

CASE OF THE FORTNIGHT
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MERKEL CELL CARCINOMA

68 YEAR OLD MALE PATIENT WITH RAPIDLY GROWING SWELLING OF 4 MONTHS DURATION.
ON THE DORSAL ASPECT OF FOREARM.
O/E FAIRLY WELL CIRCUMSCRIBED FLESH COLOURED NODULE., 2 X 1.8 CM.
NO HISTORY OF OTHER TUMOURS IN THE BODY.

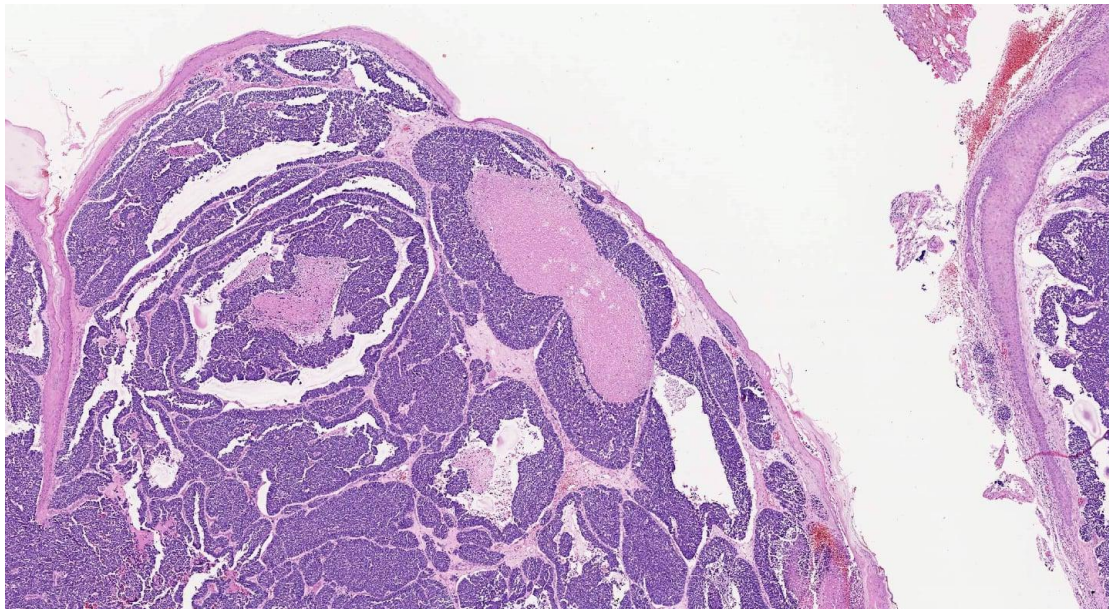


Fig 1: A nodular small round blue cell tumour centred in the dermis

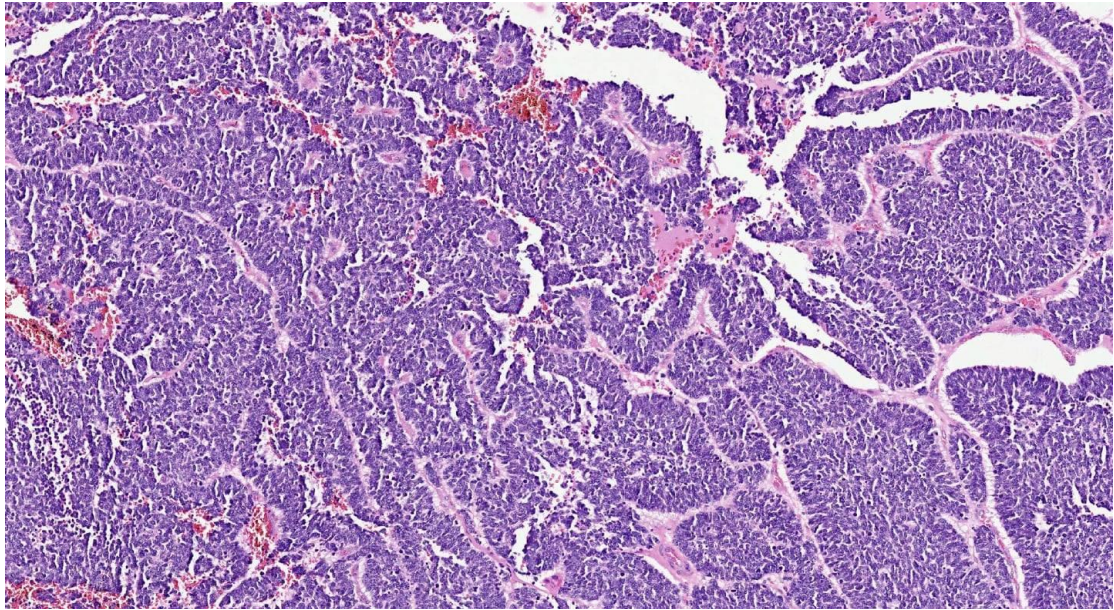


Fig 2: Tumour cells arranged as sheets, nests and trabeculae, occasional pseudorosettes

