

# Acinic Cell Carcinoma with high grade transformation and lung metastasis

Royal Cornwall Hospitals Trust, Treliske

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Thomas Greenslade

Royal Cornwall Hospital

Leonie Wheeldon and Tim Bracey

Diagnostic and Molecular Pathology, Royal Cornwall Hospital

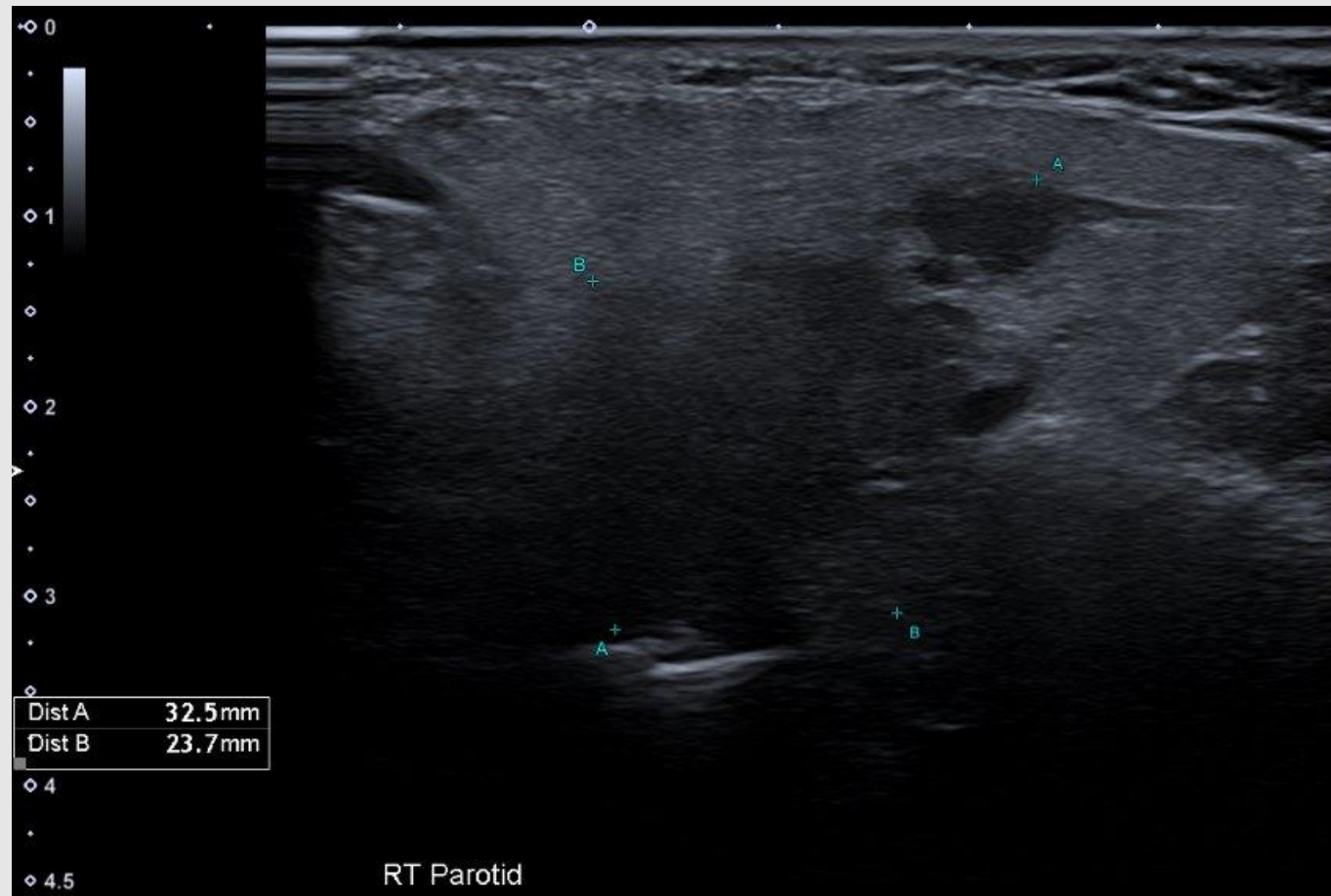
# Clinical presentation

- 69-year-old female presented to ENT with a 4-week history of a rapidly growing right parotid mass associated with pain and new onset right facial nerve palsy (House Brackmann Grade V)
- On examination: firm, tender swelling in the right pre-auricular region extending posteriorly and inferiorly below the angle of the mandible. No overlying skin changes.

