Dr Trevor W. Beer
MBChB FRCPath FRCPA
Dermatopathologist & Pathology Reviewer for WAMAS
Clinipath Pathology
Osborne Park

Melanoma and Spitz Naevi in Children

Melanoma is rare in childhood, representing no more than 2% of all skin melanomas. Diagnosis is often delayed because melanoma is unsuspected, partly due to differences in presentation and its rarity. The diagnosis is made with trepidation by pathologists, since the vast majority of childhood skin lesions are benign.

Establishing the true prevalence of juvenile melanoma is complicated by a number of factors, one being the definition of childhood or juvenile. Many studies use a cut-off age of 19 years, but this is not consistent. Cancer registry data may also be unreliable due to misclassified Spitz naevi, for example.

ABCDE + ABCDD

ABCDE criteria help to identify adult melanoma clinically (Asymmetry, Border irregularity, Colour variation, Diameter >6mm, Evolution). Many melanomas in childhood are non-pigmented (> 50% of 65 recent cases from Victoria). Additional ABCD criteria have been suggested in children: Amelanotic, Bleeding bump, Colour uniformity, De novo, any Diameter to facilitate earlier recognition. Children more often present with advanced disease due to diagnostic delay, reported in 50-60% of patients.

Childhood Melanoma in WA

A WA study (in press, *Am J Dermatopathol*) with the WA Melanoma Advisory Service (WAMAS) identified 95 melanomas in patients 19 years or younger over a 14-year period. Three patients died from melanoma. The majority of tumours, 75%, occurred between ages 13 and 19 years, similar to other studies. In all populations, juvenile melanoma is much less common before puberty.

Delayed diagnosis was evident with 21 of 23 patients presenting with Clark level 4 or 5 melanomas with Breslow thickness >1mm in 65%. A family history of melanoma was seen in 17%. A study this year in Victoria revealed 65 melanomas during a 19-year period, with seven fatalities. A decrease in juvenile melanoma has been seen in WA despite an increasing population. In Queensland, a substantial decline occurred between 1997 and 2010, attributed to safer sun exposure practices since the 1980s.

Prognosis

The outlook for childhood melanoma mirrors that in adults being primarily based on stage and Breslow thickness. The exceptions are tumours with spitzoid features. These show appearances resembling Spitz naevi and, although often metastasizing to local nodes, are less frequently lethal. Prognostication is complicated by the fact that occasional examples behave aggressively and some spitzoid melanomas in case series may be misdiagnosed Spitz naevi.

Even in children, the possibility of melanoma needs to be considered.

Treatment

Complete excision, as in adults, is required. Further investigations and treatment should be decided in conjunction with expert advice. Treatment of advanced melanoma is progressing rapidly and possible eligibility for clinical trials means that patients will get the best opportunities for positive outcomes with personally tailored, up-to-date guidance.

It is recommended that all melanoma diagnoses in childhood are reviewed by pathologists with expertise in melanocytic lesions. Referral to a multidisciplinary team (such as WAMAS) is valuable to ensure correct diagnosis and to optimise treatment and advice to patients and families.

Sophie Spitz's 'Juvenile Melanoma'

Pathologist Sophie Spitz described 12 unusual 'juvenile melanomas' in 1948. Follow-up showed benign behaviour, despite microscopy suggesting melanoma. These lesions are now called Spitz naevi. Although typically childhood lesions, they can occur in adults, reducing in frequency with age.

Diagnosis clinically and microscopically can be challenging. Lesions may clinically resemble pyogenic granulomas, haemangioma or dermatofibroma. While most are correctly identified pathologically, some are misdiagnosed as melanoma. Conversely, a leading cause of litigation in pathology is under-diagnosis of melanoma as Spitz naevus.

Distinction between Spitz naevi and melanoma may be extremely difficult or even impossible. In these histologically ambiguous tumours inter-observer agreement is notoriously poor. Even panels of international experts have demonstrated limited diagnostic conformity with agreement, at times, little better than by chance. There is now a move away from traditional benign versus malignant divisions with suggestions that a histological continuum exists between Spitz nevi at one end and Spitzoid melanoma at the other. Indeterminate lesions may be labelled Spitzoid tumours of uncertain malignant potential (STUMP) with a guarded prognosis.

Improving pathological diagnostic accuracy

Treatment and prognostication of childhood melanoma requires accurate diagnosis which can be enhanced by experience and consultation between pathologists. Molecular studies such as FISH and aCGH may assist in ambiguous cases, but such methods are still in development and not uniformly available. However, molecular techniques will ultimately lead to more accurate diagnosis, prognostication and tailored treatment for children with melanoma.

Because of the rarity of childhood melanoma, individual clinicians or pathologists may have little experience in their recognition or treatment. WAMAS provides a centralised clinical and pathological approach to melanoma, bringing together expertise and experience, so it is regrettable that its future is in jeopardy.

References available on request.