Benign Melanocytic Proliferations and Naevi
Reviewed by Dr Inara Strungs

Melanocytic proliferations and naevi often require careful clinical and/or pathological assessment to exclude malignancy. This is best achieved by accurate classification of the lesion into a recognised diagnostic category, the major ones of which are discussed below.

**Freckle (ephelis)**
A freckle (ephelis) is an area of increased melanin production without an increase in melanocyte numbers. These will often fade in the absence of UV exposure.

**Lentigo**
In contrast, a lentigo shows an increase of melanocytes in linear array along the dermoepidermal junction and persists in the absence of UV exposure. Multiple lentigines are occasionally a component of the rare hereditary syndromes with internal manifestations such as Carney syndrome.

**Labial Melanotic Macule**
Labial melanotic macule is a pigmented macule occurring on the lip that histologically demonstrates increased melanin pigment, normal melanocyte numbers and melanin spillover into the dermis. Similar lesions may occur on the penis and vulva (genital melanotic macules).
These lesions are benign, but tissue diagnosis is of importance to distinguish them from mucosal melanomas, which can demonstrate a deceptively banal clinical appearance.

**Acquired Melanocytic Naevus**
Acquired melanocytic naevi consist of aggregates of benign naevus cells. Junctional naevi consist of nests at the dermoepidermal junction, compound naevi also show dermal naevus cells and intradermal naevi involve the dermis only. Clinically distinctive variants include the halo naevus, which demonstrates a depigmented halo due to the initiation of inflammatory regression, and the Meyerson naevus, which shows an eczematous halo.

**Spitz Naevus**
Spitz naevus is a lesion occurring predominantly in children and adolescents, which despite some histologic resemblance to malignant melanoma, behaves in a benign fashion. Clinically, Spitz naevi are usually pink, red or reddish brown papules or nodules that demonstrate rapid growth over 3 to 6 months and then may remain stable for years. The face and lower limbs are common sites.
Histologically, Spitz naevi are composed of plump epithelioid and/or spindle cells, and display symmetry, maturation, Kamino bodies and lack of pagetoid spread of single melanocytes.

Fig. 1: Spitz naevus
Large dermal cells resembling melanoma.
Though there are criteria to distinguish Spitz naevi from melanoma, there are some lesions that show features at odds with the classical picture and are designated atypical Spitz naevi/atypical Spitz tumours and are considered to be of uncertain malignant potential. As with all Spitz naevi, they should be completely excised, with margins as for melanoma if sufficiently atypical, and followed up clinically. It is worth reiterating that melanomas may be misdiagnosed as Spitz naevi and vice versa, even by experts, and that some Spitz lesions defy accurate diagnosis by current methods.
Pigmented Spindle Cell Naevus

Pigmented spindle cell naevus (Reed naevus), which is no longer considered a variant of Spitz naevus, is a benign naevus that presents particularly on the thighs of young females as a darkly pigmented lesion.

Congenital Melanocytic Naevus

Congenital melanocytic naevi occur in 1% of all newborns, and congenital naevus-like naevi/early-onset naevi develop by the age of two years and may be even more common. Small (less than 1.5 cm) and medium-sized (between 1.5 and 19.9 cm) congenital naevi probably have a slightly increased risk of malignant degeneration when compared to acquired naevi, although the degree of the risk is controversial. Studies have shown a risk from 1 to 8%, with a recent review finding that it is 0.7%.

A rate of malignant degeneration of 5% has been estimated for giant (over 20 cm) congenital naevi and in one study 50% of melanomas occurred before puberty. The issues of surgical management and clinical follow-up, therefore, require early specialist consultation.

Dermal Dendritic Melanocytic Lesions

Dermal dendritic melanocytic lesions are derived from melanocytes whose embryonic migration from neural crest to epidermis has arrested in the dermis. The prototypic lesion is the blue naevus, its colour attributable to the site of pigment deep within the dermis.

The common blue naevus is a small blue or blue-black macule or papule occurring at any site. The cellular blue naevus is a larger nodular lesion occurring particularly on the buttocks, extremities, scalp and dorsal aspects of the hands and feet. The Mongolian spot is the most common congenital dermal melanocytic lesion.

These lesions are all benign and the major pathologic issue that (rarely) arises is separation from the rare malignant blue naevus and blue naevus-like metastatic melanoma.

The deep penetrating naevus is regarded by some experts as a variant of blue naevus. It is heavily pigmented, usually occurs in young adults and may have histological features that overlap with Spitz naevus and melanoma.

Dysplastic Naevus

Dysplastic (or Clark’s or atypical) naevi, found in 2-18% of the population, are larger than ordinary naevi (over 5 mm in diameter) and have irregular margins and variegated colour. They may be sporadic or familial, and are regarded as intermediate lesions of tumour progression between naevi and melanoma. Though dysplastic naevi are reported in contiguity with 1/3 or more of superficial spreading melanomas, most dysplastic naevi are stable and rarely progress to malignancy. They are a marker of increased risk of melanoma.

Dysplastic naevi have distinctive histology, though it must be noted that not all clinically atypical naevi show these features. The histology comprises random cytological atypia, architectural atypia with lentiginous hyperplasia and a stromal response (Fig. 2).

Fig. 2: Dysplastic naevus

Random cytological atypia, variation in size, shape and placement of nests of naevus cells, fibrosis and inflammation of stroma.

Most pathologists grade the atypia into mild, moderate and severe and, while the interobserver concordance may be low, a study has shown an increasing probability of a personal history of melanoma with increasing grade (Arumi-Uria et al).

Dysplastic naevi should be excised if they are clinically suspicious. There is no place for removal of all naevi in dysplastic naevus syndrome. Partial biopsy is not recommended since 1/3 of dysplastic naevi are heterogeneous in their atypia. There is not general agreement about treatment of incompletely excised dysplastic naevi, but a survey of American dermatologists indicated that 2/3 would re-excise them. Crowson et al state that the treatment of incompletely or closely excised mildly dysplastic naevi should be re-excision only if there is a residual lesion clinically, of severely dysplastic naevi should be a 5 mm margin, and are equivocal about moderately dysplastic naevi (which should probably be conservatively excised).

Lentiginous Dysplastic Naevus of the Elderly

This naevus (which some experts designate as ‘lentiginous melanoma’) occurs on the back in males, and on the legs in females over 60. It is an important precursor, rather than marker, of melanoma and should be excised with a margin of 5 mm.

Other Atypical Naevi

Various other variants of melanocytic naevi may show atypia clinically or histologically that may be suspicious for melanoma. These include naevi of special sites (acral, genital, breast, ear, flexures, umbilicus and perianal region), irritated naevi, regenerating naevi (after incomplete excision or trauma), naevi in pregnancy (which may darken and show a slight increase in mitoses) and cellular nodules in congenital naevi. It is therefore important to provide clinical details since a degree of atypia is appropriate in some naevi and does not indicate malignancy.

References


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