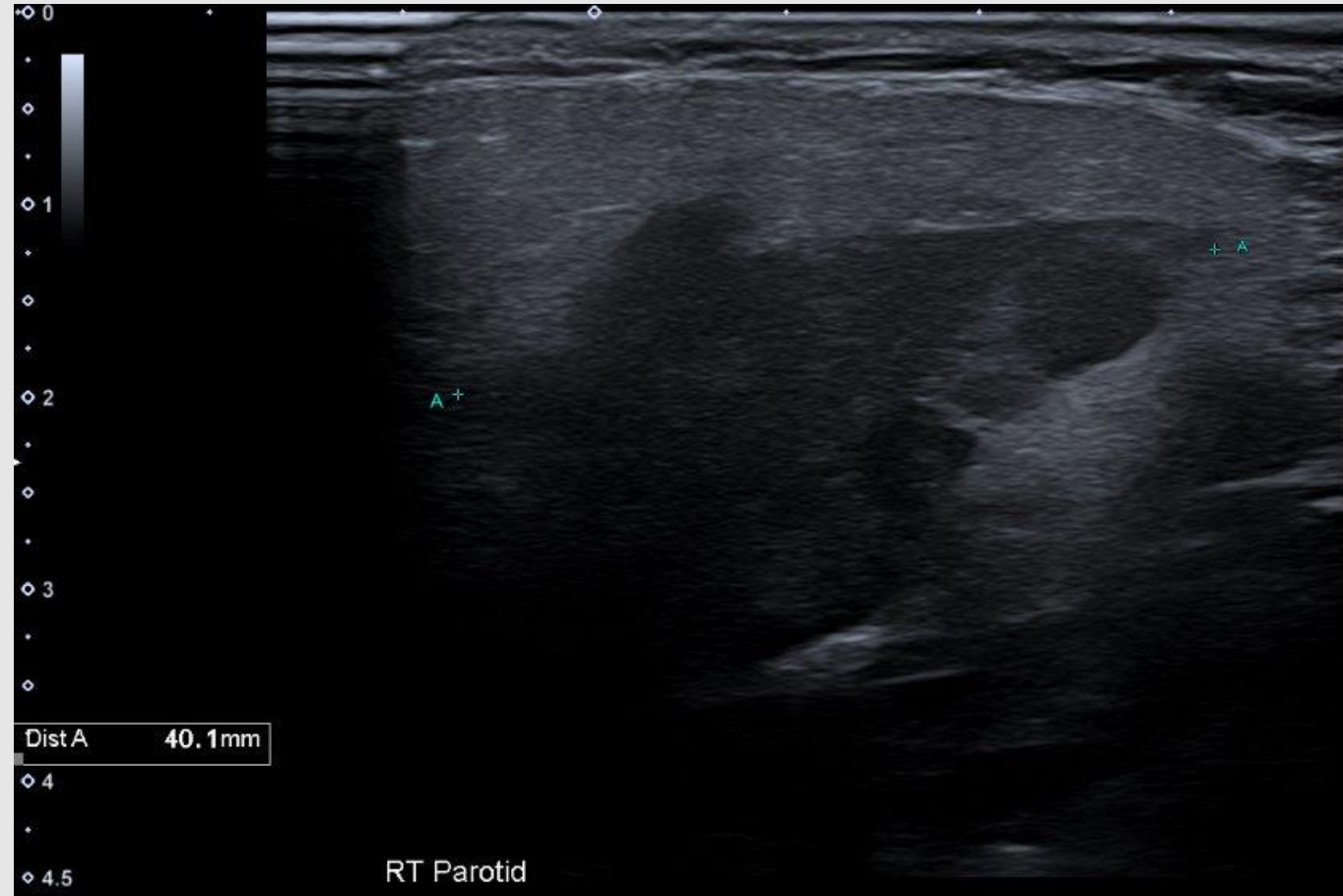
# Ultrasound appearances

- Ill-defined hypoechoic infiltrative process
- At least 33 x 23 x 40mm
- Affecting the superficial and deep lobes of the right parotid gland
- Appearances highly suspicious of a malignant salivary neoplasm
- US-guided FNA performed with ROSE