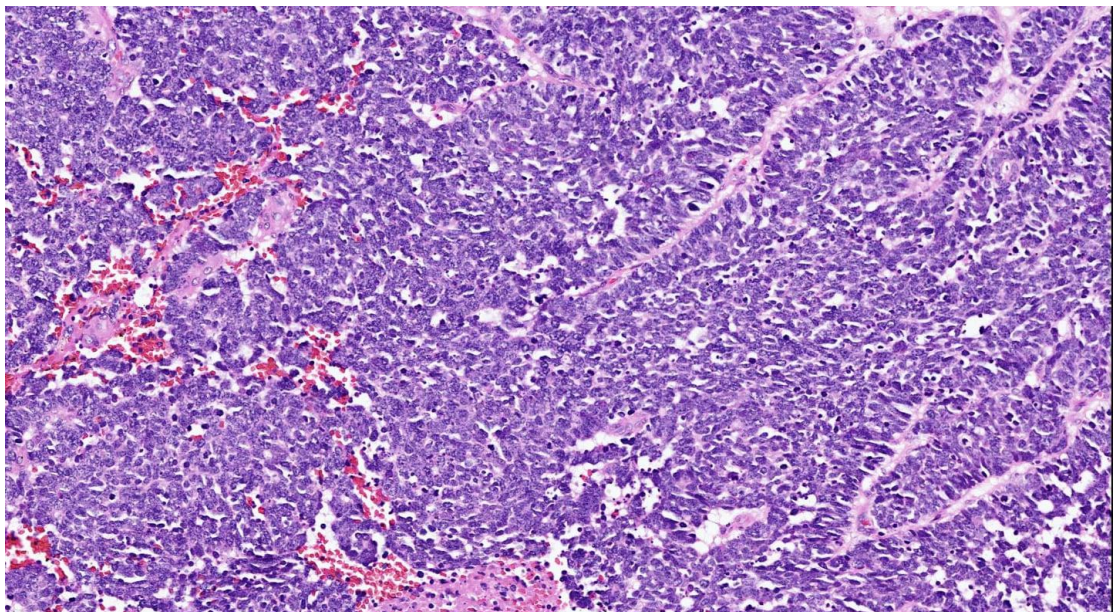


Fig 3: Tumour cells with scant cytoplasm, molded nuclei with stippled chromatin

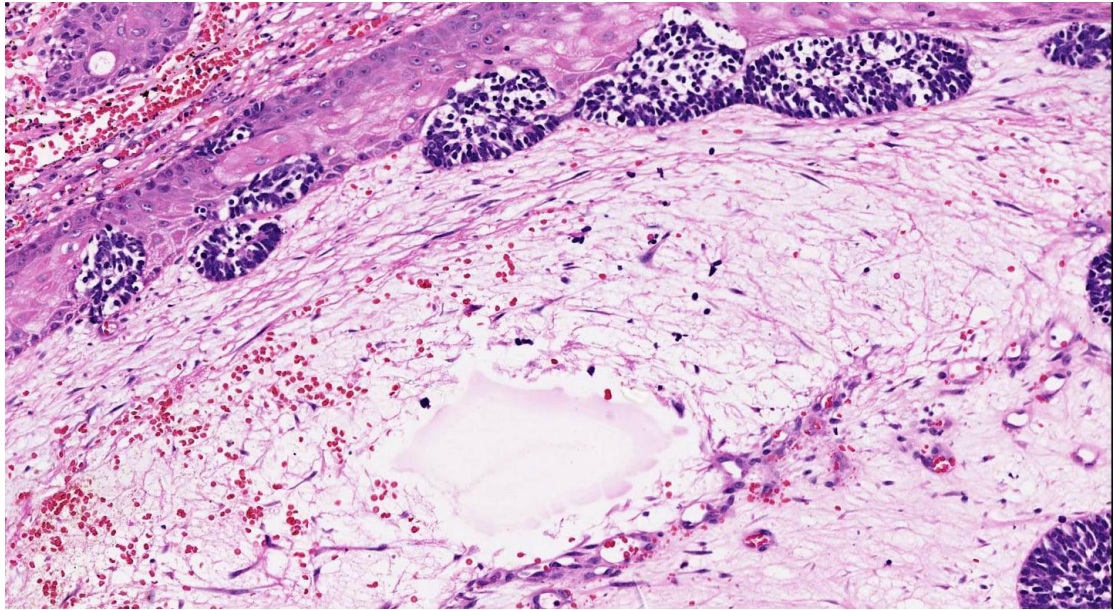


Fig 4: A rare instance of epidermotropism of tumour cells

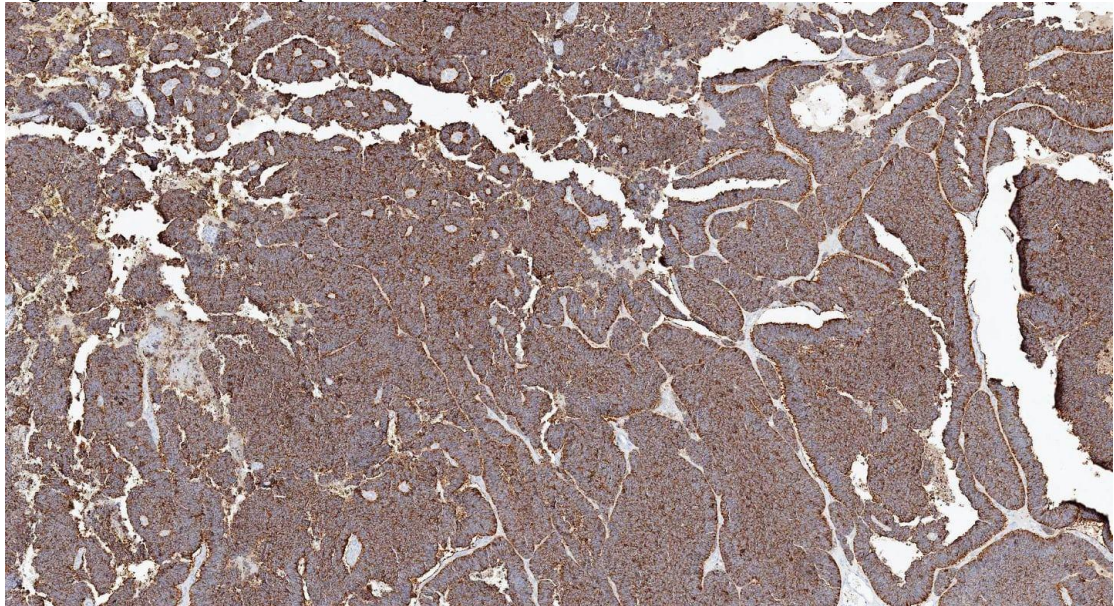


Fig 5: Diffuse positivity with Synaptophysin

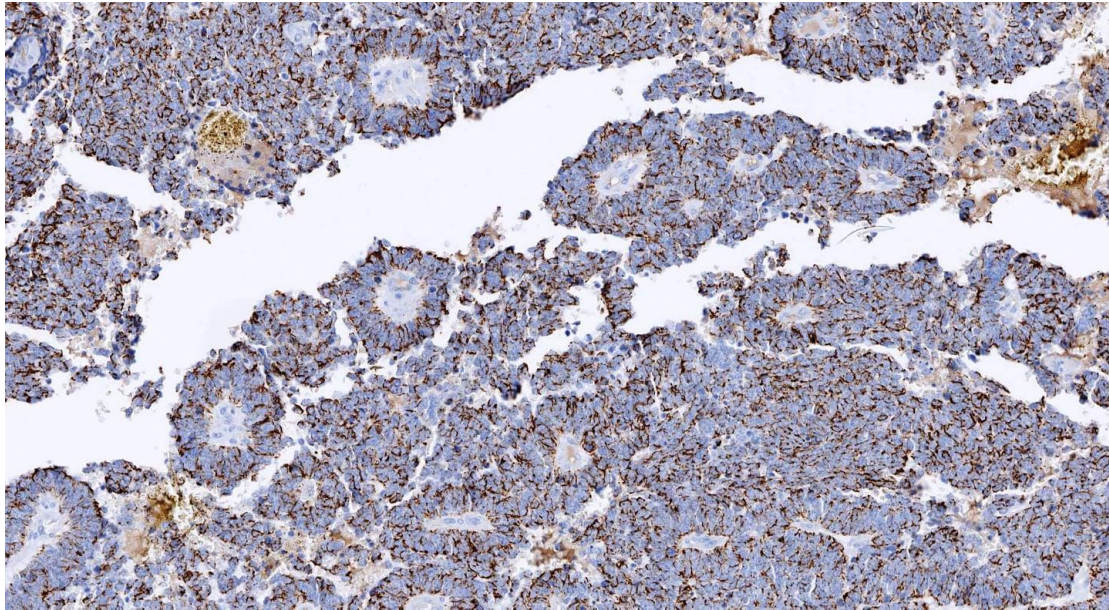


Fig 6: Characteristic paranuclear dot like positivity with CK 20 (some cases may show cytoplasmic positivity)

LEARNING POINTS:

- 1) A rare, aggressive, **primary cutaneous neuroendocrine carcinoma**.
- 2) Elderly males, sun exposed skin of head and neck/ extremities/ trunk
- 3) Immunosuppression and advanced age are risk factors.
- 4) Cell of origin- unknown, possible precursors include Pro-B/ Pre-B lymphocytes, fibroblasts, dermal mesenchymal stem cells and epithelial cells.
- 5) Clinically, **rapidly growing violaceous/ flesh coloured nodules**.
