Malformations of the nose, Clefts & Tumors of the nose

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Content

1. Congenital malformations of the anterior skull base
2. Nasal malformation in the context of cleft lip and palate
3. Chonial atresia
4. Tumors of the paranasal sinuses
Learning objectives in congenital malformation of the anterior skull base

• Distinguish: skin ectodermal or neuro-ectodermal

• Imaging: always CT (without contrast) + MRI

• Skin ectodermal:
  - Dermoid
  - Nasal dermal sinus cyst
  - always extradural
  - on / in / under the nasal bone
  - variable length of proximal fibrous stalk
  - total excision depending on cosmetics and/or infection

• Neuro-ectodermal:
  - nasal glioma
  - meningocele / meningo encephalocele
  - risk of ascending meningitis
congenital malformation of the anterior skull base

Embryology

• 8th – 9th week of gestation

• Frontal lobe gets separated from the skin by the growing frontal bone

• Different theories
  - remaining skin ectodermal or neuroectodermal cells
  - fusion of skin with neuro-ectoderm in the midline
    - either skin tears neuroectoderm → meningocoele...
    - or neuro-ectoderm tears skin → NDSC, dermoid ...

Sessions RB, Laryngoscope 1982
Skin ectodermal

Nasal dermal sinus cyst (NDSC)

Clinical findings

• Nasal pit in the midline (nasal dorsum ♦ columella) with (black) solitary hair

• Cystic tumor on nasal dorsum (hump)

• Discharge out of pit

• Infections
Skin ectodermal

Nasal dermal sinus cyst (NDSC)

Imaging findings

Fistula (sinus) can be followed from the pit above, within underneath the nasal bone
Skin ectodermal

Nasal dermal sinus cyst (NDSC)

Imaging finding

- Enlarged foramen coecum
- Bifid crista galli

Skin ectodermal Dermoid

Clinical findings

• Midline or paramedian
• Subcutaneous tumor
• Recurrent infections
Skin ectodermal Dermoid

Imaging findings

- Hypodense well defined tumor
- Without bony dehiscence
- No intracranial component (F. coecum, crista galli)

Holzmann D Eur Radiol 2004
Skin ectodermal

NDSC & Dermoid

Treatment

• Radical resection

• Approach depends on location and extension (‡ imaging is essential!)

Holzmann D Rhinology 2007, Holzmann D 2013
Neuro-ectodermal

Meningocele / Meningoencephalocele

Clinical findings

• (Unilateral) nasal obstruction
• „solitary Polyp“
• Ev. watery discharge (CSF-Leak)
• meningitis
Neuro-ectodermal

Meningocele / Meningoencephalocele

Imaging findings

• CT: bony dehiscence

• MRI: T2-hyperintense (liquor isointense) tumor
Neuro-ectodermal

Nasal Glioma
Learning objectives in nasal malformation in the context of cleft lip and palate

- Cleft types: unilateral / bilateral // lip only, palate only / partial or total
- Nasal involvement only in “anterior cleft“ (lip and alveolar process)
- Complex malformation requires detailed analysis
  - Septum
  - maxillary crest
  - nasal vestibulum
  - medial and lateral crus of the upper lateral cartilage
- Treatment
  - timing: interdisciplinary approach / respecting growth spurt
  - function (nasal obstruction)
  - aesthetic aspects (bring more symmetry)
  - consider the entire midface (upper jaw!)
nasal malformation in the context of cleft lip and palate

Cleft types

• Nasal deformities only in “anterior“ clefts
nasal malformation in the context of cleft lip and palate

If so always complex deformity

• nasal septum
  - caudal septum

• Outer cartilaginous nasal skeleton
  - lower lateral cartilage

• Skin deficits in the inner lining

• Nasal floor upper jaw
  - bony dehiscence
nasal malformation in the context of cleft lip and palate

Cleft types

complex deformity of the ala

- Lateral crus (cartilage)
- Skin deficit of the inner lining
Midfacial growth deficit † under rotated tip

