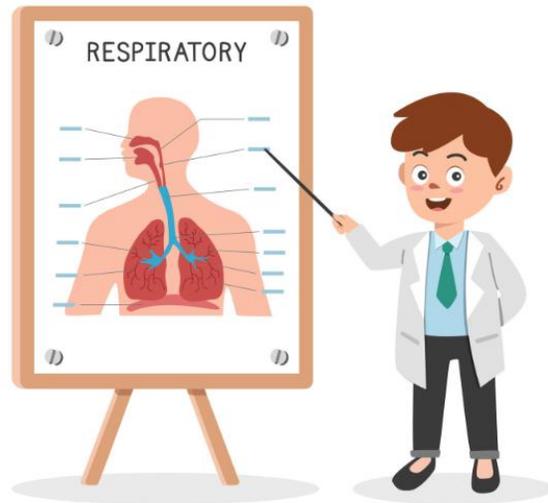


An airway obstruction with an unexpected turn



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TRANSPORT CALL...

“ Can you come for a term neonate with an obstructive airway? ”

Pregnancy uneventful, normal ultrasounds

Good start, but increasingly obstructive breathing

Difficult feeding

CPAP has no effect



CLINICAL EXAMINATION

Parameters:

- Sat 98%
- RR 76/min
- HR 135 bpm



Obstructive airway, better with chin lift → bulging faryngeal mass at laryngoscopy

Normal general clinical examination

No dysmorphic features

WHO YOU GONNA CALL??

Otorhinolaryngologists

WHAT SHOULD WE THINK OF?

