Our Dermatology Online

Review Article

Melanocytic lesions and dermoscopy in childhood: diagnosis, therapy and foloving

Irdina Drljević, Edin Bjelošević, Amir Denjalić, Kenan Drljević

Faculty of Health of the University of Zenica, Zenica, Bosnia and Herzegovina

Corresponding author: Assoc. Prof. Irdina Drljevic, MD, PhD., E-mail: idrljevic@hs-hkb.ba

ABSTRACT

Early detection of malignant skin tumors, particularly malignant melanoma in childhood, which is most malignant of all, is of utmost importance for the prevention, treatment and outcome of the disease. If an observed lesion is characterized as melanocytic, the second step can be performed in the so called "*two-step dermoscopy*" algorithm, which includes the differentiation between a benign pigmented lesion (nevus) and a malignant lesion (melanoma). Gigantic nevi, as a subgroup of large congenital nevi, may pose a huge psycho-social burden for children and their parents, primarily because of potential complications. Melanoma can develop in healthy children but also in those with identified genetic disorder or immune diseases. Dermoscopy, at the same time, has reduced the number of unnecessary biopsies and excisions of benign melanocytic lesions in childhood.

Key words: Nevi; Melanoma; Dermoscopy; Childhood; Therapy; Foloving

Knowledge of pigmented lesions dates back to ancient times beginning with Hippocrates and Celsius, who called such outgrowths *naevi nigricans*. In the 16th century they were described by Russius Lauretius and then in 17th century by Highmore as tumors resembling coal. During the 20th century those tumors had different names: melanotic tumor, melanotic cancer, anthracite cancer etc. One of recent publications is the thesis by Bonnta in Lion (1911) titled "Melanosis and melanotic tumors". In 1967 an international body within the World Health Organization was established for the evaluation of diagnostic methods and treatment of melanoma, and under its auspices comparative clinical research has been conducted worldwide [1].

Moles (nevi) are confined, circumscribed inherited skin anomalies, occurring as a consequence of embryonic development [2]. Nevus cells appear in the shape of a nest on epidermal border and epidermis, and unlike melanocytes, they have no dendritic endings, tonofilaments and desmosomes. On the other hand, melanocytes and nevocytes produce the only endogenous autochthon skin pigment – melanin. The process of melanogenesis is monitored by genes, hormones and UV radiation [3].

Early detection of malignant skin tumors, particularly malignant melanoma, which is most malignant of all, is of utmost importance for the prevention, treatment and outcome of the disease. With the production of the first portable dermatoscope in 1958 by Leon Goldman a new era in the diagnostics of pigmented skin lesion and early detection of melanoma has begun [4].

Dermatoscopy (also known as dermoscopy, epiluminescence microscopy, surface microscopy, video dermoscopy) is a diagnostic method in dermatovenerology performed by covering of a skin lesion with mineral oil, alcohol or water and examination of the lesion with handheld dermatoscope, stereo microscope, camera or digital imaging system. It enables *in vivo* visualization of skin structures that cannot be seen with the naked eye such as epidermis, dermoepidermal borders and papillary dermis [5,6].

How to cite this article: Drljević I, Bjelošević E, Denjalić A, Drljević K. Melanocytic lesions and dermoscopy in childhood: diagnosis, therapy and foloving. Our Dermatol Online. 2016;7(1):97-100.

Submission: 29.07.2015; **Acceptance:** 12.11.2015

DOI: 10.7241/ourd.20161.25

In the past decades different epiluminescence microscopy (ELM) parameters have been set to differentiate melanocytic (moles, melanoma) from non-melanocytic lesions (basalioma, spinalioma, dermatofibroma, hemangioma and seborrheic keratosis), which is the so called first step in dermatoscopy.

If an observed lesion is characterized as melanocytic, the second step can be performed in the so called *"two-step dermoscopy"* algorithm, which includes the differentiation between a benign pigmented lesion (nevus) and a malignant lesion (melanoma). With that regard several different melanocytic algorithms have been established [7].

- Pattern analysis
- ABCD rule
- Menzies method
- 7-point checklist
- 3-point checklist
- ABC-point list

Classical dermatoscopic algorithm in the diagnostics of pigmented skin lesions is the so called *pattern analysis* set by Pehambergera et al. in 1987 and further modified and supplemented by Argenzian et al. in 2000 [8,9]. It is established on the analysis of numerous dermatoscopic structures, and based on the analysis of different colors and patterns, along with the so called *clue*, can lead to a specific diagnosis. The structures are often described metaphorically (e.g. *blue-whitish veil*) and they are difficult to translate into our language, for example, structureless blue zone vs. blue veil [10].

