Adenoid Cystic Carcinoma Minor Salivary Gland Origin

Educational Session

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Content

• Case report
• Imaging
• Primary Therapy – Surgery
• Adjuvant Therapy – Radiotherapy
• Pathology
Case report

• 56y female patient

• Persistent left otalgia
  – No otological explanation

• Airway NAD

• Swallowing NAD
Case Report

- Social history
  - Non-smoker
  - Professional voice user

- Past medical history

- O/E
  - Nasal polyp
  - Clinically no other lymphadenopathy

- Mass incidentally seen left arytenoid

Micro and biopsy performed at another site
BIOPSY

Biphasic neoplasm
  -> epithelial and myoepithelial cells
Cribriform architecture
Nests with lumens containing matrix
Biphasic salivary gland neoplasms

-Adenoid cystic carcinoma
-Epithelial-myoeipithelial carcinoma
Cribriform architecture

Favoured:
ADENOID CYSTIC CARCINOMA
FLUORESCENT IN SITU HYBRIDISATION (FISH)

Approximately 50% of adenoid cystic carcinomas will have a MYB-NFIB translocation
Fish – no myb rearrangement detected
FDG-PET summary

• Primary tumour left arytenoid

• FDG avid positive node left neck

• No evidence of distant disease

• => What next?
FNA left neck level 4 & 5

- Blood and occasional lymphocytes
- No malignant cells identified
FDG-PET and AdCC

• 18F-FDG PET-CT is not helpful to rule out distant metastasis if the primary SGC does not show enhanced FDG uptake

• AdCC with relatively low FDG uptake might be obscured by the normal physiologic FDG uptake of the salivary glands;

• Salivary glands are frequently affected by inflammatory processes wherein increased FDG uptake might result in a false-positive result.

Staging

- As per major salivary gland tumours

- T1 = <2cm, T2 = 2-4cm, T3 >4cm T4a skin, bone T4b SB, pterygoid plates, carotid

- N1, N2a, N2b, N2c, N3

- M0/M1

- T1, N0, M0
Treatment of the primary?

• Principles?
  – Resection
  – Reconstruction

• Options?
Treatment options: ablation

• Primary site
  – Surgery
    • Transoral laser/robotic resection
    • Cordectomy via laryngofissure
    • Partial laryngectomy
      – Vertical
      – Supra-cricoid
    • Total laryngectomy
  – RT/CRT

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Non-surgical primary treatment

- Surgery with RT vs. RT alone
- Combined modality improved local control and survival

- 5 and 10 year local recurrence
  - 56 and 43% RT alone
  - 94 and 91% Surgery and RT

- 5 and 10 year survival
  - 57 and 42% RT alone
  - 77 and 65% Surgery and RT

Treatment of the neck

- Nil
- Surgery
  - Further staging information
- RT
- CRT

Occult nodal disease in approx. 15% elective neck dissection
  improves local control
Patients do not die of nodal disease

Reconstruction options

• Aims: restoration of function
• Options
  – Mucosal free graft
  – False cord advancement/pull-down
  – Bi pedicled muscle/perichondrial flap
  – Skin or muscular free flap
  – Muscular ligamentous/cartilagenous free flap
  – Laser reduction or injection thyroplasty
False cord advancement/pull-down
Bi-pedicled muscle flap
Free tissue transfer
Intra-operative: Ablation
Specimen

- **Left Aryepiglottic Fold**
- **Piriform Fossa**
- **Posterior Cricoid Mucosa**
- **Left Vocal Process**
- **Left Edge of Epiglottis**
- **Anterior False Cord**
- **Anterior Ventricle**
- **Left Lateral Cranarytenoid Mucosa**
- **Cricarytenoid Joint**
Intra-operative: Reconstruction
Post Operative Day 7
LEFT ARYEPIGLOTTIC FOLD

LEFT EDGE OF EPIGLOTTIS

ANTERIOR FALSE CORD

ANTERIOR VENTRICLE

LEFT LATERAL CRICABYRINTHINE MUCOSA

CRICLAYRENGID

POSTERIOR CRICOID MUCOSA

PiriFORM FOSSA

JOINT

LEFT VOCAL CORD

CENTIMETERS
Pale circumscribed lesion

15x6x6MM
-WELL DEMARCATED,
UNENCAPSULATED
-predominantly cribriform architecture
- PERINEURAL INVASION
- EXTENSION INTO SUBMUCOSAL FAT
- Adenoid cystic carcinoma 15mm
- Margins clear (closest-> deep margin= 3mm)
- Perineural invasion present
- No lymphovascular invasion
- Lymph nodes 0/22

- Stage I (pT1N0)
Adenoid cystic carcinoma

- Biphasic tumour- epithelial & myoepithelial cells
- Lumens filled with hyaline or basophilic mucoid material
- Architecture can be cribriform, tubular of solid
  - > 30% solid -> more aggressive course

- 10% of epithelial salivary neoplasms
- Most frequently parotid, submandibular and minor salivary glands
Adenoid cystic carcinoma

- 5 year survival – approximately 35%
- 80-90% die of disease in 10-15 years
- Local recurrence 16-85%
- Lymph node involvement 5-25%
- Distant metastasis 25-55% (bone, brain, liver)
- Perineural invasion is a common feature
- Other prognostic factors include tumour site, clinical stage, bone involvement and status of margins.
Post operative recovery

• Covering tracheostomy and laryngeal stent

• Both removed prior to discharge

• Discharged 3/52

• Tolerating diet and fluids

• Initial degree of aspiration

• Breathy voice good cough
Summary so far

- T1 N0 M0 adenoid cystic Ca supra-glottis
- Completely excised
- Local peri-neural invasion
Patterns of recurrence

• Between 60% - 100% of patients recur at the primary site 30yrs FU

• 10 and 20 year survival: 65% and 28%

• Disease is ‘incurable’ in the long term

• Death is from distant disease
  – Lung mets common site
  – 32 months from diagnosis to death

• Targeted molecular therapies not shown to be of benefit

