

Adenoid Cystic Carcinoma Of The Prostate With PSA-Negative Biology And Progressive Pulmonary Metastases: A Report Of Two Cases

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BACKGROUND

- **Rare basal-cell malignancy** of the prostate, with fewer than 150 reported cases.
- **Mimics acinar adenocarcinoma**; diagnosis rests on immunohistochemistry.
- **Not androgen-driven**: NKX3.1–, little or no PSA, poor response to hormone therapy.
- **Aggressive**: about one in five patients develop distant metastases.

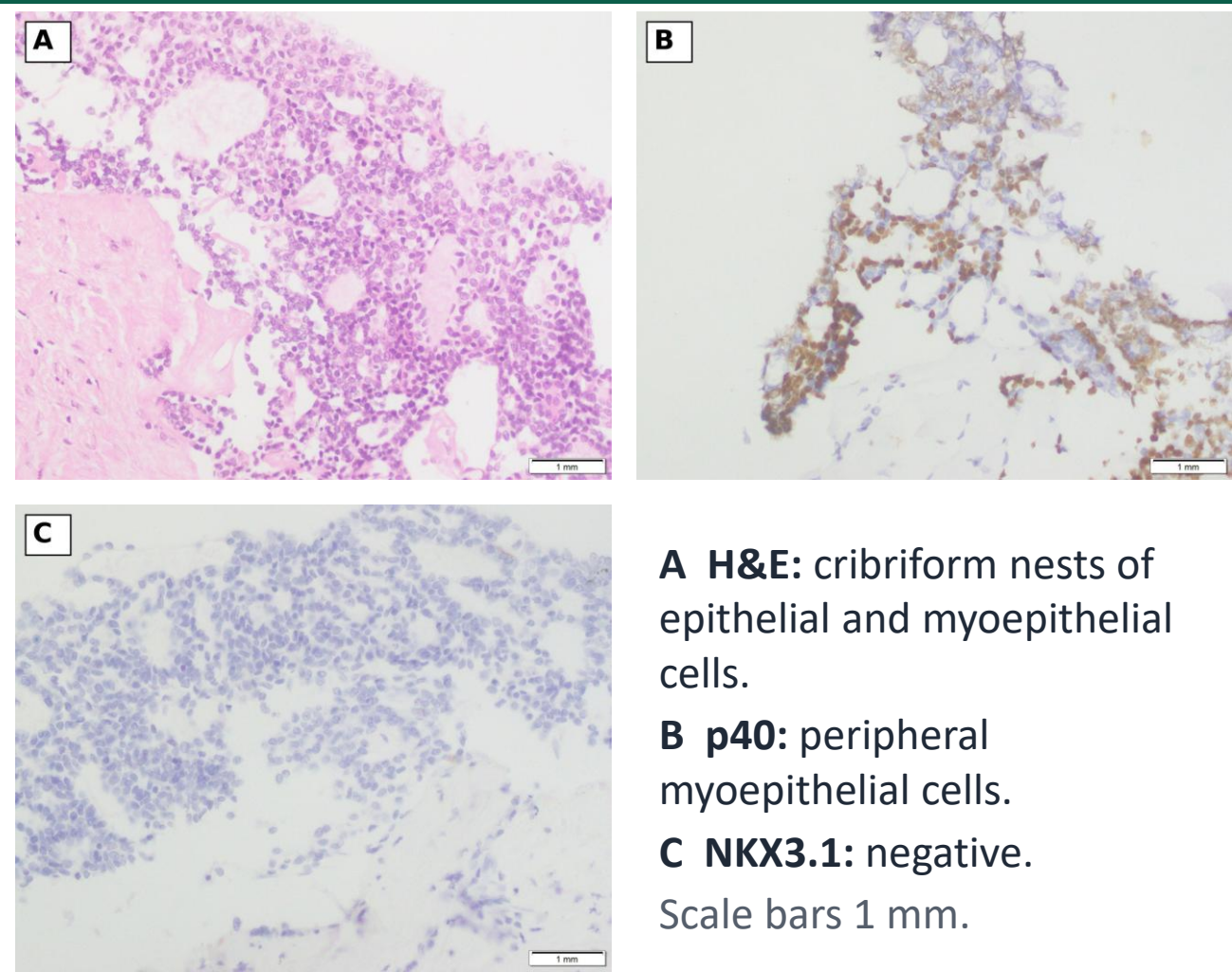
CASE 1 – 54-year-old smoker

- **Presentation**: LUTS after two TURPs; hard anal-canal mass on DRE.
- **Pathology**: reported as Gleason 4+3=7; on review a biphasic basaloid tumour with perineural invasion, CK+, p63+, NKX3.1–.
- **Stage / PSA**: T4 N0 with bone metastases; PSA 0.01–0.03 ng/mL throughout.
- **Course**: RT, ADT and chemotherapy → lung, liver and spine progression.

CASE 2 – 44-year-old ex-smoker

- **Presentation**: LUTS, urinary retention; large hard prostate on DRE; TURP + trucut biopsy.
- **Pathology (see images)**: cribriform epithelial–myoepithelial tumour; p40+, NKX3.1–, PSA–, INSM1–.
- **Stage / PSA**: ≈9 cm mass, T3a/T4 N0 M0; PSA 0.45 → 0.01 ng/mL on ADT.
- **Course**: RT, ADT and chemotherapy → regrowth and enlarging lung nodules.

IHC & Histology



DISCUSSION

- **IHC makes the diagnosis**: p63/p40+, HMWCK+, NKX3.1–, PSA– – the reverse of acinar adenocarcinoma. Case 1 was mislabelled Gleason 4+3=7 until this profile was recognised; INSM1– excluded neuroendocrine carcinoma.
- **Biology**: a subset carries the MYB–NFIB fusion of salivary ACC; PTEN loss and EGFR/HER2 overexpression are reported, but no drug target is established.