# Ultrasound image showing maximum height (A) and width (B)

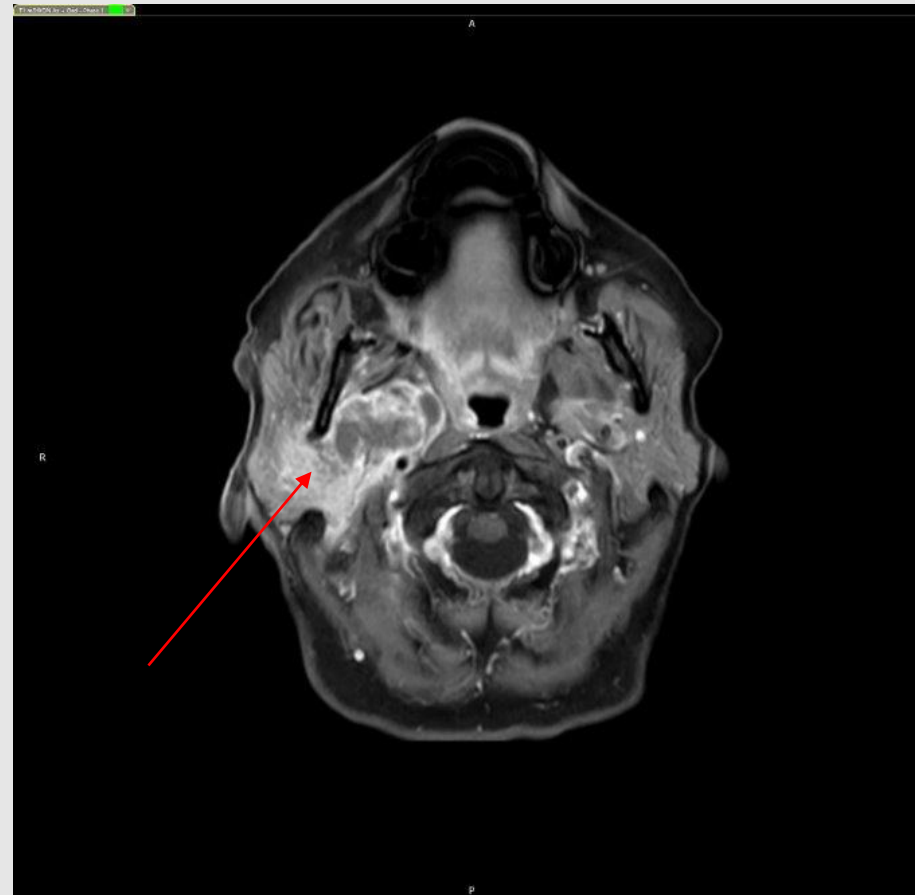


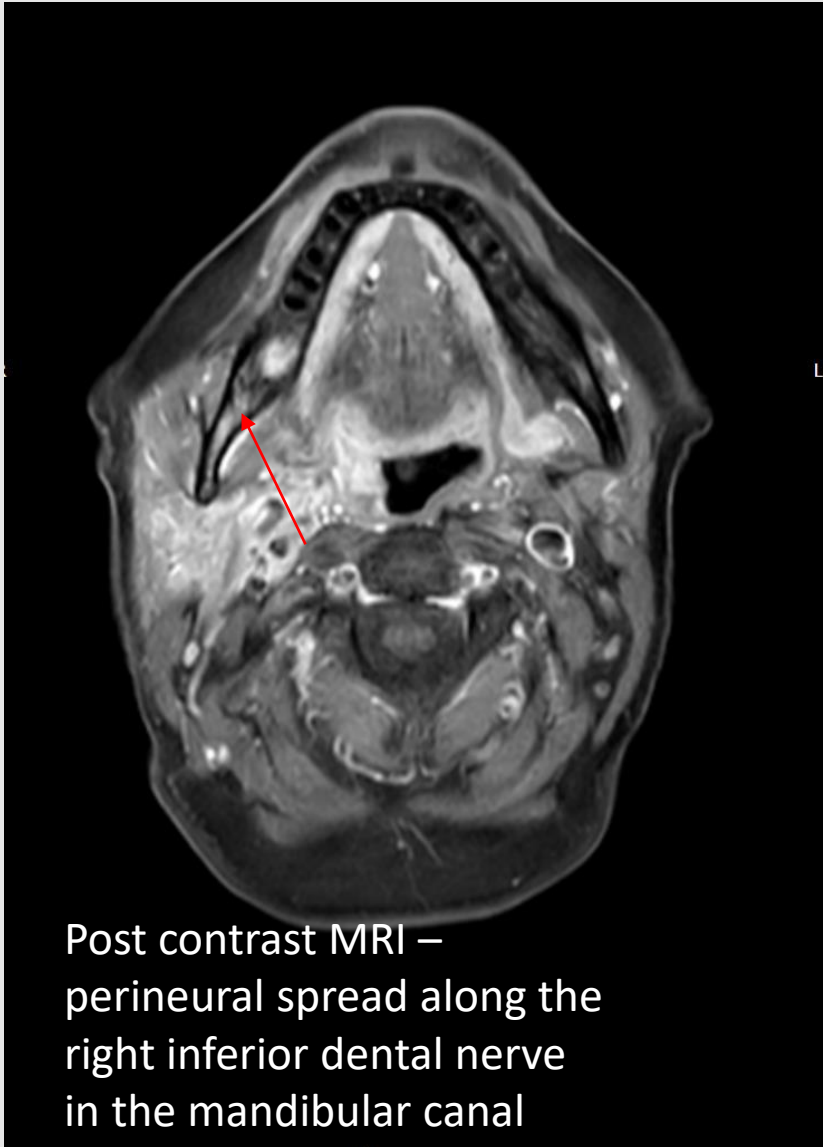
# Ultrasound image showing maximum length (A)



