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.

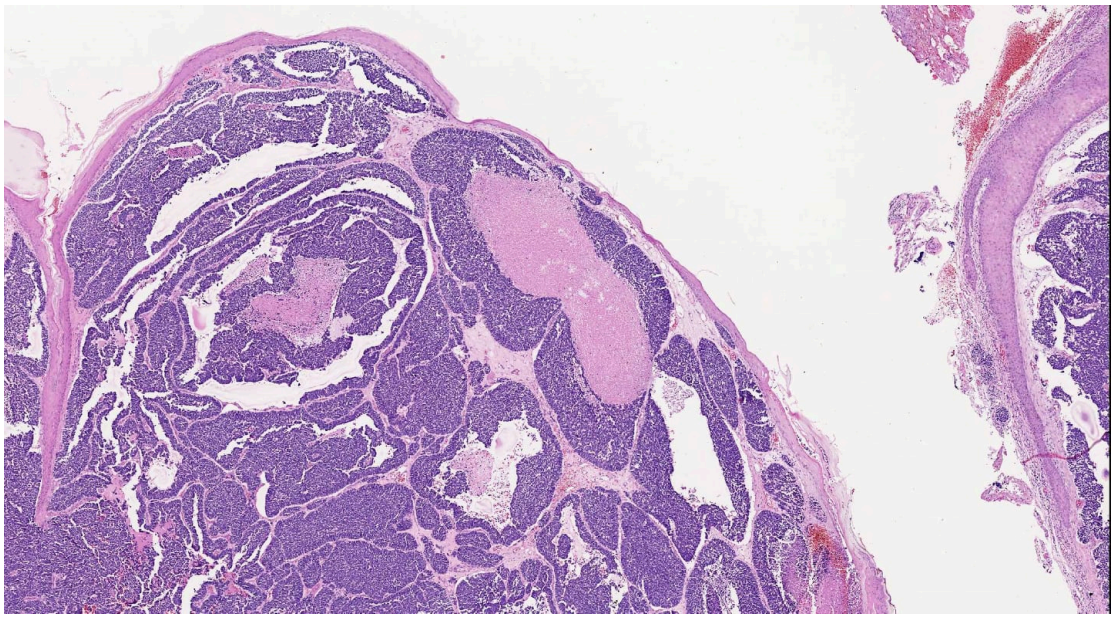

Fig 1: A nodular small round blue cell tumour centred in the dermis

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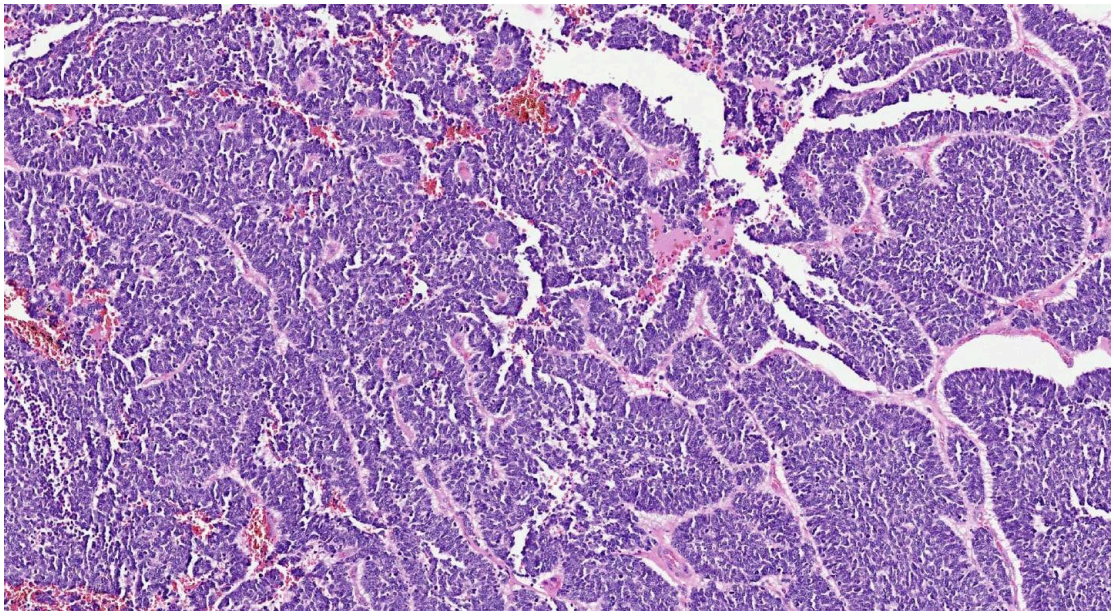

Fig 2: Tumour cells arranged as sheets, nests and trabeculae, occasional pseudorosettes