Retreated upper jaw

After Le Fort I Osteotomy
Learning objectives in Choanalatresia

• Unilateral / bilateral

• bilateral
  - Emergency in the newborn ❌ secure airway
  - syndrome associated vs. non syndromic
  - Dg: auscultation (no airflow) or smallest feeding tube
  - Rx: to exclude other malformation of the central skull base
  - Tx: transanosal surgery

• unilateral
  - never emergency !
  - Dg: see bilateral
  - Rx: to include sphenoid in surgical treatment
  - Tx: transanosal surgery (no stenting)
Choanal atresia

- Unilateral / bilateral
- Syndromic e.g. CHARGE
  C Coloboma
  H Heart defects
  A choanal Atresia
  R growth Retardation
  G Genital anomalies
  E Ear malformation

- Non syndrome associated
Unilateral choanal atresia

Sy: Nasal obstruction in newborns and children, unilateral discharge

Dg: lack of breath sounds, no passage of feeding tube, nasal endoscopy

Rx: only if surgery is attempted

Th: transnasal endoscopic surgery

no stent!

Holzmann D J Laryngol Otol 2002
bilateral choanal atresia

Sy: Neonatal respiratory distress + asphyxia!
Emergency!!

Dg: lack of breath sounds, no passage of feeding tube, nasai endoscopy

Th: secure nasal airway !!
Transnasal surgery
(stent ??)
Learning objectives in Paranasal sinus tumors

• Benign tumors
  - Inverted Papilloma ‡ malignant transformation (?)
  - Arteriovenous malformation (Haemangioma)
  - Angiofibroma

• Malignant tumors
  - Rare tumors / different entities
  - Prognosis worsening ‡ dural and/or orbital involvement
  - Dg: clinical findings + biopsy
  - Rx: always CT + MRI
  - Tx: transanasal surgery (craniofacial resection) + IMRT
  - Follow-up: depending on diagnosis (life long in most)

www.WHO.int/publications (free download or purchase as hard copy)

European Rhinologic Society
Supplementum 22
European Position Paper on Endoscopic Management of Tumors of The Nose, Paranasal Sinuses and Skull Base
www.rhinologyjournal.com (free download)
Sinunasal Papilloma

Note: originating from the Schneiderian Membrane (Schneiderian Papilloma)

Class.
- inverted
- oncocyctic (columnar cell)
- exophytic

Dx: clinically „unilateral polyp“
cave: frequently negative biopsy

Th: transnasal surgery

Progn.: recurrence rate < 4%\(^1\)
(external approaches: >20%)  

Holzmann, D J Laryngol Otol 2006
## Sinunasal Papilloma

<table>
<thead>
<tr>
<th></th>
<th>inverted</th>
<th>oncocytic</th>
<th>exophytic</th>
</tr>
</thead>
<tbody>
<tr>
<td>basic</td>
<td>frequently HPV Θ</td>
<td>HPV Θ / &gt;50 J “columnar cell”</td>
<td>more Θ / 20-50 y / HPV Θ 6/11</td>
</tr>
<tr>
<td>Loc.</td>
<td>Lat. nasal wall &amp; ethmoid</td>
<td>Maxillary + ethmoid sinus</td>
<td>Septum</td>
</tr>
<tr>
<td>Rx</td>
<td>Sclerosis = side of attachm. (ca.60%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Malign. transform.</td>
<td>2 (Literature?)</td>
<td>17% Ca at time of diagnosis</td>
<td>rare</td>
</tr>
<tr>
<td>DD</td>
<td>Solitary polyp</td>
<td>Low-grade Adeno-Ca</td>
<td></td>
</tr>
<tr>
<td>Th</td>
<td>Transnasal surgery</td>
<td>Transnasal surgery</td>
<td>Excision</td>
</tr>
<tr>
<td>Recurrence</td>
<td>Mostly after 2-3 y</td>
<td>25-35% (!)</td>
<td>Frequently multilocular !!</td>
</tr>
</tbody>
</table>

