• 4 year old girl

• Permanent rhinorrhea, watery eyes, extensive snoring and exclusive mouth breathing

• Hypertrophic polyps and purulent conjunctivitis repeatedly diagnosed

• Half a year later a flattened nasal pyramid appeared and imaging studies were initiated
CT scan and MRI imaging:

- Complete congestion of the nasal cavity
- Compression of the left orbita
- Infiltration of the anterior cranial fossa

→ Local aggressive growth, but focal sclerotic rim present

→ MRI resembling cartilarginous tumor with atypical homogeneous uptake of contrast media
Histology of the biopsy
Histology

• Spindle cell proliferation with quite some nuclear pleomorphism

• Areas of new bone and cartilage formation

• Spread below respiratory mucosa

→ Worrisome «atypia» and osteoid formation → osteosarcoma?
→ consultation of an expert in osteosarcoma
Expert diagnosis: Suggestive of osteosarcoma but «a kind of weird»
Histology of the curetage
Nasal Chondromesenchymal Hamartoma
Two years after diagnosis she is fine with no signs of recurrence
Nasal Chondromesenchymal Hamartoma

• Rare, predominantly pediatric nasal lesion (but can also affect adults!)

• Reminiscent with the chondromesenchymal hamartoma of the chest wall

• Radiology often suggests malignancy: poorly defined, aggressive growth pattern, contrast media uptake and intracranial extention

• Mixture of stromal and chondroid areas (no atypical mitosis!), can be very cystic (DD meningoencephalocele)

• May be associated with a germline DICER1 loss of function mutation (pleuropulmonary blastoma)
Differential diagnosis: Osteosarcoma
Data from our Basle registry for bone tumours: osteosarcoma

- Osteosarcomas from 1976 to date: n=1574
- Localisation:
  - nose, sinus maxillaris, orbita, fossa canina, os zygomaticum and palatinum → 53 cases
- Age: median and average 41 yrs (6 to 92 yrs)
- Distribution: sinus maxillaris (21) > os palatinum (9) > orbita (7)
Age: only one case under 10 years of age

Youngest child was 6 years old: OS in the orbita post radioation therapy after retinoblastoma

Localisation:
only 4 cases in the nose (range: 13-26 yrs)
Three main studies about osteosarcoma in very young children (<5 years)

- Cooperative Osteosarcoma Study Group (COSS): 27 children
  - 26 long bones, 1 orbit (secondary to radiation therapy for retinoblastoma)
- French cancer society: 15 children
  - 14 long bones, 1 rip
- The St. Jude Children's Research Hospital: 8 children
  - 6 long bones, 1 maxilla (low grade), 1 orbita (but from 1976 with no pathology review and no clinical information (secondary?))
Be very cautious with the diagnosis of an osteosarcoma in the nose in a child under the age of 10 years!
When you deal with a lesion composed of spindle cells, bone and cartilage formation in the nose, consider nasal chondromesenchymal hamartoma in your differential diagnosis.
CHONDROMESENCYHYMAL HAMARTOMA
Additional cases from the registry

• Newborn, boy, right nasal cavity, cartilarginous tumor of unknown dignity
• 56 yrs, man, ethmoid bone, sarcoma
• Newborn, boy, chest wall, aneurysmal bone cyst