A Series of Unfortunate Events: Bilateral Thalamic Injury

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Disclosures

I keep 6 honest serving-men
(They taught me all I knew);
Their names are What and Why and
When
And How and Where and Who

Rudyard Kipling
Outline

• Case history
• AVM and imaging
• Thalamic vasculature
• Bilateral thalamic injury
• Function and structure of thalamic nuclei
• Case outcome
• Treatment of bi-thalamic injury syndromes
65y M
Business owner, married

PMH

- Hypertension, GERD, DDD, retinal nevus, renal calculi, elevated creatine, BPH
Cerebellar AVM

Timeline:
April 2014: Severe occipital headache at home
- Local hospital imaging: subarachnoid cerebellar and 4th intraventricular hemorrhages
- Criticall Hospital: Angiogram: cerebellar vermis AVM

http://www.medfriendly.com/cerebellar-vermis.html
A Glimpse Ahead...

Sequelae

- Quadriplegia
- Severe neuropathic (?) pain
- Dystonia
- Depression
- Cognitive impairment
Congenital Anomaly

July 2014: Cerebral angiogram at UHN: Superior cerebellar vermis AVM

- drained to vein of Galen
- vein of Galen terminated at straight sinus
- retrograde flow to deep system and bilateral cerebral veins
- Evidence of venous hypertension in midline thalamic region usually drained by the deep system
The straight sinus ends in the back-end part of the sagittal superior sinus. In this area, this sinus fuses in the confluence of the sinuses also called “Torcular Herophilus”

Arteriovenous Malformations (AVM)

- 0.1% general population
- 3% of all strokes
- 9% of all subarachnoid hemorrhages
- Sporadic congenital malformations
- Direct arterial to venous connections without intervening capillary networks
- Arterial supply and venous drainage can be single or multiple vessels
- High flows can lead to aneurysm development
- Annual hemorrhage rate of 3%
Cerebellar AVMs

- <15% of all cerebral AVMs

Recent retrospective review of 500 AVMs at USCF

- 60 cerebellar AVMs (12%):
  - 30% were vermian (most common)
  - >93% of vermian AVMs presented as hemorrhage
  - Vermian AVMs – poorest outcomes
  - Avg. age=41 at presentation

Rodríguez-Hernández et al., 2012
Diencephalon

http://www.highlands.edu/academics/divisions/scipe/biology/faculty/harnden/2121/notes/cns.htm
Dorsal thalamus
AKA “thalamus”

• Paired bodies
• 3 x 1.5cm
• Bulk of diencephalon
• Lateral border of 3rd ventricle
• Bordered by internal capsule laterally
• Joined by massa intermedia (runs through 3rd ventricle)
• Divided into 3 larger groups of nuclei by the “internal medullary lamina”
• Plus the caudal ‘pulvinar’

Linn et al., 2007
Fact and Folly
AVM Embolization

Image 1/3: Post embolization moving up from cerebellum
Thalamic drainage

Upper and middle thalamus
  – Internal cerebral vein

Lower and posterior
  – Vein of Rosenthal (basal vein) or posterior mesencephalic veins

Central and upper
  – Superior thalamic vein (largest thalamic vein)
    - this runs parallel to the internal cerebral vein and joins it or the Vein of Galen

http://neuroangio.org/venous-brain-anatomy/deep-venous-system/
2. Straight sinus
3. Internal cerebral vein
4. Vein of Galen
5. Basal vein (Vein of Rosenthal)
6. Thalamostriate veins
7. Transverse sinus
Physical exam findings April 2015

- Normal cranial nerves II-XI
  - Normal swallowing, DAT, no respiratory difficulty
- Reflexes: 1+ upper
- Lower limbs: too painful to test
- Increased tone in all limbs and rigidity
- Yelling out in pain with any sensory stimulus
- Limited R. sided active motor ability - inconsistent
  - Some automatic movements, but inconsistent
  - Wiggles R. fingers, toes and slight movement of R. ankle
• Sensory exam
  – Pain with any intervention
  – Trunk and upper extremity: diminished light touch and pinprick
  – Lower extremity: no pinprick or light touch
  – Proprioception intact to great toes with large movement only
Rigid Quadriplegia

Dystonic rigidity:

- Neck forward flexion and ‘anterocollis’ to the left ‘laterocollis’ + ‘torticollis’ to the right
  - Related to left sternocleidomastoid hyperactivity
  - Right SCM hypertonic and overstretched
- R. elbow flexion: 70 degrees
- L. elbow flexion: 100 degrees
- L. wrist flexion: 30 degrees
- Shoulders internally rotated, hands resting rigidly against body
- L. knee flexion: 50 degrees
Cognitive assessments

