

PIGMENTED LESIONS IN CHILDHOOD AND PREGNANCY

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Health&A's Annual Women's and Children's Health Update
2nd March 2019
Melbourne, Australia



PIGMENTED LESIONS IN CHILDHOOD AND PREGNANCY

OBJECTIVES OF THIS PRESENTATION

- To assist recognition of high risk skin lesions encountered in childhood and adolescents (and no risk & low risk lesions!)
- To outline changes that may occur in pigmented lesions during pregnancy, and the features and management of suspected melanoma in pregnant women

PIGMENTED LESIONS IN CHILDHOOD AND PREGNANCY

IMPLICATIONS FOR GENERAL PRACTITIONERS

- Pigmented skin lesions in children, adolescents and pregnant women often present the GP (and skin specialists and pathologists!) with diagnostic and management challenges

PIGMENTED LESIONS IN CHILDHOOD AND PREGNANCY

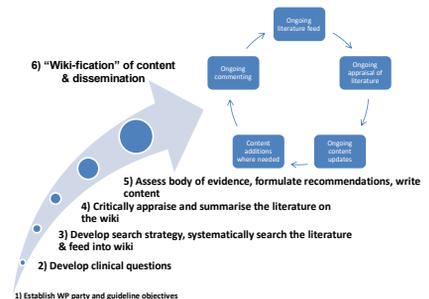
IMPLICATIONS FOR GENERAL PRACTITIONERS

- Most pigmented lesions presenting in these populations are benign but the possibility of melanoma must always be considered
- It is therefore important for the GP to have a thorough understanding of the more common pigmented lesions in childhood, adolescence and pregnancy and recognise suspicious features that should prompt further action

PIGMENTED LESIONS IN CHILDHOOD AND PREGNANCY

- This topic (and many others) are covered in new evidence-based Melanoma Management Guidelines
- These guidelines were produced by a working party convened by Cancer Council Australia
- On a Wiki platform, so can be readily accessed electronically, and easily updated

THE WIKI WAY OF DOING GUIDELINES



HOW TO ACCESS THE NEW NATIONAL GUIDELINES ?

- Access the guidelines at:

wiki.cancer.org.au/Australia/Guidelines:Melanoma

(Email guidelines@cancer.org.au to be notified of updates)

PIGMENTED SKIN LESIONS IN CHILDHOOD AND ADOLESCENCE

- Pigmented lesions in childhood and adolescence can be either congenital or acquired
- Acquired pigmented lesions most often develop in fair-skinned individuals, frequently as a result of sun exposure and/or use of tanning beds

CONGENITAL MELANOCYTIC NAEVI

(small or medium size - <2% of body surface area)

- Present in approximately 1 in 100 newborns
- Risk of malignant transformation very low (<<1%)

GIANT CONGENITAL NAEVI

- Rare (1 in 20,000 newborns)
- Defined as involving >2% of total body surface area (projected adult size >20cm)
- Most commonly on the trunk, have a verrucous surface with irregular pigmentation and often have associated satellite melanocytic lesions
- May extend into the dermis, subcutaneous tissues and muscle, making surveillance for malignant transformation extremely difficult

GIANT CONGENITAL NAEVI

- Lifetime risk difficult to quantify, but a large systematic review reported the development of melanoma in 2% of children with GCN
- Malignancy most often occurs in GCN >40cm (74%) and in those with satellites (94%). The highest risk for transformation is during childhood
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- These melanomas are not limited to the skin, and may develop viscerally or manifest as primary tumours within the central nervous system in association with neurocutaneous melanosis
- Lifelong surveillance of GCN is of critical importance

ACQUIRED PIGMENTED SKIN LESIONS

FRECKLES

- Common in fair-skinned children
- Benign aggregations of melanocytes
- Completely flat and impalpable

ACQUIRED PIGMENTED SKIN LESIONS
SIMPLE MELANOCYTIC NAEVI
(COMMON "MOLES")

- Benign proliferations of melanocytes
- Usually appear in childhood in sun-exposed areas
- Can increase in size during puberty

ACQUIRED PIGMENTED SKIN LESIONS
SIMPLE MELANOCYTIC NAEVI
(COMMON "MOLES")

- Intradermal naevi
- Junctional naevi
- Compound naevi

ACQUIRED PIGMENTED SKIN LESIONS

- The majority of melanomas in all age groups arise without an associated naevus
- HOWEVER
- Both size and number of acquired melanocytic naevi have been linked to melanoma risk

DYSPLASTIC NAEVI

- Acquired atypical pigmented lesions
- Display some of the more conventional features of melanoma, (asymmetry and colour variegation)
- Do not meet the diagnostic threshold for a clinical diagnosis of melanoma

DYSPLASTIC NAEVI

- Histologically - architectural disorder and cytological atypia (absent in common acquired and congenital naevi)
- Lack features of malignancy that occur in melanomas

BLUE NAEVUS

- An uncommon variant of dermal naevus that can often be worrying to the uneducated eye
- Typically well-circumscribed, dome-shaped lesions
- Usually <10mm in diameter, on the dorsum of extremities, scalp and buttock in children or adolescents