Kraft M, Simmen D Kaufmann T, Holzmann, D *Laryngoscope* 2003
inv. papilloma: sclerosis = side origin

exophytic papilloma
TNM

T – primary tumor of maxillary sinus

T1 only in mucosal layer (no bony infiltration or destruction)

T2 bony infiltration of hard palate, middle meatus

T3 Infiltration of maxillary sinus back wall, subcutaneous tissue pterygopalatine foramen, inferior a/o medial orbital wall, ethmoid

T4a Infiltration of anterior orbital space, skin of nose/cheek, pterygoid plate, infratemporal fossa, cribriform plate, sphenoid, frontal sinus

T4b orbital apex, dura, brain, middle fossa, cranial nerves other than V2, nasopharynx, clivus
TNM

T – Primary tumor of the nasal cavity and ethmoid

T1  Tumor limited in one area of the nasal cavity or ethmoid without bony infiltration

T2  Tumor in two areas on one side or invasion into an adjacent region within the nasoethmoidal complex with or without infiltration of bone

T3  Tumor infiltrating inferior or medial wall of orbit, maxillary sinus hard palate or cribriform plate

T4a Infiltration of anterior orbital space, skin of nose/cheek, pterygoid plate, infratemporal fossa, cribriform plate, sphenoid, frontal sinus

T4b Orbital apex, dura, brain, middle fossa, cranial nerves other than V2, nasopharynx, clivus
Despite different in outcome and treatment they have great deal in common

• **Symptoms:**
  - (unilateral) nasal obstruction
  - recurrent epistaxis or bloody discharge (unilateral), hyposmia/anosmia
  - at late stage: double vision, visual impairment, proptosis, facial tenderness

• **Diagnosis**
  - Typical history with leading symptoms
  - nasal endoscopy with suggestive findings
  - **CT (with or without contrast):** mass, bony erosions
  - **MRI:** to check infiltration of cranial fossa dura (or brain) and/or periorbit (or orbit)
  - **Biopsy & diagnostic exploration:**
    - under general anaesthesia with frozen section (reliable material)
    - office based biopsies: not representative material
Despite different in outcome and treatment ‡ they have great deal in common

• Further staging:

  PET-CT (or if available PET-MRI):
  - in all sinonasal Melanoma
  - in all N+

  Ultrasound
  - in any other malignant pathology
Despite different in outcome and treatment they have great deal in common

- **Treatment**: (decision on interdisciplinary tumorboard)
  - Surgery: trend to endonasal >> craniofacial resection
  - IMRT intensity modulated radiotherapy
  - Proton beam therapy
    - young patients
    - tumors close to visual pathway (optic nerve, chiasma opticum)
  - Chemotherapy
    - induction Chemo-Radiation Therapy
    - neoadjuvant
  - Systemic Therapy (Immunotherapy)
Despite different in outcome and treatment they have great deal in common

Follow-up

• Nasal endoscopy every
  1st year: every 6 weeks
  2nd&3rd year: every 3 months
  >3 y every 6 months

• Imaging
  MRI
  1st year: every 3 Months
  > 1 year every 6 – 12 months
  PET-CT/MRI
  - in case of recurrence
  - sinonasal melanoma: part of follow-up imaging
Despite different in outcome and treatment ‡ they have great deal in common

outcome

• Tumor biology

• T-stage
  T3 ‡ significant worsening of prognosis

• More invasive surgery (e.g. orbital exenteration) – does not necessarily result in better outcome
Orbital involvement has an impact on outcome

Adeno-Ca

**FIGURE 4.** Survival distribution by T classification reveals T4 disease to have significantly lower survival ($p < .001$).

