Birthmarks classification, differential diagnosis and follow-up

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Nevus, birthmark or beauty mark in Czech, is a medical term that deserves a detailed treatise. This article will deal with the actual birthmark classification, the same as their differentiation from other melanocyte skin signs such as benign melanocytic spots but also malignant melanomas. It will also present the follow-up procedure recommended by dermatologists. The regular birthmark observation is the elementary precondition for an early diagnosis of its malignant transformation into a melanoma and an early therapeutic intervention.

In the clinical practice birthmarks are most times confused with so-called melanocyte spots (ephelides, melasma, cafe au lait spots and lentigines) caused by simple hyperplasia and hyper function of melanocytes, however, these lesions are totally benign. The other group birthmarks are frequently mistaken for are malignant melanomas. The late diagnosis of these if not revealed in time have fatal consequences for the patient.

Ephelides

Ephelides, freckles is a genetically conditioned congenital pigmentation that appears mainly in red-haired and light-haired individuals. They are predominantly found in central areas of a face, on arms, on the upper third of a chest or back. Freckles are sharply circumscribed maculas of circular, oval or of irregularly uneven shape, usually smaller then 5 mm. They are flat so undetectable to the touch. The colour varies from a yellowish shade through red to different shades of brown and they do not transform into a melanoma. Freckles are diagnosed by its clinic appearance, the histologic screening is not necessary. Freckles are just a cosmetic problem for the affected, usually they are not treated.

Melasma

Melasma, syn. chloasma ( obr. 2 ) is obtained melanin pigmentation most often found in middle-aged women. It is caused by the combination of UV radiation and hormone oscillation (pregnancy, taking
contraceptives). It appears as dark brown spots mostly in lateral areas of the face, lesions are completely benign. However, as they appear in visible areas, they are often treated for cosmetic reasons. The only effective therapy is their removal by means of a highly effective laser. The prevention is the only one; that is using photo-protective preparations.

**Café-au-lait spot**

A solitary café-au-lait spot appears approximately in 15% of inhabitants and forms in early childhood or is already present when born. In children it is on average 1-3 cm big, in adults it reaches even 20 cm and more. Café-au-lait spots are sharply bordered, even coloured light brown spots, because of their colour often compared to coffee with milk. During next life they have stable appearance. They do not cause diagnostic problems, they are benign and do not transform into melanomas. They frequently accompany other diseases like for example neurofibromatosis. For cosmetic reasons they can be removed again by a highly effective laser.

**Lentigo simplex**

*Lentigo simplex* is an interstage between a finding in a normal skin and a junctional melanocytic nevus. Lentigo simplex is an intense-brown, sharply circumscribed, oval or cicular macula, usually up to 5 mm big and usually individual, appearing in both sexes at any age and anywhere on the skin. Its colour does not depend on the sun exposure. It can appear even on palms, soles, nail beds and a mucous membrane. It can be differentiated from a junctional nevus only by histology. The therapy is not necessary, just for cosmetic reasons – it can be removed by means of a laser.

**Lentigo solaris (lentigo senilis, liver spot, age spot)**

*Lentigo solaris* appears mainly in older people the same as other signs of a chronic skin lesion caused by UV radiation, its typical location being the face and the back of hands. The deciding factor is not the age of the patient but a long-term, repeated exposure of the skin to UV radiation effects. In spite of this lentigo solaris appears mainly in people over 60 years old and preferably in those with lighter skin. The spots are from
several square milimetres to several square centimetres large, they are oval or irregular, sharply circumscribed, light or even dark brown. If the exposure to UV radiation is avoided, the spots turn pale. Lentigo solaris is a benign lesion. It can also appear in people who repeatedly go to sunbeds. For their potential removal for cosmetic reasons laser is indicated.

**Nevi**

Birthmarks are circumscribed skin lesions appearing due to an embryonic development disorder. From a practical point of view they have a big importance as 20-30% of cases clinically change during the life and in some cases can they transform into a malignant melanoma. The classification of birthmarks have gone through many changes in the last few years, however, I consider the below mentioned classification the most comprehensible. Regarding the great amount of kinds of birthmarks I am mentioning here just the most frequent ones. Birthmarks are divided into 3 basic groups: vascular, adnexal and pigmented.

**Vascular nevi**

Among the largest vascular birthmarks there is the naevus flammeus (syn. naevus vinosus, port wine stain) which is present already at birth and continues developing along with the child’s growth. Clinically it is a violet red, sharply circumscribed lesion with a slightly elevated surface of a size from several milimetres up to a large area covering a big part of the body. It appears very often in the face. For the patient it is cosmetically depressing but at present it can be easily removed by means of a special laser.

The second most well-known vascular birthmark is nevus araneus (spider nevus) of a typical look with a central spot capillary and ray extensions radiating outwards resembling a spider. The therapy consists either in sclerotization or in a laser therapy.