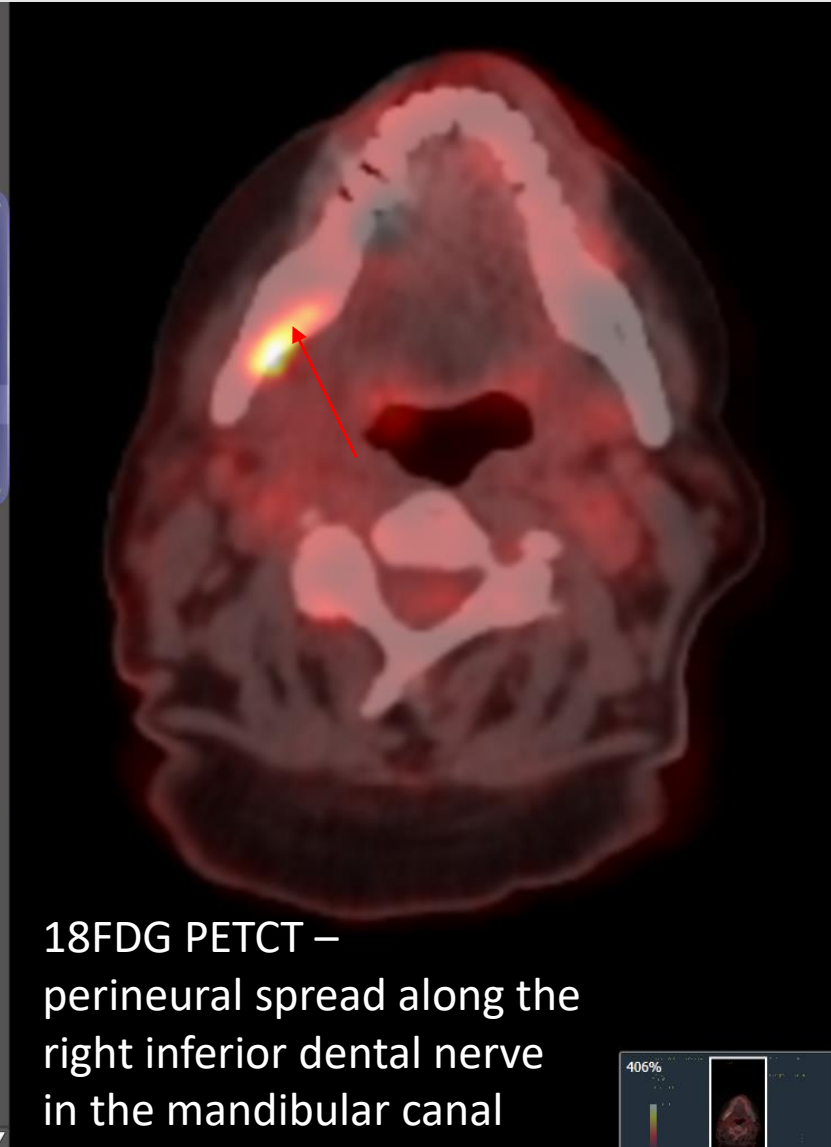
# Pre-treatment Head MRI, axial view. T1 weighted image.

Large 45 x 24 x 29 mm part solid, part cystic tumour in the right parotid with extensive perineural spread





Post contrast MRI –  
perineural spread along the  
right inferior dental nerve  
in the mandibular canal



18FDG PETCT –  
perineural spread along the  
right inferior dental nerve  
in the mandibular canal

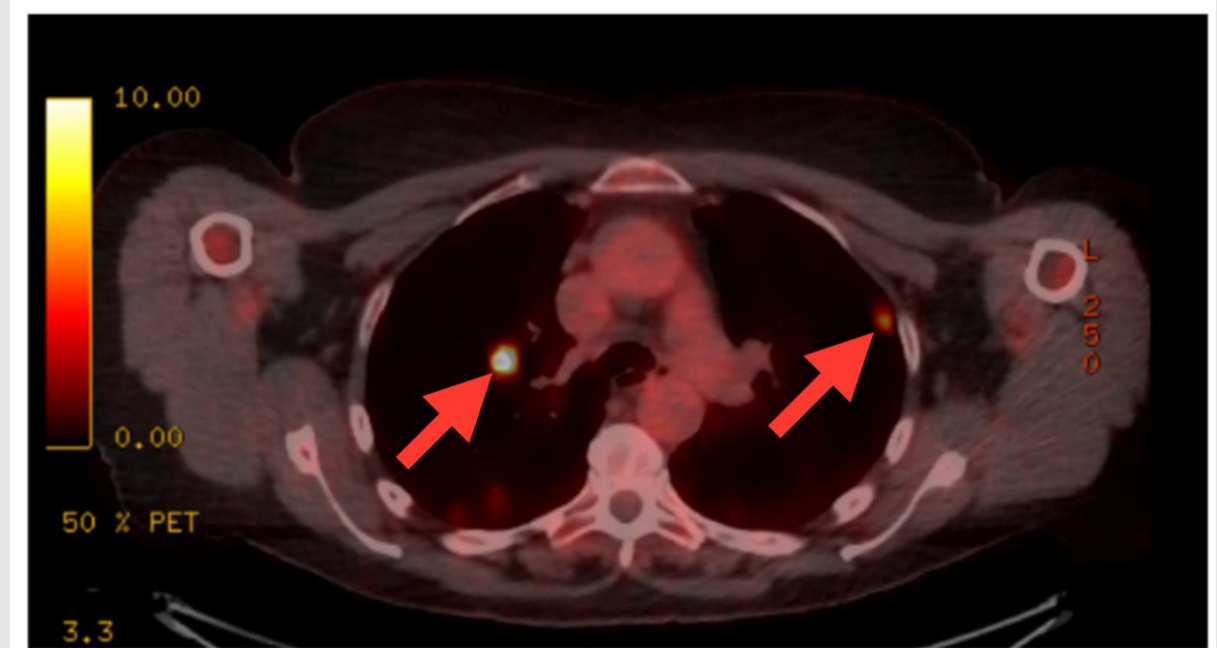


# Lung metastases

PET scan showing lung metastases (arrows)

An analysis of the U.S.A SEER database found distant metastases in 1.1% of cases.