Melanocytic neoplasia in childhood can be divided into three classes: congenital moles (small, medium size, big and gigantic), acquired moles and melanoma. Congenital melanocytic nevi are found in 1-6% of newborns and are defined as benign neoplasia present already *in utero*. They include moles found during the birth but also tardive nevi, which become visible shortly after the birth. According to their size they are divided to small nevi, up to 1.5 cm in diameter, medium-size 1.5-19.9 cm and large ones of 20 cm and over [11].

Gigantic nevi, as a subgroup of large congenital nevi, are found in 1:500.000 newborns and they pose a huge psycho-social burden for children and their parents, primarily because of potential complications: melanoma, rhabdomyosarcoma, and manifestations of neurocutaneous melanocytosis. Nevus spilus and segmental speckled lentiginous nevi are also classified as congenital melanocytic lesions found in 0.2% of newborns. They need to be distinguished from agminated nevi, which lack brownish background. In early childhood sebaceous nevi composed mainly of fully formed sebaceous glands located in dermis are commonly found on the scalp. In around 30% of cases they can develop into basal cell carcinoma. From the time of birth or during early childhood a verrucous mole with hyperkeratotic surface, linear and/or bizarre shape located mainly unilaterally may occur. Its prognosis is good and malignant alteration is not known [12].

Specific dermoscopic structures, which can be found in congenital nevi include hypertrichosis, perifollicular hyper/hypopigmentation, multi-component pattern and structures that are found in seborrheic keratosis, so called *milia-like cysts* [13].

Melanoma incidence in children below the age of 14 is generally low worldwide, however, its increase has been evident over the past decades. Around a half of melanoma in children occur *de novo*, and a half from pre-existing melanocytic lesions [14].

Melanoma can develop in healthy children but also in those with identified genetic disorder or immune diseases. A special risk comes from the gigantic congenital nevus of over 50 cm in diameter, and a relatively high risk of malignant alteration exists for a small congenital nevus of 1.5-2 cm in diameter [15].

In terms of clinical but also dermoscopic examinations, pitfalls occur in vascular lesions (capillary and cavernous hemangioma) in childhood, particularly if they are thrombosed. In case of an injury of such lesions, they can be of livid and black color, they can bleed and be affected by a secondary infection and definitely resemble melanoma or pyogenic granuloma [16]. Differential diagnosis may also include angiokeratoma, which is a benign vascular lesion with dilated capillaries located below hyperkeratotic and acanthotic epidermis. Multiple angiokeratoma appear mainly before puberty and they are mostly symmetrically distributed on the skin of the scrotum, gluteus and periumbilically [17].

Furthermore, melanocytic nevi, which occur several months or years after the birth (so called acquired melanocytic nevi), such as the blue nevus (*naevus coeruleus vs. blue naevus*), Spitz and Reed *naevus*,

and Clark's nevus are a clinical, dermoscopic but also histopathological challenge for establishing a proper diagnosis.

Atypical vs. dysplastic nevi are defined as acquired melanocytic nevi with diameter over 6 millimeters, irregular borders and different colors, therefore, in terms of dermoscopy several types of such moles can be differed. Special attention should be paid to moles with an eccentric hyperpigmented patch or focally pronounced or prominent reticular pigment network [18].

Nevocellular nevus of *"Halo"* type (Sutton's *naevus*), which is clinically presented as depigmented halo around the junction mole, is relatively common in childhood. The mole in the center of the halo may completely disappear leaving behind a leucodermic patch resembling vitiligo. Children with a higher number of such lesions need to be observed and dermoscopic monitoring should be recommended [19].

A real challenge in clinical diagnosis is posed by hypopigmented and depigmented melanocytic lesions, because in everyday clinical practice, melanoma is rarely suspected in children, especially amelatonic or hypomelanotic type of tumors. Juvenile melanoma is a special form of a complex nevocellular nevus in children, which is most often located on the face. Clinically, it is recognized as a dome-shaped, red and brownish in color, with smooth surface and hard to elastic consistence. Although malignant alteration is very are, surgical treatment is recommended, i.e., full removal of the lesion and histopathological verification [20].

Neurocutaneous melanosis is a very rare disease in children manifested with rather large hyperpigmented areas located mainly in the bathing trunk site. It is described within the neurocutaneous syndrome, which involves nevocellular nevi on the brain, spinal cord and meninges. Having in mind symptoms originating from the central nervous system (e.g. convulsions) and the development of hydrocephalus at a later stage, the prognosis is very bad and there is no successful treatment. Malignant alteration is possible [21].

A special risk is found in children with a higher number of moles (more than a hundred) and atypical nevus syndrome, especially in case of a hereditary AMS type (a*typical mole syndrome*). Anatomic site of the moles and child's phototype are of special importance for several reasons. Namely, regions exposed to light as well as sites of permanent or occasional mechanic irritation are given special importance, particularly in terms of recommendations for prophylactic excision biopsy. On the other hand, dermoscopically, such lesions have special algorithms for recognition because they belong to so called *specific-site lesions*, which particularly involve lesions of the face, hairy part of the head, palms, soles and nails as well as all *milky-line lesions* affecting *mammae* areola, umbilicus, vulva and penis [22].