- 6) **Microscopy: Small blue round cell tumour** in the dermis and/ or subcutis.
Cells with high nuclear: cytoplasmic ratio, **salt and pepper chromatin**.
Sheets/ nests/ occasional trabeculae/ pseudo-rosettes
Mitoses/ apoptosis/ necrosis.
Squamous differentiation maybe seen.
- 7) **Immunohistochemistry:**
Neuroendocrine markers
CK 20- paranuclear dot- like positivity
Neurofilament
SAT B2
Often , Merkel Cell Polyoma Virus (see below)

8) TWO MAJOR SUBSETS BASED ON VIRAL STATUS:

Feature	MCPyV-positive	MCPyV-negative (UV exposure related)
Incidence	More common	Less common
Morphology	Pure	Pure/ combined
Prognosis	Better	Worse
Tumor infiltrating lymphocytes	Brisk	Few
IHC	Rb pos (+++) P53 pos (+) P63 pos (+)	Rb pos (+) P53 pos (+++) P63 pos (+++)
Genetics	Tumour mutational burden	Tumour mutational burden

	low No Recurrent RB1/ P53 mutations No UV mutation signature	high Recurrent RB1/ P53 mutations UV mutation signature
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9) Treatment: Excision, lymph node resections, Immune-checkpoint-inhibitors.

10) *Data for inclusion in pathology reports:*

- Maximum dimension of tumor (gross or microscopic)
- Tumor confined to dermis/subcutis: yes/no
- Involvement of underlying muscle, fascia, bone or cartilage: yes/no
- Lymphovascular involvement: yes/no
- Distance from surgical margin
- Local microscopic “satellite” deposits of tumor: yes/no
- Distance between main tumor and deep surgical margin
- Morphology (pure neuroendocrine or combined): pure or combined
- Merkel cell polyomavirus status: positive or negative
- Immunohistochemical profile
- P63 expression: positive or negative

REFERENCES:

- 1) Noreen M. Walsh, Lorenzo Cerroni; Merkel cell carcinoma: a review, Journal of cutaneous pathology <https://doi.org/10.1111/cup.13910>
- 2) WHO classification of skin tumours, 4th ed.