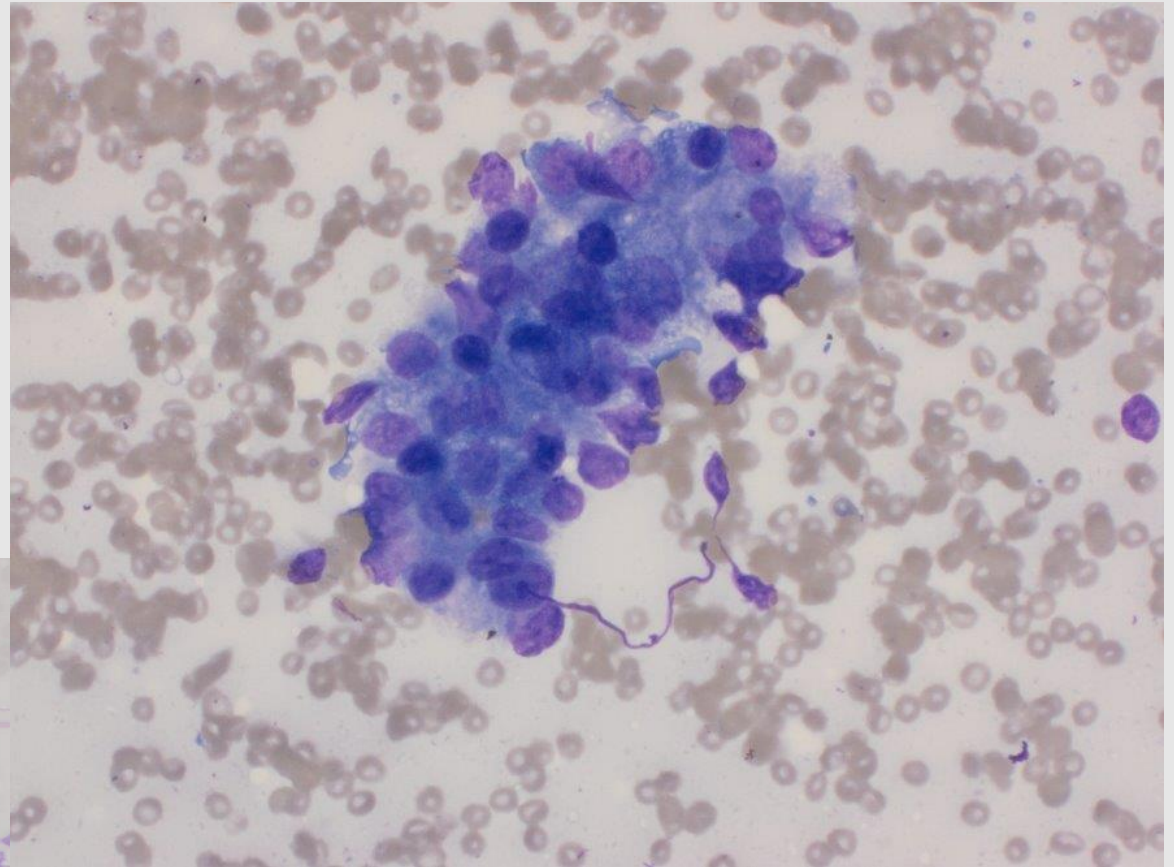
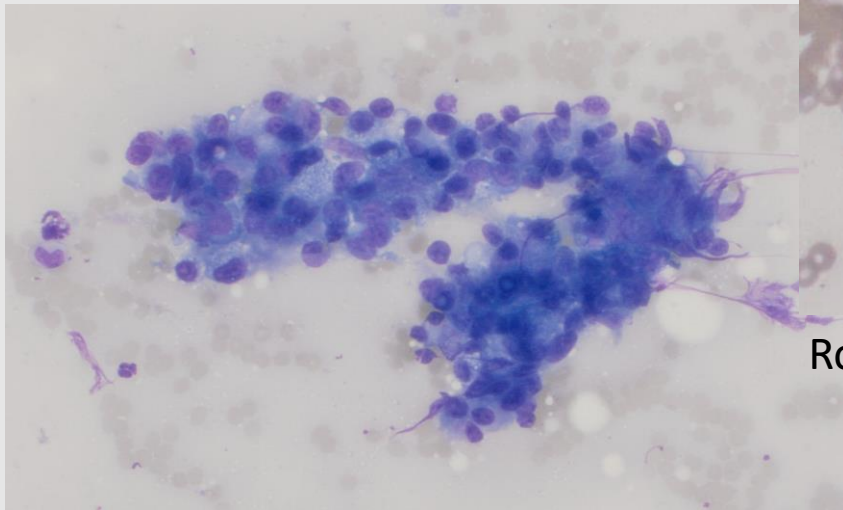
Most commonly involving lungs (44%), bone (40%), liver (12%) and brain (4%)