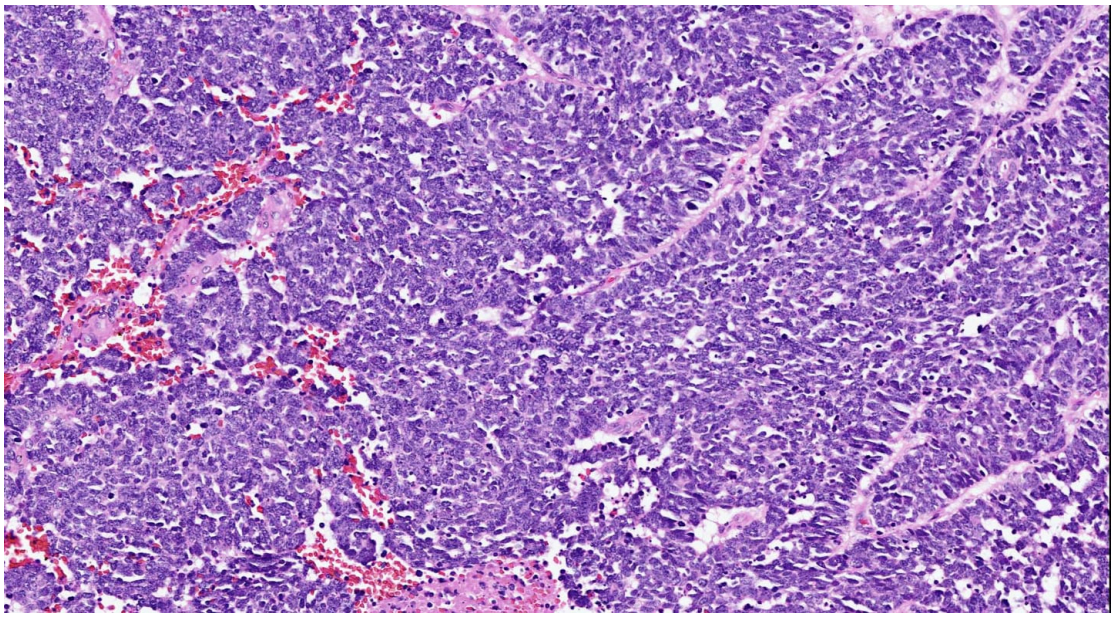

Fig 3: Tumour cells with scant cytoplasm, molded nuclei with stippled chromatin

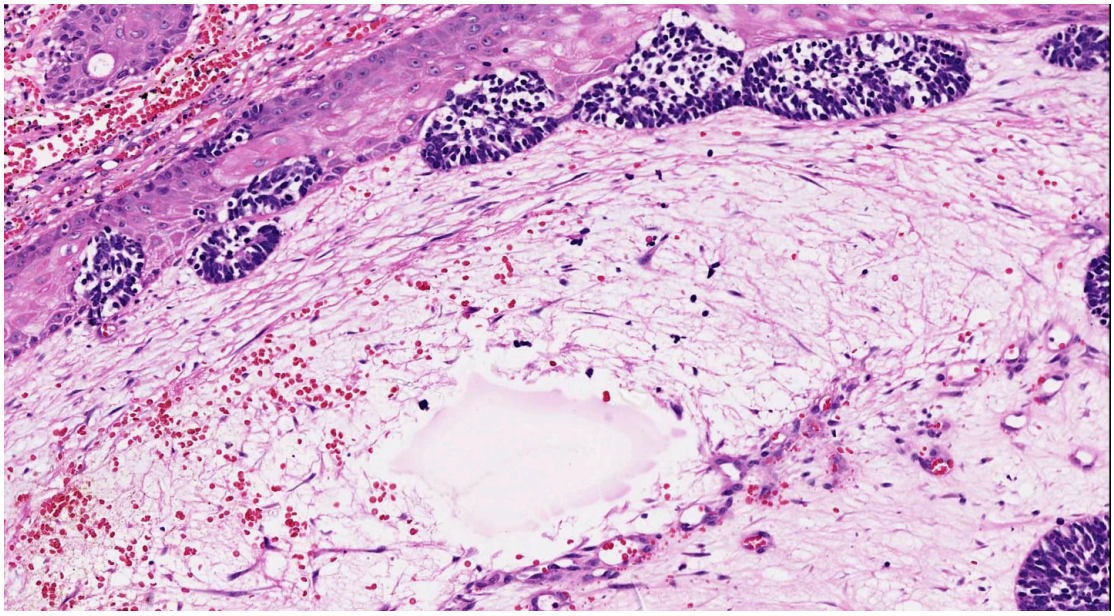

Fig 4: A rare instance of epidermotropism of tumour cells

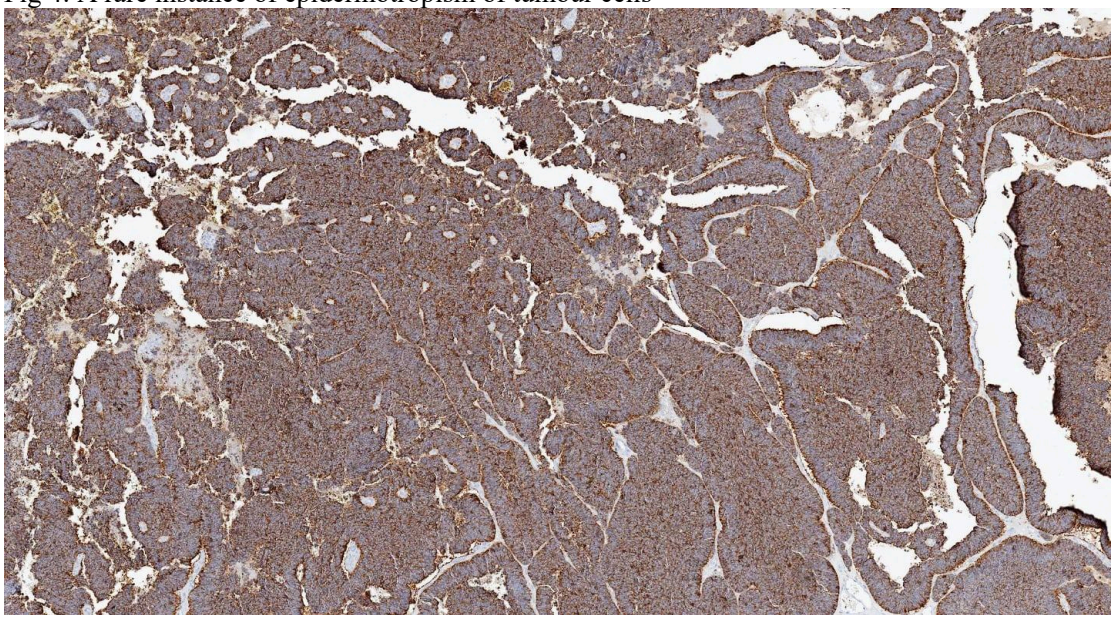

Fig 5: Diffuse positivity with Synaptophysin

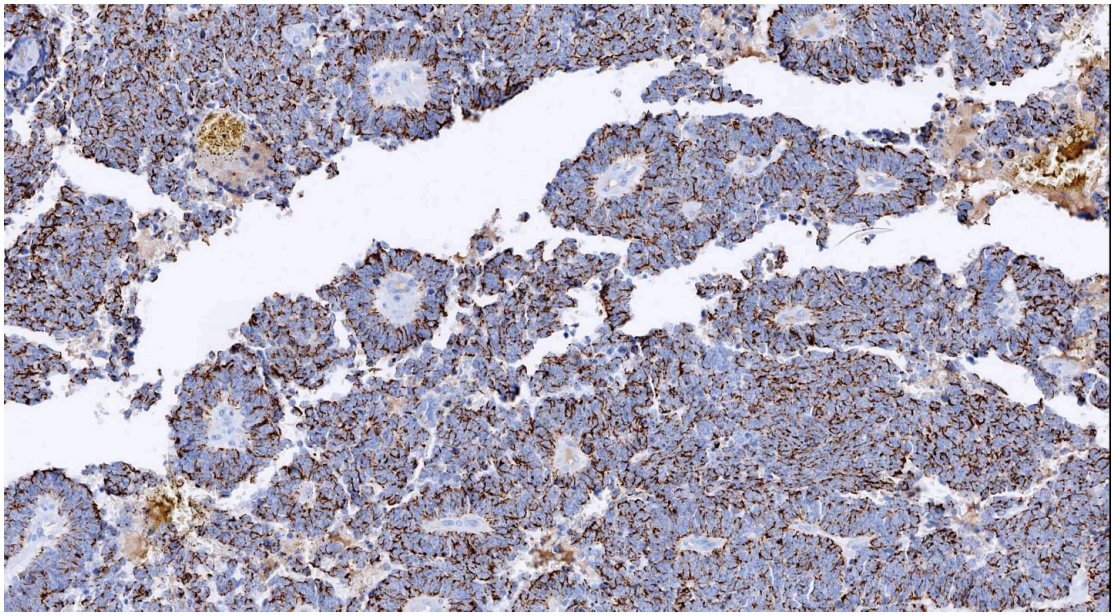

Fig 6: Characteristic paranuclear dot like positivity with CK 20 (some cases may show cytoplasmic positivity)

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

9) Treatment: Excision, lymph node resections, Immune-checkpoint-inhibitors.

10) <u>Data for inclusion in pathology reports:</u>

- 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
- Immunohistchemical 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 classifcation of skin tumours, 4th ed.