**Adnexal nevi**

The most common adnexal birthmark is naevus sebaceus. It is a congenital soft yellow-brown lesion of a papillomatosis surface of a size from 1 to 6 cm. It appears mostly in the scalp and gradually grows. Due to a danger of its transformation into a basal-cell carcinoma a laser excision surgery is recommended.
Pigmented birthmarks

Among pigmented birthmarks there is first a group of melanocytic birthmarks and their other histological sub-types and also defined clinical units: *Naevus spilus, Naevus Spitz, Halo naevus, Naevus coeruleus.*

**Melanocytic birthmarks**

*The melanocytic birthmarks* appear by accumulation of melanocytes (histological term – nests) in different areas of the skin (epidermis, corium or both) and according to that they are histologically divided into sub-types – junctional, intradermal or compound. They are either congenital (present immediately after the birth) or acquired (appear later, usually increasing until the age of 20-30).

*The congenital ones* are usually from 1 to several tens of cm and they are individual. The most well-known representative of the group is Naevus Becker.

*The acquired nevi* in adults very rarely exceed the size of 2 cm and are nearly always multiple. They usually start to appear from 6 months of age and their amount gradually increases until the age of 30. They can also appear later than that. According to different sources there can be found on average about 20-40 birthmarks in a thirty-year old white person. However, the amount is very variable. They are the genetic factors that have the decisive influence on the amount and clinical appearance of the birthmarks. Changes in melanocytic lesions in the childhood and puberty are physiological and their signs are changes in size, colour and form. After puberty such clinical changes in melanoctytic lesions can be a sign of a gradual development of a tumour.

*Junctional nevus* looks like a spot which cannot be found by means of palpation. The size varies from 1 mm (like a needle mark) to 10 mm in diameter on average, the colour is pink, dark brown to black, the surface is smooth.

*Compound nevus* The compound nevus has a semi-spherical shape (from papules to bumps), a rather dark brown colour with a smooth surface, just slightly wrinkled.
**Intradermal nevus** appear usually in the face, on the neck, chest and during adulthood they usually appear as semi-spherical, circumscribed papules or humps up to 1 cm in diameter, of verrucous look, often with hairs.

The riskiest sub-type includes the so-called dysplastic birthmarks. They are usually 10-15 mm large, unclearly circumscribed and unevenly pigmented. The centre is darker, the borders usually lighter. The diagnosis of a dysplastic birthmark is confirmed by histology. These dysplastic nevi are very difficult to distinguish from a melanoma. This kind of nevi most often turns into a malignant melanoma. A long-term and regular monitoring is crucial for this group.

**Naevus Spilus**
It is a large-size light brown pigmented spot with pinhead-size pigmentation scattered in it. Rarely a melanoma may appear in darker spots.

**Naevus Spitz** (earlier juvenile melanoma)
It is a completely benign, quickly growing (weeks) semi-spherical hump of reddish colour, about 1cm big, histologically reminds a malignant melanoma.

**Halo naevus**
There is a typical whitish up to 2 cm wide oval areola (halo) around the birthmark; it is a sign of an immunological reaction against melanocytes. It is recommendable to protect the light halo by UV protection preparations.

**Naevus coeruleus**
There is a typical whitish up to 2 cm wide oval areola (halo) around the birthmark; it is a sign of an immunological reaction against melanocytes. It is recommendable to protect the light halo by UV protection preparations.

**Differencial diagnosis of birthmarks - malignant melanoma**

A melanoma is a disease of the middle age, the number of patients mostly increases at the age from 40 to 50. The tumour is very rare in children before puberty. The melanoma prevalence in white people is
about 1/100 – 2/200. The melanoma appears more frequently in women (55% women, 45 % men) with a primary tumour especially in the face and lower limbs, whereas in men it is more usual on the chest. The melanoma also runs in families, above all in those with a higher appearance of different melanocyte diseases. Genetics plays a decisive role in the appearance of melanocyte lesions and the melanoma. Among the risky factors of melanoma appearance there are:
positive family anamnesis of the melanoma
a big amount of melanocyte lesions on the skin
a lighter type of skin
presence of a congenital birthmark.
The melanoma appears in genetically predisposed people together with a presence of different outer or inner factors; the most discussed among them being the effect of the UV radiation. In 2/3 of the ill the melanoma forms on clinically absolutely normal-looking skin. As in most adults the increase of other melanocytic symptoms does not continue (as frequent below the age of 30), the formation of the melanoma should not pass unnoticed by the patient or the doctor. In the resting 1/3 the melanoma forms in an already existing melanocytic lesion (in so-called melanoma precursors), e.g. in a congenital birthmark, lentigo maligna and different so-called dysplastic nevi. The ill should not ignore skin symptoms like the increase in shape, change of the colour, growth over the surrounding area level or bleeding. The ABCD rule is to be applied by means of which we watch the following changes:
A (Asymetry), B (Border), C (Colour), D (Diameter). The above mentioned changes of an originally stable melanocytic lesion can be a signal of a beginning melanoma formation.