DERMOÏD CYST

TERATOMA

GLIOMA

VALLECULAR CYST

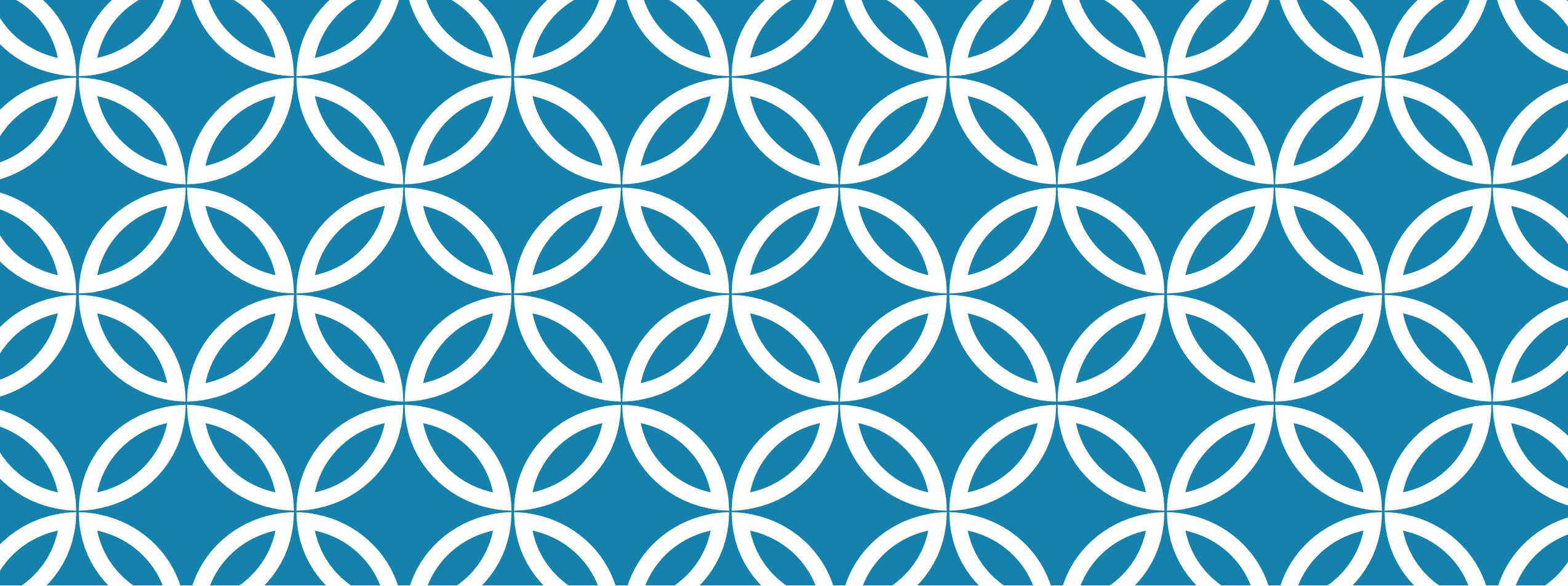
ENCEPHALOCELE

HEMANGIOMA

LYMPHANGIOMA

NEUROBLASTOMA

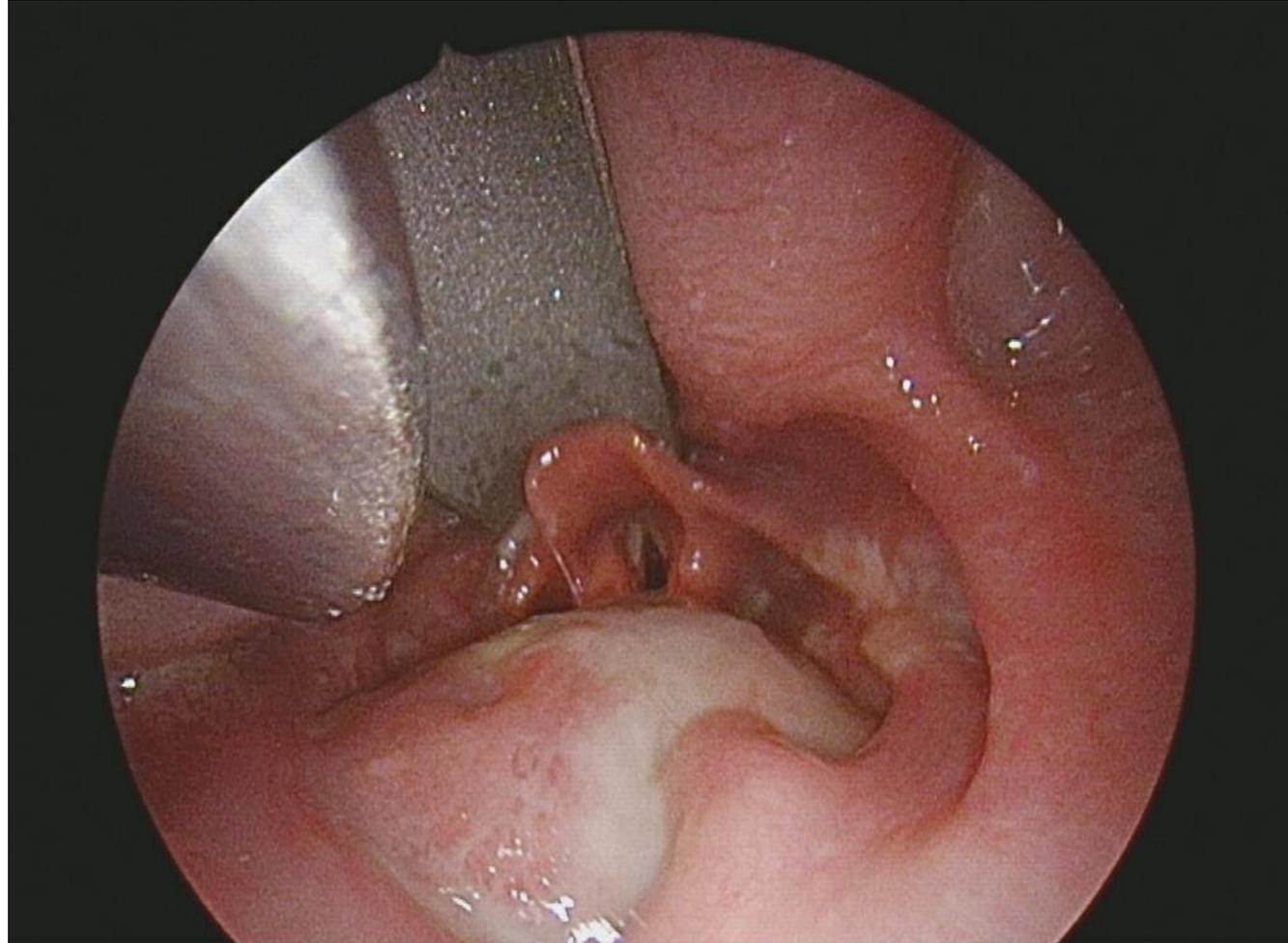