• Oriented to ‘hospital’, not specific name or date
• Pt did not reply unless personally relevant questions asked
• Closed eyes shortly after responding, even without significant pain medication
• Evidence of decreased thought processing and decreased short term memory
• Some evidence of decreased long term memory
• Behavioural issues began to emerge:
  – Inhibition: profanity, sexual remarks, suspicion, confabulation
  – Irritability
Treatment

• Trials of
  – DANTROLENE SODIUM, TIZANIDINE and BACLOFEN
• Botox
• Glutathione, curcumin
• Lorazepam, opiates pre-intervention
• Physiotherapy
  – Better ROM with upright position on tilt table
  – Pain limited
• Occupational therapy
  – Splinting not possible due to severe flexion and pain
Bilateral thalamic injuries

Wernicke’s Encephalopathy  Deep Cerebral thrombosis  Basilar tip thrombosis

http://qjmed.oxfordjournals.org/content/103/11/891
http://radiopaedia.org/articles/deep-cerebral-vein-thrombosis
http://radiopaedia.org/articles/top-of-the-basilar-syndrome
Under the Hood
Figures 2a to 2f. Imaging anatomy of the thalamus. Each thalamic body is divided by Y-shaped bands of white matter (internal medullary lamina, arrowheads) into three larger nuclear groups: the anterior nuclei, which lie in the fork of the Y (asterisk), the ventrolateral nuclei, situated laterally (thick arrow), and the medial nuclei, lying medially (curved arrow). The pulvinar joins the thalamic bodies on the dorsocaudal side (thin arrow).

a, b) Transverse (a) and coronal (b) T1-FLAIR-weighted ST-MRIs of an adult test person. c-f) Transverse T2- (c) and T1-weighted (d) and coronal (e, f) T2-weighted images of an infant with cistic degeneration of the gray matter of the thalamus. Only the medullary lamina are preserved and well.
Thalamic Nuclei

Nuclei of the Thalamus

ROSTRAL

CAUDAL

Inferior colliculus
Auditory area
Optic tract
Visual area

http://homepage.ntlworld.com/teversal/myweb/CNS/thalamus.htm
Major relay to prefrontal cortex
• *Integration of information with mood, emotion, olfactory*

Projects to the cingulate gyrus
• *attention, memory storage*

Receives nociceptive spinothalamic fibres
• RAS

Sensory Areas 3, 1, 2

Motor Areas 4, 6, 8

Nuclei of the Thalamus

Pulvinar

“Pulverizer”

• Association areas from temporal, parietal, occipital
• Attention and neglect
• Behavioural orientation to stimuli
Thalamic function in the News

Bilateral anoxic thalamic injury common in vegetative state (VS) coma

Lobotomy related damage
Rosemary Kennedy
1918 – 2005
Central Pain Syndrome

• Déjerine and Roussy syndrome – obsolete term
• Central, neuropathic disorder involving the spinothalamic pathway
• Etiology within the thalamus: Ventroposterior nuclei

http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2659949/
Madame Jossaume

• Dejerine and Roussy, 1911

“...among the most spectacular, distress, and intractable of pain syndromes”
Madame Jossaume

“Fifteen days later, the paralysis had receded to be replaced by severe and unrelenting pain over the entire left side. This pain was described as ‘burning and crawling’, particularly over the left temple, ear, nostril, and tongue.

Three months later, Madame Jossaume was taken to see Jules Déjerine, a well-known Parisian neurologist. Déjerine’s thorough examination revealed a complete absence of touch sensibility in the left hand and forearm, hypoesthesia (decreased sensitivity to stimulation) of the left arm, and mirrored signs in the left foot and thigh. The left side of her face was similarly insensible to touch, while the mucous membranes of the left mouth, palate, pharynx, tongue, cornea and conjunctiva all demonstrated hypoesthesia. ...

Remarkably, the patient displayed a complete absence of proprioception (awareness of body position) in her left arm. A similar, albeit less severe phenomenon was observed in the leg.
...And above all, hope

DBS for intractable pain (Bittar et al., 2005; Nandi and Aziz, 2004)

Schiff et al, 2007  [http://www.nature.com/nature/journal/v448/n7153/abs/nature06041.html](http://www.nature.com/nature/journal/v448/n7153/abs/nature06041.html)

References

• Pearl, Gregg, Gandhi (2011). Cerebral Venous Development in Relation to Developmental Venous Anomalies and Vein of Galen Aneurysmal Malformations. Seminars in Ultrasound, CT, and MRI, 32(3):252-263.