BLUE NAEVUS

- Blue appearance due to pigment in the dermis with preferential scattering & reflection of blue vs red light (the "Tyndall effect")
- Can be confused with the "blue-white veil" seen on dermoscopic evaluation of some melanomas
- Histologically, composed of pigmented dendritic dermal melanocytes
- Rare transformation into melanomas has been reported, most commonly on the scalp

NAEVUS SPILUS

(SPECKLED NAEVUS, SPECKLED LENTIGINOUS NAEVUS)

- May be congenital or acquired
- Appears as a collection of pigmented macules and papules on a background of hyperpigmentation
- Can be up to 20cm in diameter
- May be distributed in a dermatomal pattern
- Malignant transformation rare

HALO NAEVUS

- An acquired (usually compound) naevus which develops a surrounding symmetrical "halo" of depigmentation
- Mainly occurs in adolescents
- Can be a source of concern both for parents and practitioner, as halos can also appear around melanomas (but usually in an asymmetrical fashion)

HALO NAEVUS

- Is a benign process, and over time (usually within 12–18 months) the central naevus can regress and the depigmented skin regain normal pigmentation
- Histologically, a dense lymphocytic infiltrate is seen associated with a common acquired naevus with bland, mature features

MEYERSON NEVUS

- A benign melanocytic naevus with an eczematous halo
- The eczema usually resolves within weeks (much quicker if topical steroid used!)
- The residual naevus does not require removal

SPITZ NAEVUS

- Benign - usually appearing in the first decade as a solitary, well-circumscribed, pink or lightly pigmented papule <1cm in diameter
- Most often on head, neck or an extremity
- Histologically, overlap with some features of melanoma, making accurate diagnosis difficult - however when the constellation of histological features is present in an appropriate clinical context, Spitz naevi can be confidently diagnosed

SPITZ NAEVUS

- Lesions with some histological features of Spitz naevi but also more concerning atypical features are often referred to as "atypical Spitzoid tumors"
- These are categorised as "melanocytic tumours of uncertain malignant potential" (MeiTUMPs)
- Assessment by an experienced dermatopathologist is highly desirable

PIGMENTED EPITHELIOID MELANOCYTOMAS

- Most commonly arise in children, adolescents and young adults
- Dermal lesions that may mimic melanoma clinically and pathologically
- Histologically, they have infiltrative borders and are composed of large, heavily pigmented epithelioid and spindled melanocytes

PIGMENTED EPITHELIOID MELANOCYTOMAS

- Frequently spread to regional nodes, but the clinical course is indolent
- Regarded as borderline or low-grade malignancies
- Should be managed like other MeiTUMPs or melanocytomas

"MeiTUMPS" ("MELANOCYTOMAS")

- For some melanocytic tumours with atypical histological features, it can be difficult or impossible (even for acknowledged experts) to predict biological behaviour from pathological assessment
- Such pigmented lesions in children and adolescents are ultimately designated as "MeiTUMPs" or "melanocytomas"

"MeiTUMPS" ("MELANOCYTOMAS")

- Because of the lack of diagnostic certainty, the histology of these lesions should be reviewed by an expert dermatopathologist, and the patients managed at a high-volume referral centre
- The lesions should be excised with at least 5mm surgical margins, and the decision to perform a sentinel lymph node (SLN) biopsy made on a case-by-case basis

MELANOMA IN CHILDHOOD AND ADOLESCENCE (CAM)

- Rare - the most recently reported melanoma incidence rates in Australia were:

Childhood melanoma (age 0-14 years)

2.0 per million per year (= 50/year)

Adolescent melanoma (age 15-19 years)

24.3 per million per year (= 600/year)

CHILDHOOD AND ADOLESCENT MELANOMA (CAM)

PRESENTATION AND DIAGNOSIS

- Making the diagnosis of CAM can be difficult because they often do not present with the classic features of adult melanomas
- When compared to adults, more young melanoma patients have a family history of melanoma and atypical naevi, suggesting an underlying genetic driver
- However, recent genomic data indicate that UV radiation damage still plays an important aetiologic role in CAM (solar UV & tanning beds)

CHILDHOOD AND ADOLESCENT MELANOMA (CAM)

PRESENTATION AND DIAGNOSIS

- A high index of suspicion is necessary when evaluating young patients with multiple or atypical naevi
- Clinically, melanomas arising in young patients, especially pre-pubertal children, do not always follow the ABCDE detection criteria used for adults
- Rather, they often present with amelanotic, papulo-nodular lesions that are commonly of a single colour.

