- Inadequate CSF production?
- Rapid CSF absorption?

NEVER

• ? CSF rhinorrhea–otorrhoea ?

Lack of causal association between spontaneous intracranial hypotension and cranial cerebrospinal fluid leaks

Clinical article

Wouter I. Schievink, M.D.,¹ Marc S. Schwartz, M.D.,^{1,2} M. Marcel Maya, M.D.,³ Franklin G. Moser, M.D., M.M.M.,³ and Todd D. Rozen, M.D.⁴

Departments of ¹Neurosurgery and ³Radiology, Cedars-Sinai Medical Center; ²House Clinic, Los Angeles, California; and ⁴Department of Neurology, Geisinger Specialty Clinic, Wilkes-Barre, Pennsylvania

Evolution of brain sagging in bvFTD



2007 – headache only



2008 – bvFTD





2014



2

0

2

2015 - postop

2017 - lawsuit

Pre- and post surgery PET in bvFTD



Outcome of behavioral-variant FTD in SIH



Frontotemporal dementia in spontaneous intracranial hypotension

Solution (SIHDAS I or II) after treatment

1 / 7 patients who had undergone Chiari decompression (14%)

VS

20 / 22 patients who had not undergone Chiari decompression (91%)

P=0.003

Superficial Siderosis in Spontaneous Intracranial Hypotension

- Frequency: 5% (usually asymptomatic)
- ✤ N=46
- 27 women/19 men
- Age: 52.2 years (34-82 years)
- More common: Ventral type 1a CSF leak *

Long symptom duration

Superficial Siderosis

- Diffuse hemosiderin deposits in CNS
- About one-third have underlying type 1a ventral CSF leak
- Hearing loss and ataxia

MRI in Superficial Siderosis due to SIH



Intraspinal hemorrhage – ventral spinal CSF leak

















Idiopathic Spinal Cord Herniation in Spontaneous Intracranial Hypotension

- ✤ Frequency: 1%
- ℬ N=8
- Age: 45.2 years (34-52 years)
- More common: Ventral type 1a CSF leak

Long symptom duration

Spontaneous spinal cord herniation



Ventral spinal CSF leak – delayed sequelae (spinal cord herniation)







Bibrachial Amyotrophy in Spontaneous Intracranial Hypotension

- Frequency: 0.5%
- ℬ N=6
- All men
- Age: 45.2 years (24-52 years)
- More common: Ventral type 1a CSF leak

Long symptom duration

Ventral spinal CSF leak – delayed sequelae (Bibrachial amyotrophy or ALS-like syndrome)



Bibrachial amyotrophy in SIH





Post-op



22 year old with bibrachial amyotrophy





Post-op brain MRI



Pre-op





Cedars: 1/17



2/17



3/17



5/17

Non-aneurysmal Diffuse SAH in Spontaneous Intracranial Hypotension

- ✤ Frequency: 0%
- ✤ N=2
- Both women
- Age: 56.2 years (51-78 years)
- More common: ?

Ventral spinal CSF leaks – delayed sequelae (non-aneurysmal SAH?)



Thank you CSFLEAK

Save the Date! Saturday October 14, 2017 13 2018 Intracranial Hypotension Symposium Spinal csf leak