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Dermoscopic findings of congenital melanocytic nevi. Hair component and multifocal hypopigmentation.

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Summary The dermoscopic findings of 78 congenital melanocytic nevi are examined. Multifocal hypopigmented areas are characteristic of these nevi and contribute to their polymorphism. These regularly distributed areas are shown in 31/78 nevi and often -18/31 nevi- are centered by hypertrophic hairs. This finding is more evident than in normal skin, probably due to the presence of hypertrophic hairs and to the contrasting peripheral pigmentation.

Key words Congenital melanocytic nevus, dermoscopy, multifocal hypopigmentation, hypertrophic hairs.

Some recent reports in pediatric dermatology (3, 7) were aimed at singling out the dermoscopic characteristics of congenital melanocytic nevi (CMN). To better understand the dermoscopic differences between acquired and congenital nevi, we should first underline their different clinical features and pathological findings.

From a clinical point of view, the congenital nevi, besides being present at birth, are characterized by greater changes of number, size, shape, surface, color and therefore by greater polymorphism. Moreover, congenital nevi are clinically characterized by associated hair alterations.

From a pathological point of view, besides their greater size, the epidermis of CMN is characterized by a verrucous or lentiginous hyperplasia, the latter being similar to that one of dysplastic nevi. Moreover, in the dermis the nevus cells are distributed superficially in a band-like pattern or infiltrate the whole dermis in the deeper and usually more extensive nevi. However, the nevus cells tend to deepen along the hair follicles, often hypertrophic, or to be distributed in single file among the collagen bundles.

We do not know very well the dermoscopic characteristics of the congenital nevi (1, 2, 3, 4, 5, 6, 7, 8). Among the latter, Braun et Al. (3) underline the presence of thicker and hyperpigmented hairs. Taking into account the clinical features, this finding is not surprising, whereas the presence of hypopigmented multifocal areas (1) is striking. According to other Authors (7, 8), the latter are regularly distributed around the hair follicles especially in the nevi with reticular pattern. Argenziano et Al. (1) in 17 congenital nevi showed multifocal hypopigmentation in 7 cases. In some cases the hypopigmented areas were clearly distributed around hypertrophic hairs, although the Authors did not mention this finding.

This report is aimed at verifying whether a relationship exists between the hypopigmented multifocal areas and the hypertrophic hairs in the congenital melanocytic nevi. The identification of these areas could be a useful sign to recognize the congenital nevi and to confirm their benign nature, in contrast with so many other

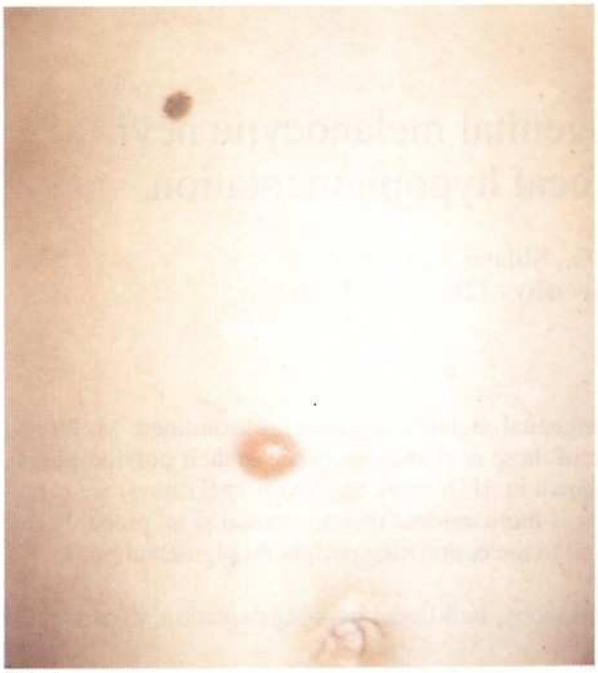

Fig. 1

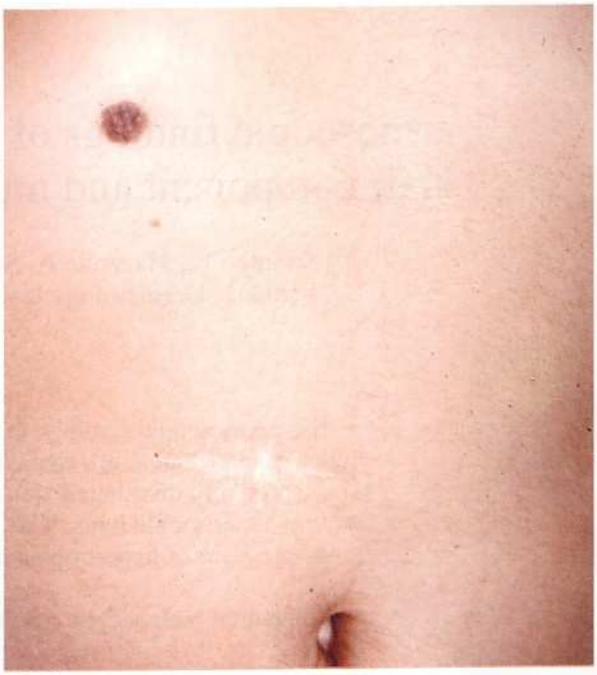

Fig. 2

Fig. 1, 2, 3: This little girl has two relatives with melanoma, one of whom living. She underwent removal of periumbilical dermatofibroma when aged 5. She also has a congenital melanocytic nevus, which you can see at the age of 5 (Fig. 1) and 10 (Fig. 2). Dermoscopy of the nevus at the age of 10 showed a polymorphic pattern, due to the presence of a typical net, regularly distributed dots and areas of multifocal hypopigmentation, which were centered by hypertrophic hairs (Fig. 3).