A melanoma is according to the clinical and histopathological picture devided into 5 types:

- melanoma in situ
- superficial spreading melanoma (SSM)
- lentigo maligna melanoma (LMM)
- nodular melanoma (NM)
- Acral lentiginous melanoma (ALM)

The most common type is the superficial spreading melanoma (50-70%), the nodular melanoma forms about 15-30%, the lentigo malignant melanoma 5-10% and the acral lentiginous melanoma is rare (2%). The tumour tends to form metastases, most times in the local lymph nodes.
But a melanoma can metastasise into any organ, even many years after removing the primary tumour.

**Melanoma in situ**

It is usually a brown spot of several millimetres with a smooth shiny surface and it is unclearly bordered from the healthy skin. It does not have any specific diagnostic signs and can not be differentiated from benign melanocytic nevi. The diagnosis can be just histologic – the tumour changes are just in the epidermis and tumour melanocytes do not exceed the basement membrane into the dermis.

**Superficial spreading melanoma (SS)**

The most common form of melanoma during which an unevenly coloured, smooth, dark several centimetres big lesion appears. The tumour is mainly on the back and lower limbs but can be found anywhere on the body. In men it is most often fount on the back, in women on the lower legs. It looks like a small brown dot which gradually becomes bigger and changes until reaching the above described picture. Such development takes from several months to several years. The ill usually state that the growth is obvious 6-12 months before the diagnosis is made. There are first changes in the horizontal level and later some part gets easily hurt, does not heal and that is the reason why the ill go to see a doctor.

**Lentigo maligna a lentigo maligna melanoma**  
**(melanosis circumscripta preblastomatosa Dubreuilh)**

Lentigo maligna is a slowly growing unevenly brown pigmented lesion on the skin exposed to UV radiation, with a histologic picture melanoma in situ. A lentigo maligna melanoma represents a completely developed melanoma formed in such a lesion. Lentigo maligna forms almost exclusively on the face, more often in women than men (2 : 1), usually in older age (over 60 years old). A light brown, at first smooth, circumscribed patch without subjective difficulties appears on the skin. Gradually in many years it slowly grows and changes its colour so it is more visible and noticeably unevenly pigmented. In some areas there is regression, i.e. a certain defensive organism reaction, when the tumour disappears. The size of the tumour can be even several centimetres in diameter (2-5 cm). Lentigo maligna stays without changes for many years and then it transforms into a lentigo maligna melanoma when from
the histological picture of the melnana in situ a developed melanoma forms. This melanoma type is considered to be more prognostically favourable than other types.

**Nodular melanoma**

The melanoma appears as a gradually growing swelling, the vertical growth dominating. The tumour may appear anywhere on the skin starting like a small brown macula and changing into a papule and an elevated semi-spherical wide sessile swelling. The tumour surface is smooth and shiny, but when hurt it suppurates, bleeds and does not heal.

**Acral lentiginous melanoma**

A form of a melanoma that differs from the others just by the fact that it appears in the acral locations on palms, soles, fingers, nail beds and around nails and also on mucous membranes, above all of the mouth cavity and genitals.

The acral lentiginous form appears rather in older patients. If growing under a nail it looks at first like light bleeding and can cause big diagnostic difficulties. Among special forms of the melanoma there is the so-called amelanotic melanoma. The melanin pigment is missing there and it is practically impossible to make the diagnosis from the clinical picture. The lesion is rather reddish and impossible to be told apart from other tumours.

**Follow-up**

Although people are already aware of the necessity of having their changing pigmented birthmarks checked, just a small part of them realizes the importance of checking them over time. Many patients after the first check-up and assurance that it is not a tumour become less cautious and then do not consult their doctor any more. Nevertheless, the birthmark development and their malignant change can be detected in time just during regular check-ups. In the dermalogist’s examination the patient should be instructed about the necessity of regular examining and it is the best if the next appointment is immediately made. The next check-up is usually in one year, preferably after the summer season when the UV radiation affects birthmarks with the highest intensity. In case of a positive anamnesis or other risky factors it is necessary to shorten the time period to 6 months.
The most modern way how to diagnose birthmarks is by means of a digital dermatoscope. This device can in a non-invasive way, just by an intensive scan of the lesion, examine any pigmented formation, assess its risks comparing more than 50 characteristics and save the picture in its memory. During the check-up it can easily compare the picture from the last check-up with the actual examination and without any doubts will find out whether there has been a change or not. A change into a malignant tumour can be diagnosed in time then, which will significantly increase the chance of survival. Regarding its high cost it is available just in a few centres in the Czech Republic. An examination by a digital dermatoscope should be done by an experienced dermatologist who can interpret the results well and also judge other clinical aspects and decide on the adequate action.

The examination by a digital dermatoscope is done by the dermatologic centre Lasermed, Nedvědovo nám. 3, 147 00 Prague 4 – Podolí, Czech Republic. Tel. 734480916, www.lasermed.cz

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