**TABLE 1.** T classification for primary sinonasal adenocarcinoma.

<table>
<thead>
<tr>
<th>T classification</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>T1</td>
<td>Tumor is confined to the ethmoid sinus with or without bone erosion.</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor invades 2 subsites in a single region or extends to involve an adjacent region within the nasoethmoidal complex, with or without bony invasion.</td>
</tr>
<tr>
<td>T3</td>
<td>Tumor extends to invade the medial wall or floor of the orbit, maxillary sinus, palate, or cribriform plate.</td>
</tr>
<tr>
<td>T4a</td>
<td>Tumor invades any of the following: anterior orbital contents, skin of nose or cheek, minimal extension to anterior cranial fossa, pterygoid plates, and sphenoid or frontal sinuses.</td>
</tr>
<tr>
<td>T4b</td>
<td>Tumor invades any of the following: orbital apex, dura, brain, middle cranial fossa, and cranial nerves other than (V2), nasopharynx, or clivus.</td>
</tr>
</tbody>
</table>
Tumor specific aspects

**Squamous Cell Carcinoma**

**Etiology:** Ni, chlorine-phenole, thorotrast, smoking
HPV: in malignant inverted papilloma

**Biology:** aggressive

**Loc:**
- a) maxillary sinus (separate TNM-Staging)
- b) Ethmoid

**Progn:**
- 5-year survival:
  - maxillary sinus 42%
  - nasal cavity 60%
Tumor specific aspects

**Sinonasal Undifferentiated Carcinoma SNUC**

**Note:** Frequently metastasis and orbital/intracranial involvement at time of diagnosis

**Biology:** very aggressive !!

**Loc:** maxillary sinus; ethmoid

**Progn:** 5-year survival 20%; median survival: 18 months

Morand GB, Holzmann, D et al *Oral Oncol* 2017
Tumor specific aspects

Intestinal Type Adeno-Ca

Risk factor: Wood and leather dust
(cave: industrial disease ‡ SUVA!)

Patho: 4 subtypes

Epid: 10 – 20 % of all sinonasal malignancies
Loc: between septum and middle turbinate

Progn: local recurrence 50% (!!), LN-metastasis 10%, distant metastasis 20%
5-year survival 40% (depending on subtype)
Life long follow-up

Tumor specific aspects

Adenoidcystic Carcinoma

Epid.: Patients at any age (11 – 92 years)

Etio.: no risk factors

Loc.: Maxillary sinus (60%), nasal cavity (25%), skull base

Progn: bad in general but long living with disease (10-year survival 7%)

long survival with pulmonary metastasis (years)

Note: „once ACC always ACC“
Tumor specific aspects

Non Intestinal Type Adeno-Ca

Risk factor: not known!

Patho. High-grade / Low-grade

Epid: rare
Loc: „everywhere in the paranasal sinuses“

Progn: bad (3-year survival 20%)
Life long follow-up
Tumor specific aspects

Olfactory Neuroblastoma

- Risk factor: not known!
- Etiop/Pg: from olfactory epithelium
- Patho: Hyams grading (correlation with outcome?)
- Epid.: 2-3% of all malignant sinus tumors / age: 2 – 90 years
- Loc.: superior nasal meatus (Assoc. with Jakobson’s canal)
Olfactory Neuroblastoma

Staging: Kadish (5-year survival):

A: limited to nasal cavity (75-91%)
B: in paranasal sinuses (68-71%)
C: intracranial (41-47%)

TNM (Dulguerov)

Progn: overall survival
      5-year: 78% / 10-year: 71% / 15-year: 68%
      low-grade: 80% / high-grade: 40%
Tumor specific aspects
Sinonasal Melanoma

- Risk factor: not known!

- Patho: Mutation: frequently wild type for BRAF, C-KIT, NRAS

- Epid.: increasing incidence (!)
  < 1% of all melanoma / < 5% of all head and neck tumors

- Clin: > 70% no metastasis at diagnosis

- Staging & F.U with PET

- Th: Surgery + RT ev. novel immunotherapy (e.g. anti-PD-1 inhibitor)

Surgical concepts in paranasal sinus & skullbase tumors

transnasal endoscopic approaches:

- transcribriform

- expanded transsphenoidal

- transclival

- transpterygoid
Classification of endoneurosurgical approaches

**transcribriform**
Foramen coecum
→ Planum sphenoidale

**transsphenoidal**
Planum sphenoidale
→ Sella floor

**transclival**
Sella floor
→ odontoid
Thank you for your attention