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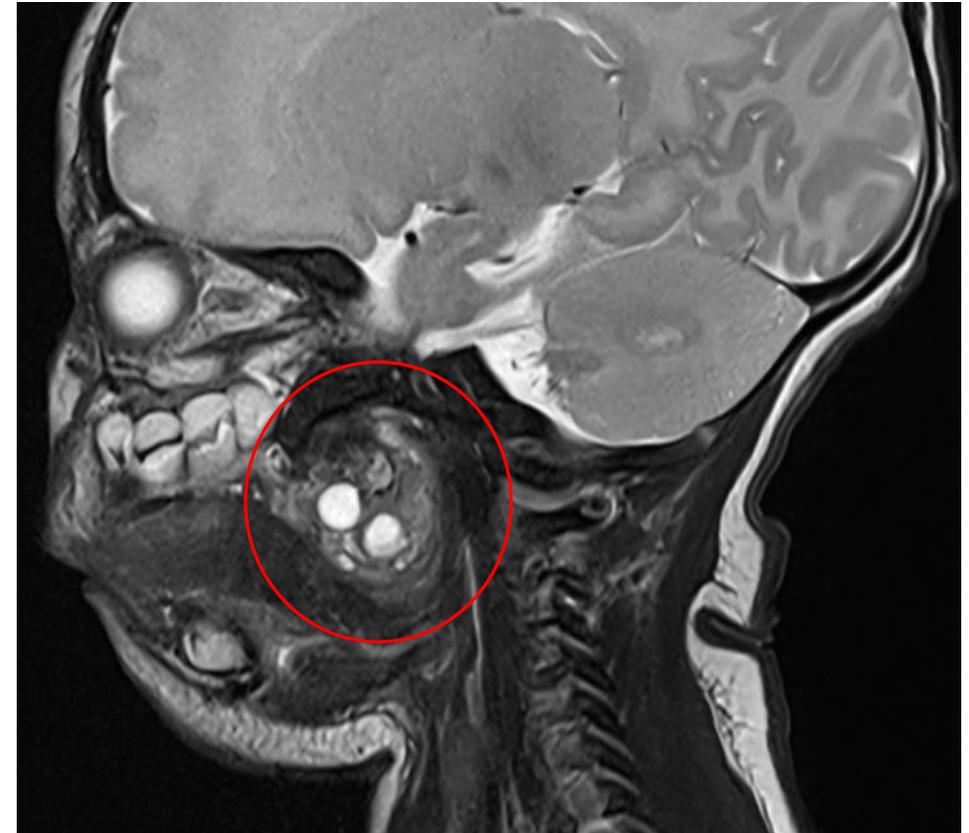
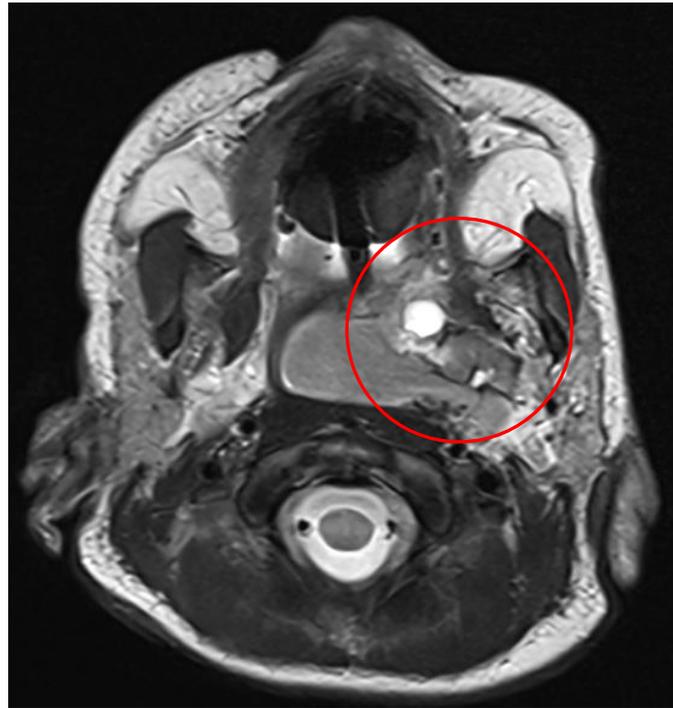
**SO WE START LOOKING FOR
ANSWERS...**