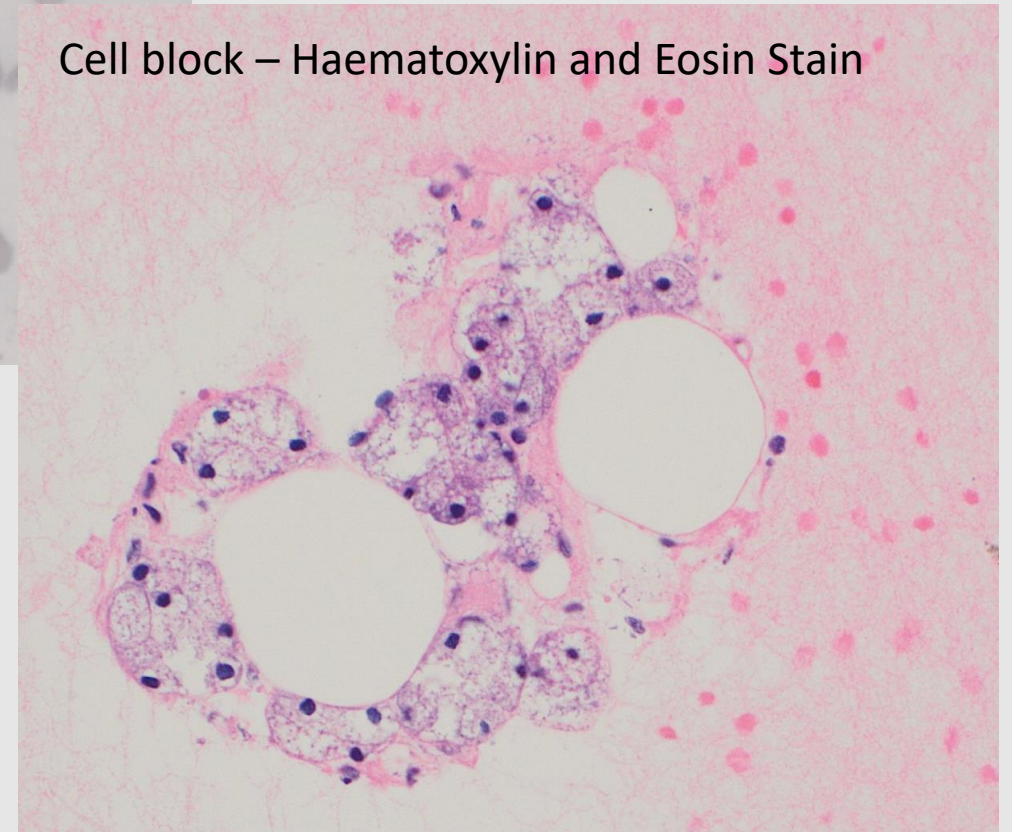
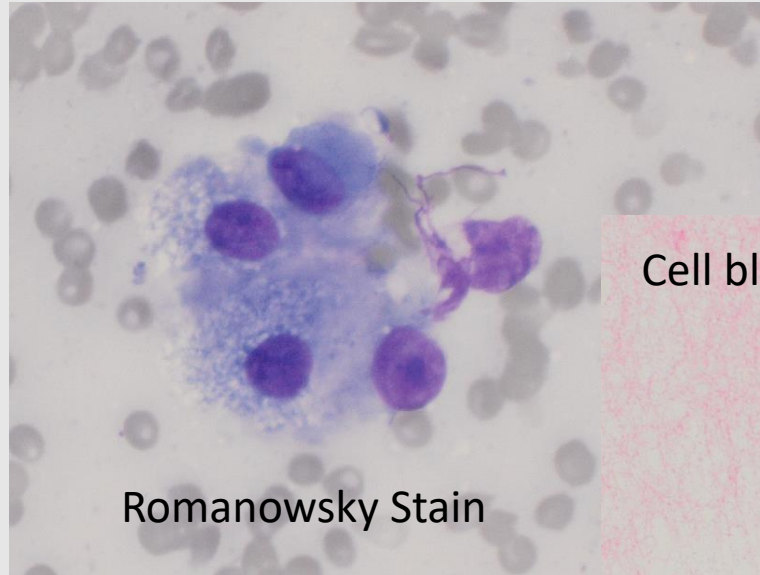
# Cytology of the right parotid lesion

- Uniform population of epithelioid cells
- Loosely cohesive
- Uniform round eccentrically placed nuclei
- Conspicuous nucleoli
- No necrosis or mitotic activity