Dermoscopy, as a non-invasive and pain-free diagnostic procedure, has given a new morphological dimension to nevi, their dermoscopic monitoring and as well as early detection of atypical melanocytic lesions and melanoma in childhood. At the same time, it has reduced the number of unnecessary biopsies and excisions of benign melanocytic lesions in childhood.

REFERENCES

1. Putnik M. Istorijat U. Putnik M. Maligni melanom kože-klinika i terapija. Beograd: Institut za onkologiju i radiologiju, 1979:16-7.
2. Markovic SN. Erickson AL, Rao DR, Weenig RH, Pockaj BA, Bardia A, et al. Malignant Melanoma in the 21st Century, Part1: Epidemiology, Risk Factors, Screening, Prevention, and Diagnosis. Mayo Clin Proc. 2007;82:364-80.
3. Drljeviæ I. Influence of detrimental radiation of sunrays on an increase in malignant melanoma and non-melanoma malignant skin tumors (master thesis). Sarajevo: Sarajevo University; 2006.
4. Goldman L. A simple portable skin microscope for surface microscopy. Arch Dermatol. 1958;78:246.
5. Bahmer FA, Fritsch P, Kreusch J, Pehamberger H, Rohrer C, Schindera I, et al. Terminology in surface microscopy. J Am Acad Dermatol. 1990;23:1159-62.
6. Marghoob AA, Braun RP, Kopf AW. Atlas of dermoscopy. New York: Taylor Francis; 2004.
7. Marghoob AA, Braun RP, Kopf AW. Atlas of dermoscopy. New York: Taylor Francis; 2004.
8. Pehambereger H, Steiner A, Wolff K. In vivo epiluminiscence microscopy of pigmented skin lesion. I. Pattern analysis of pigmented skin lesions. J Am Acad Dermatol. 1987;17:571-83.
9. Stolz W, Riemann A, Cognetta AB, Pillet L, Abmayr W. ABCD role of dermoscopy: a new practical method for early recognition of malignant melanoma. Eur J Dermatol. 1994;4:521-7.
10. Bahmer FA, Fritsch P, Kreusch J, Pehamberger H, Rohrer C, Schindera I, et al. Terminology in surface microscopy. J Am Acad Dermatol. 1990;23:1159-62.
11. Šitum M. Malignant Tumors of Skin and Soft Tissue. Medicus. 2001;10:237-44.
12. Tannous ZS, Mihm MC Jr, Sober AJ, Duncan LM. Congenital melanocytic nevi: clinical and histopathologic features, risk of melanoma, and clinical management. J Am Acad Dermatol. 2005;52:197-203.
13. Zalaudek I, Manzo M, Savarese I, Docimo G, Ferrara G, Argenziano G. The morphologic universe of melanocytic nevi. Semin Cutan Med Surg. 2009;28:149-156.
14. Ingordo V, Iannazzone SS, Cusano F, Naldi L. Reproducibility of

dermoscopic features of congenital melanocytic nevi. Dermatology. 2008;217:231-4.
15. Zalaudek I, Manzo M, Ferrara G, Manzo M, Savarese I, Argenziano G. Problematic melanocytic lesions in children. Expert Rev Dermatol. 2009;4:249-61.
16. Wolf I. Dermoscopic diagnosis of vascular lesions. Clin Dermatol. 2002;154:1108-11.
17. Garzon MC, Enjolras O, Frieden IJ. Vascular tumors and vascular malformations: evidence for an association. J Am Acad Dermatol. 2000;42:275-9.
18. Krengel S. Nevogenesis: new thougts regarding a classical problem. Am J Dermatopathol. 2005;27:456-65.
19. Inamadar AC, Palit A, Athanikar SB, Sampagavi VV, Deshmukh NS. Unusual course of a halo nevus. Pediatr Dermatol. 2003;20:542-3.
20. Ferrara G, Argenziano G, Soyer HP, Chimenti S, Di Blasi A, Pellacani G, et al.The spectrum of spitz nevi: a clinicopathologic study of 83 cases. Arch Dermatol. 2005;141:1381-7.
21. Bittencourt FV, Marghoob AA, Kopf AW, Koenig KL, Bart RS. Large congenital melanocytic nevi and the risk for development of malignanat melanoma and neurocutaneous melanocytosis. Pediatrics. 2000;106:736-41.
22. Elder DE. Precursors to melanoma and their mimics: nevi of special sites. Med Pathol. 2006;19:4-20.

Source of Support: Nil, **Conflict of Interest:** None declared.