- **Behaviour**: ~20% metastasise in larger series; both our patients progressed in the lung despite RT, ADT and carboplatin–paclitaxel. Lenvatinib, active in salivary ACC, is planned for Case 1.

TABLE 1 – The two cases side by side

Feature	Case 1 (54 y)	Case 2 (44 y)
Initial diagnosis	Acinar adenoca., Gleason 4+3=7	Adenoid cystic carcinoma
Immunohistochemistry	CK+, p63+, NKX3.1–	p40+, NKX3.1–, PSA–, INSM1–
PSA (ng/mL)	0.01–0.03 throughout	0.45 → 0.11 → 0.03 → 0.01
Stage / spread	T4 N0; bone → liver, lung	T3a/T4 N0; lung only
Treatment	RT 55 Gy; ADT; carboplatin–paclitaxel ×6	RT 30 Gy; ADT; carboplatin–paclitaxel ×6
Course	Progression; lenvatinib planned	Response, then lung progression

TAKE-HOME MESSAGES

- 1 A basaloid prostatic tumour, or an aggressive prostate cancer that does not fit a low PSA, should raise the possibility of adenoid cystic (basal cell) carcinoma.
- 2 Confirm with a basal-cell panel: p63/p40+, HMWCK+, NKX3.1–, PSA–; INSM1– excludes neuroendocrine carcinoma.
- 3 PSA cannot be used for diagnosis, staging or follow-up – rely on CT/MRI and bone scan.
- 4 The tumour is not androgen-driven, readily spreads to the lung and has no standard systemic therapy: manage in a multidisciplinary setting and consider MYB testing.

Key references: Iczkowski KA et al. Am J Surg Pathol 2003;27:1523-9. Ali TZ, Epstein JI. Am J Surg Pathol 2007;31:697-705. Bishop JA et al. Hum Pathol 2015;46:1204-8. Cozzi S et al. Curr Oncol 2022;29:1866-76. WHO Classification of Tumours, 5th ed. IARC; 2022. Consent for publication obtained; no conflicts of interest.