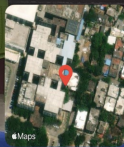
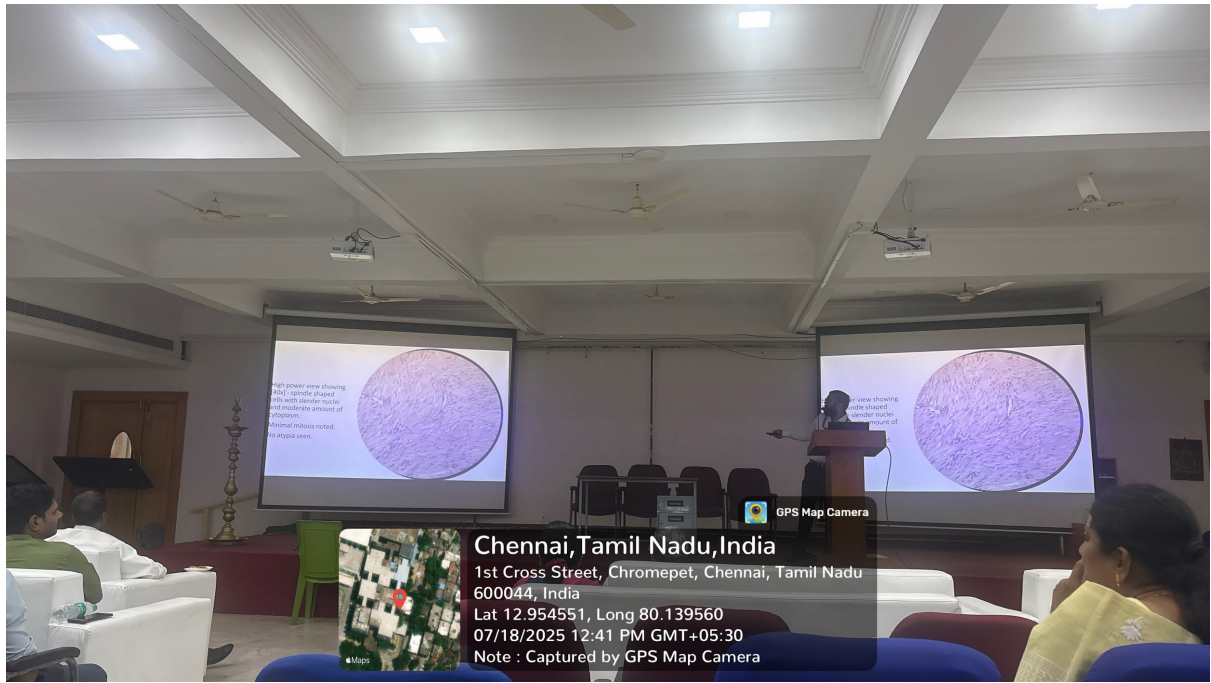


Surgery Path IDM

TOPIC - Spindle cell neoplasm- Dermatofibrosarcoma Protuberans (DFSP)

PRESENTER - Dr. Klinton

1. Definition and Nature: DFSP is a slow-growing, low-grade malignant dermal tumor with a high propensity for local recurrence. It is characterized by a storiform (cartwheel-like) pattern of spindle cells and strong CD34 positivity.
2. Clinical Case Summary: A 25-year-old female presented with a recurrent swelling over her left leg, first noted after trauma. It showed progressive growth, with recurrence after excision and current presentation over 8 years.
3. Imaging and Biopsy: MRI showed a recurrent, complex cystic lesion involving the skin and subcutaneous tissue. FNAC was hemorrhagic and inconclusive; biopsy revealed spindle cells with storiform arrangement.
4. Histopathology: The tumor was composed of slender spindle cells arranged in whorls and sheets, infiltrating subcutaneous fat with minimal mitosis and no atypia. No tumor was found at margins on frozen section and final HPE.
5. Immunohistochemistry (IHC): Tumor cells were strongly CD34 positive; negative for S100 and SOX10. Ki-67 showed low proliferation (2%), supporting a diagnosis of DFSP.
6. Microscopic Features: Classic DFSP shows bland spindle cells in a storiform pattern, infiltrating dermis and subcutis. Occasional myxoid changes and honeycomb infiltration of fat are seen.
7. Gross Findings: Typically presents as a grey-white nodular mass involving dermis and subcutis. Lesions can become protuberant and multi-nodular in advanced/recurrent cases.
8. Genetics and Pathogenesis: Commonly associated with t(17;22)(q22;q13) translocation resulting in the COL1A1-PDGFB fusion gene, often seen in ring or derivative chromosomes.
9. Differential Diagnosis: Includes cellular schwannoma, MPNST, monophasic synovial sarcoma, and spindle cell melanoma—distinguished via IHC and morphology.
10. Treatment and Prognosis: Mainstay of treatment is wide local excision with clear margins. DFSP has low metastatic potential but high local recurrence rate, especially if not excised completely.



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