CHILDHOOD AND ADOLESCENT MELANOMA (CAM)

PRESENTATION AND DIAGNOSIS

- Due to their non-conventional appearance, the differential diagnosis includes juvenile xanthogranuloma, pyogenic granuloma, haemangioma and atypical melanocytic lesion
- It has been suggested that the conventional ABCDE criteria used in adults are inadequate in children and alternative ABCDE criteria have been proposed

CHILDHOOD AND ADOLESCENT MELANOMA (CAM)

ALTERNATIVE ABCDE CRITERIA

	Conventional diagnostic criteria in adults	Proposed diagnostic criteria in children
A	<u>A</u> symmetry	<u>A</u> melanotic
B	<u>B</u> order irregularity	<u>B</u> leeding or <u>B</u> ump
C	<u>C</u> olour variegation	<u>C</u> olourless or <u>C</u> olour uniformity
D	<u>D</u> iameter >6mm	<u>D</u> e novo, or <u>D</u> iameter <6mm
E	<u>E</u> volution	<u>E</u> volution

Adapted from: Cardaro et al. Pediatric melanoma: results of a large cohort study and proposal for modified ABCD detection criteria for children. J Am Acad Dermatol 2013 Jun;68(6):913-25.

CHILDHOOD AND ADOLESCENT MELANOMA (CAM)

PRESENTATION AND DIAGNOSIS

- For suspicious pigmented lesions in children, as in adults, an initial excision-biopsy with narrow margins should be performed for diagnostic purposes
- Pathological assessment by an experienced dermatopathologist is recommended, as pigmented lesions, especially with Spitzoid characteristics, can be extremely difficult to characterise as benign or malignant

CHILDHOOD AND ADOLESCENT MELANOMA (CAM)

DIAGNOSIS

The use of sophisticated tests is increasing in an effort to improve characterisation of atypical lesions of uncertain malignant potential:

- fluorescence in situ hybridisation (FISH),
- comparative genomic hybridisation (CGH)
- gene expression analysis

CHILDHOOD AND ADOLESCENT MELANOMA (CAM)

MANAGEMENT

- Surgical excision remains the mainstay of treatment
- Excision margins should parallel those for adults, based on Breslow thickness
- Consideration of SLN biopsy in children and adolescents is also as in adults, based on prognostic features of the primary tumour (i.e. thickness, ulceration, mitotic rate)
-

CHILDHOOD AND ADOLESCENT MELANOMA (CAM)

PROGNOSIS

- The prognosis for children is better than for older patients with comparable pathological features
- Improved outcomes are seen when CAM patients receive initial surgical care at specialised centres

PIGMENTED LESIONS IN PREGNANCY

- Physiological changes in melanin production (due to oestrogen stimulation and higher-than-normal levels of MSH) cause common pregnancy-induced pigmentary phenomena (melasma on the face, linea nigra on the abdomen)
- While enlargement and darkening of melanocytic naevi occur in more than 10% of pregnancies, primarily in the first trimester, the rate of malignant transformation is not increased

PIGMENTED LESIONS IN PREGNANCY

- Naevi excised from pregnant patients can display histological atypia and/or increased mitotic activity
- The pathology request form should clearly state that the patient is pregnant or has given birth in the last 3-6 months
- In women with the dysplastic naevus syndrome, clinical change in naevi was 3.9 times higher in pregnancy than when non-pregnant

PIGMENTED LESIONS IN PREGNANCY

- Traditional criteria for dysplasia, both clinical and dermoscopic, or the appearance of new, clinically-concerning pigmented lesions, should not be attributed to gestational changes and should instead prompt immediate biopsy

MELANOMA IN PREGNANCY

- Pregnancy-associated melanoma (PAM) is defined as a diagnosis of melanoma made during pregnancy or in the first 12 months post-partum
- Melanoma is the most common malignancy in pregnancy, accounting for 1/3 of pregnancy-associated malignancies in Australia

PRESENTATION AND DIAGNOSIS

- PAM often presents a clinical challenge
- Diagnostic delays may account for increased thickness of PAMs compared to controls
- Changing naevi in pregnancy should be monitored closely and those that meet the conventional ABCDE criteria should undergo excision-biopsy to allow complete histological evaluation

MANAGEMENT

- Melanocytic lesions in pregnant women should be managed as in non-pregnant females
- Surgery remains first line treatment, if possible delayed until the second trimester to avoid the small increased risk of miscarriage or foetal damage from anaesthetic agents in the first trimester

MANAGEMENT

- Pregnancy is not a contraindication to SLN biopsy, and indications are the same as for non-pregnant patients
- Lymphoscintigraphy with technetium99 is safe in pregnancy
- Blue dye should be avoided because of a possible risk of teratogenicity and the small risk of anaphylaxis

MANAGEMENT

- Transplacental metastasis is exceedingly rare but careful routine gross and histological evaluation of the placenta is recommended

PROGNOSIS AND MANAGEMENT

- The prognostic significance of pregnancy on a melanoma diagnosis is uncertain
- Given the infrequent incidence of PAM and the need to provide care for both mother and foetus, these patients should be managed at a specialist melanoma referral centre

CONCLUSIONS ⁽¹⁾

- The assessment and management of pigmented skin lesions in children, adolescents and pregnant women can be challenging
- A sound knowledge of the more common pigmented lesions and worrying features in them is essential for the general practitioner

CONCLUSIONS ⁽²⁾

- **Early diagnosis and referral of patients with high-risk lesions to an appropriate specialist service is key to improving outcomes**