OPERATING ROOM — DIRECT LARYNGOSCOPY



MRI



MRI

*“**Heterogeneous mass** with overlying fat as well as **diffuse non-solid contents**, centered in the nasopharynx with extension into the surrounding tissues: image primarily consistent with **surinfected teratoma.**”*

Significant mass effect with narrowing of upper airway.

BACK TO THE OPERATING ROOM

Debulking of the mass → multicystic aspect, no pus

- Smooth extubation a couple of hours later, no more need for respiratory support
- Started on antibiotics, but culture showed only commensals
- Still difficult feeding with need for tube feeding, pain?



1 WEEK LATER - ANATOMOPATHOLOGY

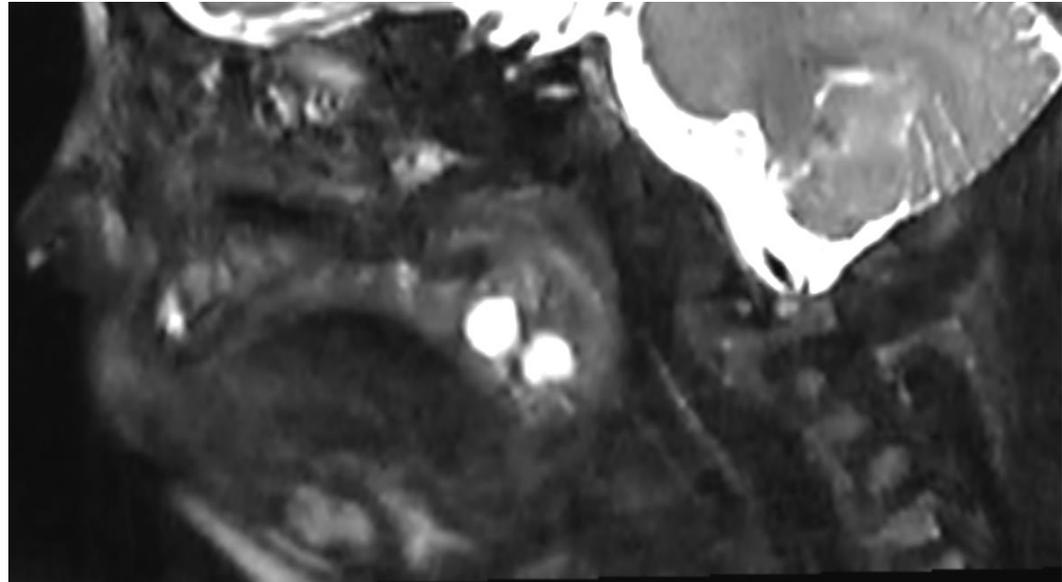
*“Ectopic brain tissue with meninges, covered with fibrinopurulent debris, with dysplastic but somewhat age-matured cortical structure highly consistent with **meningoencephalocele.**”*

Look for a connection!



BACK TO THE MRI + CT

No connection of the mass with
intra-cranial structures



EVOLUTION

- Sporadic short desaturations, min 80%
- Problematic feeding
- Very unhappy baby
 - Pain?
 - Cramps? Reflux?
 - Neurological behaviour?
 - Respiratory discomfort?
 - Social needs? Comfort and attention?
- Impression of increasingly obstructive airway



POLYSOMNOGRAPHY

Severe OSAS

→ nasopharyngeal CPAP

→ second maximal debulking procedure

Anatomopathology

- Mature CNS tissue: cortex, white matter, ventricle with plexus, meningeal tissue
- Ectopic embryonal remnant



DIAGNOSIS

Definitive diagnosis: **meningoencephalocele without residual connection**

Positive evolution since then, discharge home at age 1,5 months.



BACKGROUND

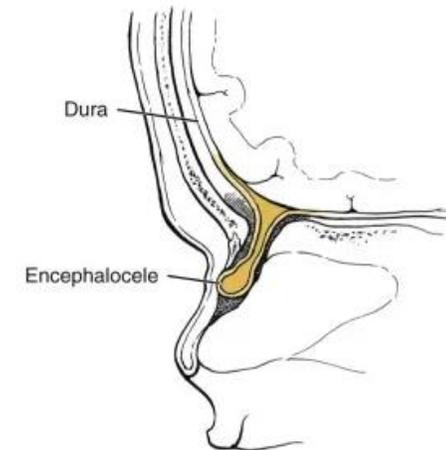
Nasopharyngeal meningoencephalocele = type of neural tube defect with herniation of cranial contents into the nasopharynx through a skull base defect

In this case → most likely transsphenoidal through a patent craniopharyngeal canal

Incidence 1/35,000

Symptoms

- Nasal obstruction
- Clear nasal fluid (liquor)
- Meningitis
- Headache



BACKGROUND

Imaging: high-resolution CT and/or MRI

Histopathology: mature neuroglial tissue, meninges

Surgical repair

- Resection of the meningoencephalocele
- (Multilayer) closure of the bony defect

TAKE HOME MESSAGES

1. Thinking before cutting
2. Adequate imaging
3. Connection of meningoencephalocele can obliterate



THANK YOU FOR YOUR SUGGESTIONS!