Romanowsky Stain

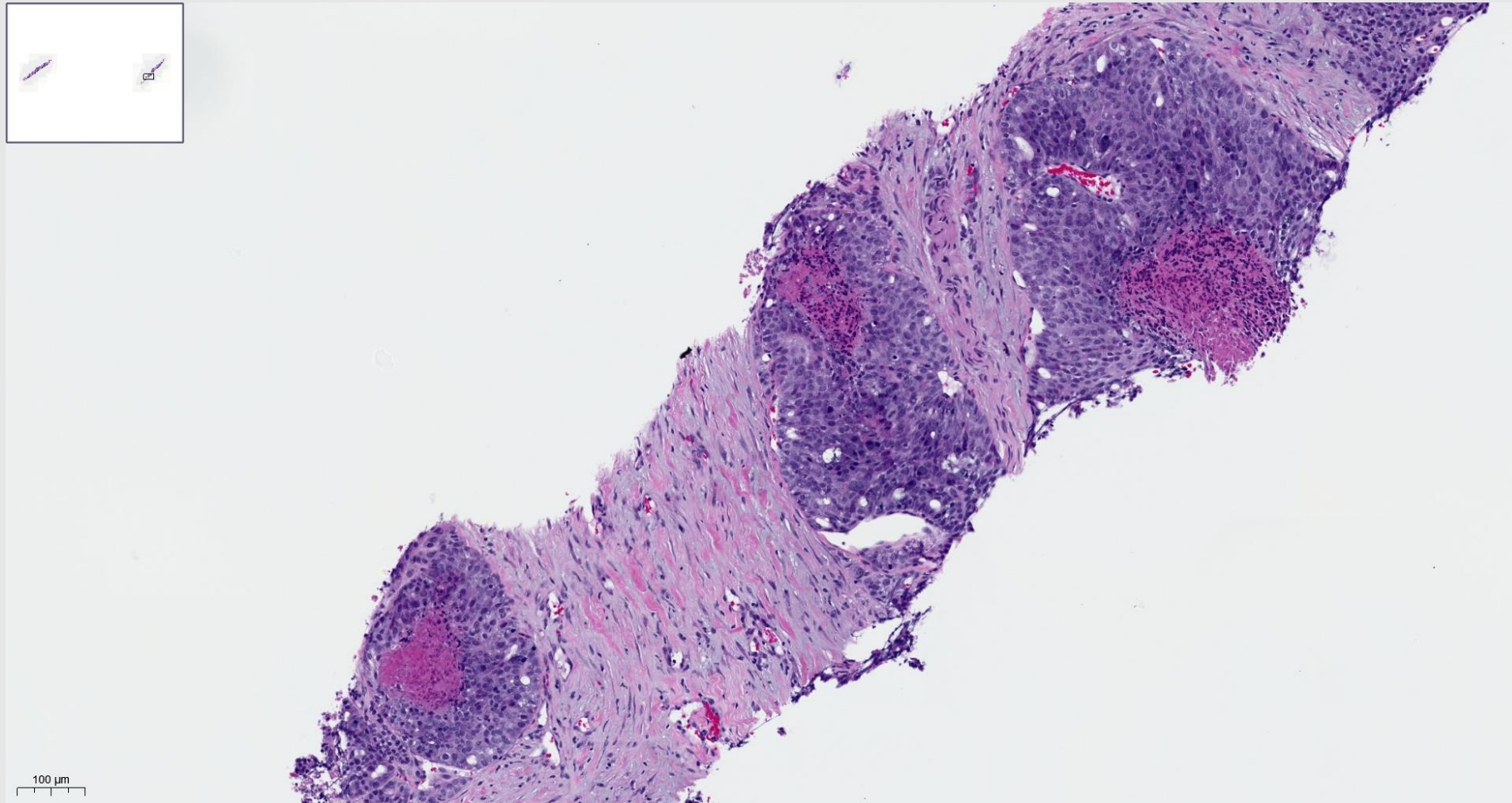
# Cytology of the right parotid lesion



- Low nuclear to cytoplasmic ratio
- Polygonal cells
- Delicate vacuolated cytoplasm with basophilic quality
- Zymogen granules



