Short Course 11

Pigmented lesions of the skin

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Problematic melanocytic nevi

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Whether the diagnosis of any particular nevus is problematic or not depends upon a variety of factors, including the experience and enthusiasm of the pathologist, the nature of the specimen (shave vs. punch vs. excisional), the quality of the sections (and their staining), the hour of the day or day of the week in addition to the problems relating to the ever-increasing range of histological variants that we are obliged to recognize. Reporting nevi, difficult or otherwise late on a Friday afternoon is probably always a mistake particularly if it follows rather than precedes the customary preweekend celebration (the TGIF club)! This presentation focuses on some of the more commonly misdiagnosed and problematic nevoid variants.

Neonatal and childhood congenital nevi

In the majority of cases, congenital nevi in neonates are not significantly different from similar lesions arising in children or adolescents. Rarely however, they may be characterized by extremely worrisome histological features including a junctional component (which may be lentiginous and nested) composed of pleomorphic epithelioid nevus cells with abundant cytoplasm and enlarged hyperchromatic or vesicular nuclei containing prominent eosinophilic nucleoli (1, 2). One or two mitoses may be seen. Particularly disturbing is the presence of pagetoid spread, (sometimes to the extent that it mimics in situ melanoma) and involvement of the appendage epithelium. The dermal component, when present, may contain similar cells or else be composed of more banal nevus cells showing maturation with depth and rendering the benignity of the lesion more obvious.

Similarly disturbing features may very occasionally be seen in congenital nevi excised from young children. Although the lesions appear clinically benign, their biological potential is as yet unknown. A modest re-excision, if the nevus cells approach anywhere near to the margins would be prudent.

Desmoclastic nevus

This nevus which was first described by Barr et al. (3) as a variant of Spitz nevus is still occasionally a source of diagnostic difficulty. It most often presents on the extremities of young adults. Many lesions are devoid of melanin pigment and clinically present as a flesh-colored to yellow or erythematous papule/nodule which may be misdiagnosed as a fibrous histiocytoma.

Histologically, the nevus presents as a symmetrical, dome-shaped dermal nodule covered by attenuated or acanthotic epidermis. Some lesions show small foci of residual junctional activity and melanin pigment is often evident. Frequently, however, the lesion is solely intradermal when it may be confused with a fibrohistiocytic tumor, particularly epithelioid cell fibrous histiocytoma (4). It is typically composed of epithelioid nevus cells with abundant eosinophilic cytoplasm and large, round, to oval vesicular nuclei containing prominent eosinophilic nucleoli. Intranuclear cytoplasmic pseudoinclusions are common and mitotic figures are occasionally present. The nevus cells which are embedded in a dense, sclerotic connective tissue stroma, usually show maturation with depth. Less frequently the nevus is composed solely of spindle cells which may result in confusion with atrophic fibrous histiocytoma. Desmoplastic nevus can be distinguished from epithelioid fibrous histiocytoma by its paucicellularity, absence of even a focal storiform growth pattern and S100 protein/HMB 45 expression. Epithelioid fibrous histiocytoma often expresses smooth muscle actin and muscle-specific actin.

Recurrent nevus

A major diagnostic problem occasionally encountered following shave biopsy specimens, is the phenomenon of recurrent nevus (pseudomelanoma) (5). Re-excision specimens commonly show melanocytic hyperplasia particularly overlying the site of dermal scarrring. If the nevus has been incompletely excised, regrowth of the junctional component may sometimes be accompanied by disturbing melanocytic cytology including nuclear pleomorphism, nucleolar prominence and occasional mitotic figures. Focal pagetoid spread may even be present. Clues to the benignity of the process include the presence of dense, horizontally orientated fibrous tissue in the superficial dermis and residual dermal nevus cells deep to the scar tissue. In those examples where the cytological changes are marked, review of the original biopsy specimen is always advisable. It is worth remembering that the original lesion may of course have been a dysplastic nevus or in situ/invasive melanoma.

Deen Denetratina nevus

This uncommon nevus variant is a common source of diagnostic difficulty. Particular problems relate to its precise histogenesis and biological potential. It presents as a densely pigmented papule or nodule on the face, neck or shoulder of young adults and as such may be clinically mistaken for melanoma (6).

Histologically the epidermis may be uninvolved although, not uncommonly, foci of lentiginous or nested junctional activity are seen (7). The nevus presents a wedge-shaped appearance with its broad base uppermost, extending into the deeper dermis or subcutaneous fat as one or more often bulbous extensions. Although in the original descriptions the nevus was said to be composed of a homogenous spindle cell population, in my own experience it more often consists of a superficial epithelioid nevus population which gradually merges with spindle cells in the deeper reaches. Cytoplasm is usually finely pigmented giving a dusty, and often gray appearance. Nuclei may be hyperchromatic and smudged or vesicular with prominent small nucleoli. Intranuclear cytoplasmic
Spindle cell melanocytic tumors

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Not uncommonly, the diagnosis of spindle cell melanocytic tumors presents problems to the diagnostic histopathologist (1). For practical purposes, these can be divided into two types, as follows: i) is the tumor melanocytic? ii) is the melanocytic tumor benign or malignant? Both of these questions will be briefly addressed in this presentation.

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References