Answer to Quiz 3:

Dermatofibrosarcoma protuberans

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Dermatofibrosarcoma protuberans (DFSP) are cellular dermal neoplasms which initially present as firm, flesh-colored, morphea-like plaques that eventually become lobulated protuberant nodules, occasionally with ulceration . It usually involves the skin of the chest, back, and thighs is particularly in the third and fourth decades, and there is a slight male predisposition. The average diameter of the tumour is 5 cm in diameter, forming asymmetrical expansive and infiltrating cellular nodules within the dermis, with frequent extension into the underlying subcutaneous fat and soft tissue. Clues to the diagnosis include the hypercellularity of the neoplasm and a tendency to infiltrate the deep dermis as intricate and lacelike trabeculae, imparting a honeycomb effect. Cells are characteristically arranged into a cartwheel-like "storiform "configuration.

In contrast to fibrous histiocytomas DFSP have a cellular rather than collagenous background, do not contain foam cells, and haemosiderin deposits are unusual. Collagen fibers in DFSP do not show birefringence under polarize light (Barr et al,1986), but some authors have not found to be a consistent differential features (Okun et al, 1988).

Mitotic figures including atypical mitoses may be seen and multinucleated tumour cells are sometimes present. Necrosis is generally minimal. Focal myxoid degeneration may be present. Rarely, DFSP may contain dendritic cells which produce melanin pigment (Dupree et al 1985). These pigmented cells which synthesize melanosomes and express S-100 protein (Fletcher 1988) probably represent colonization of DFSP by dendritic melanocytes. The variant of DFSP which contains melanin pigment is known as Bendar tumor. This should not be confused with predominantly spindle cell form of malignant melanoma. Tumour cells in DFSP including majority of cells in Bendar tumours are negative for S100 protein, permitting distinction from spindle cell malignant melanoma and diffuse forms of neurofibroma.

DFSP is prone to local recurrence (30-50 %) after simple excision. Wide local excision tend to lower the reported rate of local recurrence. Recurrences may manifest several years after initial surgery, suggesting a need for long-term follow-up. Metastases are very rare and tend to occur in lesions that have locally recurred. Spread to regional lymph nodes and to the lung have been described. DFSP is almost certainly fibroblastic or perineural fibroblastic in origin rather than fibrohistiocytic .

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