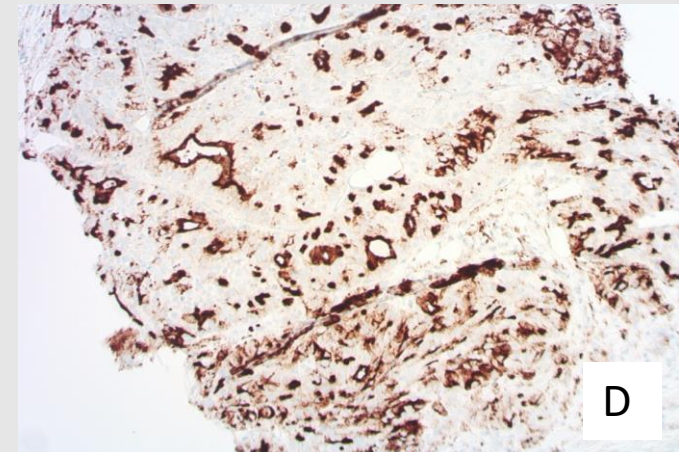
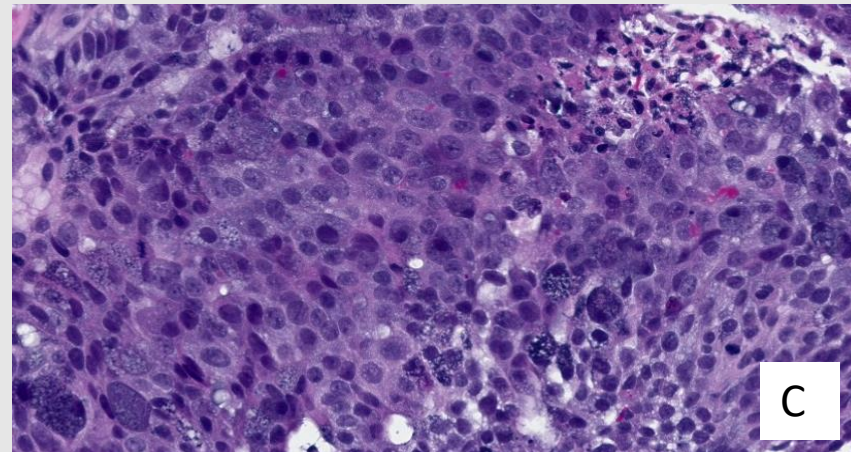
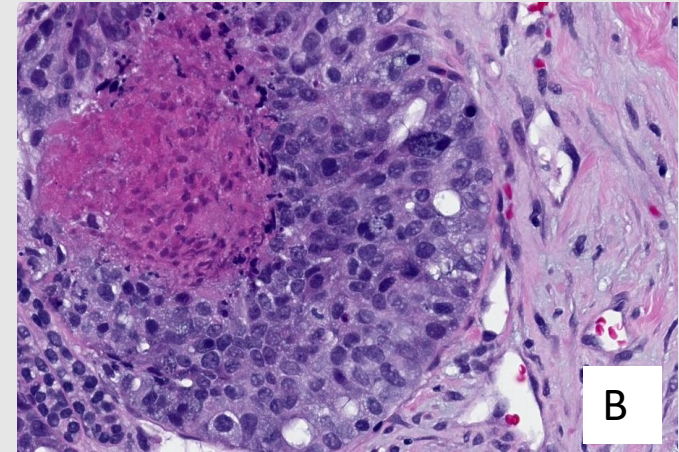
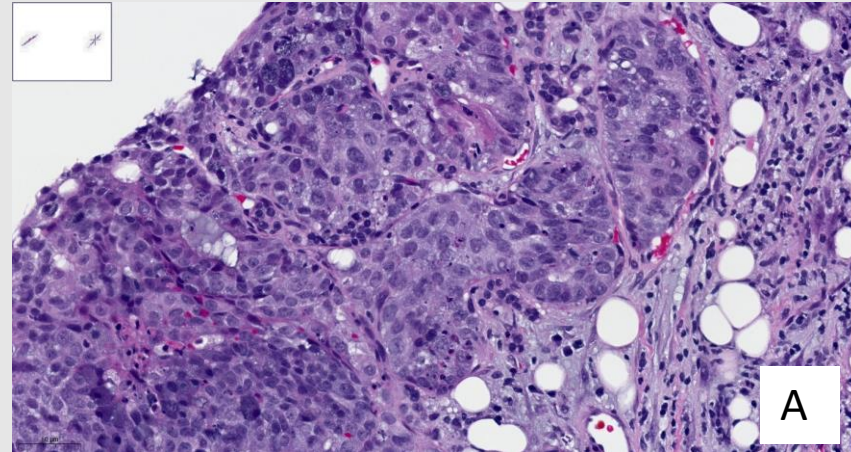
# Core biopsy histology - 10x magnification





# Core biopsy histology - medium and high power

- At higher power basophilic zymogen granules can be appreciated consistent with acinic cell carcinoma
- Comedo-type necrosis and mitotic figures indicating high grade transformation
- Characteristic DOG1+ immunohistochemical pattern (D) of acinic cell carcinoma (AcCC)



# Further predictive and molecular studies

- HER2, androgen receptor, PDL1 and NR4A3 FISH were requested
- HER2 and androgen receptors are rarely altered in acinic cell carcinoma but can provide therapeutic oncological targets
  - These were negative
- The PDL1-CPS score was 3 (patients are considered suitable for Pembrolizumab therapy when CPS is at or above 1)
- NR4A3 is an oncogenic driver event in salivary AcCC and FISH testing detected a gene rearrangement confirming the diagnosis

# AcCC – Cytopathology and Histopathology

- Usually, a low-grade malignancy which can be challenging on cytology.
- Beware similar appearances to:
  - Normal acini (differentiated by ++ acinic cells and bare nuclei, absent adipocytes and ductal epithelium)
  - Oncocytic lesions - Warthin tumour, oncocytoma (cell border less sharp in AcCC, with acinar cell arrangement, and absence of amorphous debris), oncocytosis
- Histologically varied – solid, microcystic, papillary cystic, and follicular
  - Graded by serous acinar cell differentiation. The more common well-differentiated tumours have a better prognosis - 5-year survival, 83% compared to 50% in poorly differentiated tumours

# AcCC – How it presents

- 10-15% of salivary gland epithelial malignancies
- Primarily found in the parotid gland, but comprise only 3% of parotid tumours
- Female: Male ratio of 1.5:1
- Can affect all age groups, mean age 50 years
- Rarely aggressive. Typically presents as an asymptomatic, slow-growing mass of 1-8cm

# AcCC - Imaging

- WHO classification of salivary gland pathology (2022) does not include imaging criteria for diagnosis
- Main role is to characterise the lesion and its anatomical neighbours



# AcCC – management and prognosis

- Salivary gland malignancies are rare, and research is mostly drawn from observational studies or case reports
- Large retrospective cohort study in the U.S.A: overall 10-year survival was 93.81%
- Poor prognosis if painful, tethered to surrounding tissue, or involving the facial nerve
- Treatment
  - Partial or total surgical resection of the associated gland
  - Consideration of adjuvant radiotherapy or systemic oncological therapy

# Key Points

- Usually found in the Parotid.
- Slightly more common in females
- Affects a wide age range
- Diagnosis on cytology is possible but complicated due to morphological mimics and variations.
- DOG1 staining is a useful adjunct, showing strong positivity.
- High grade transformation signifies an aggressive tumour with increased risk of metastases and worse clinical outcomes

# References

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