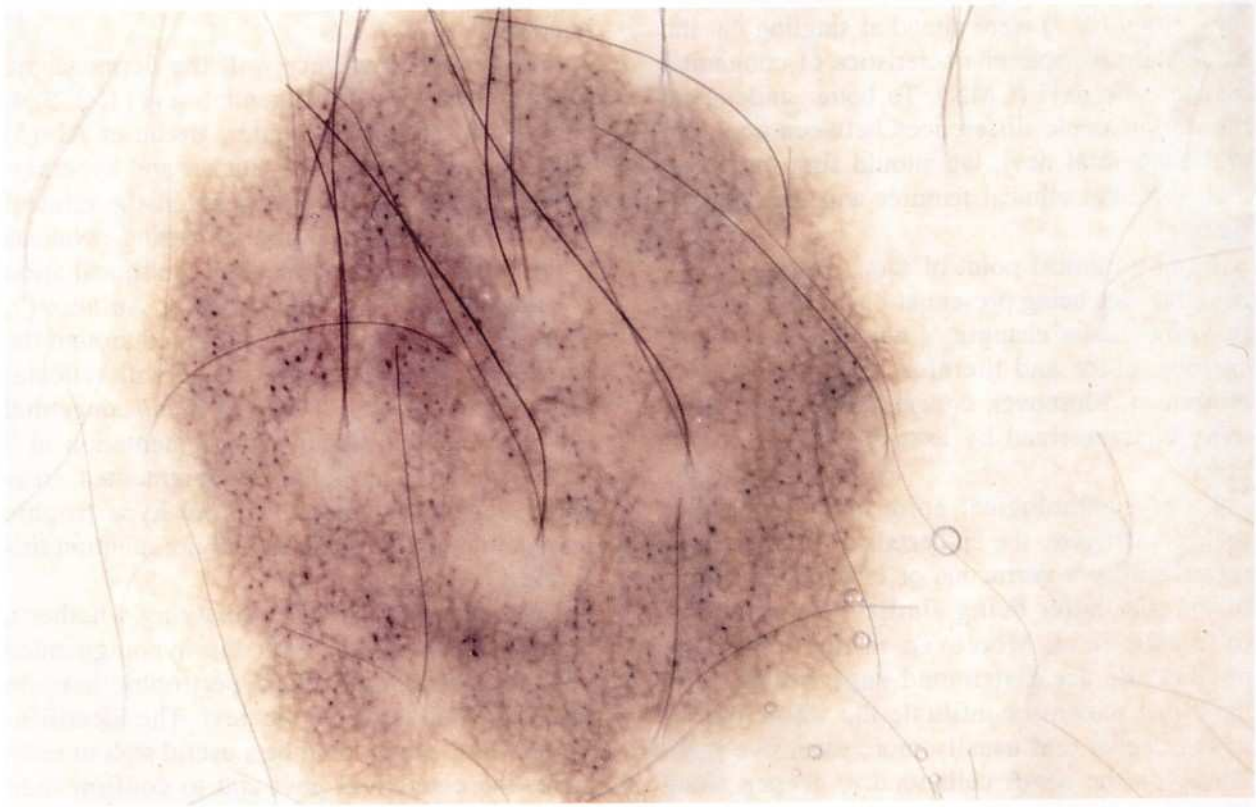

Fig. 3

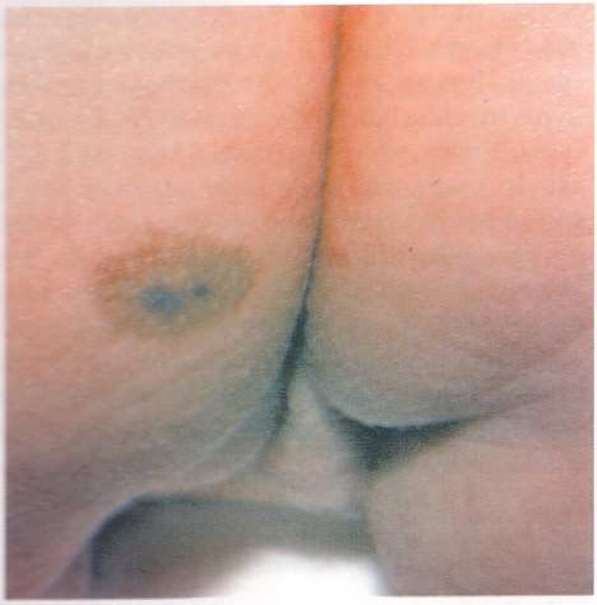

Fig. 4

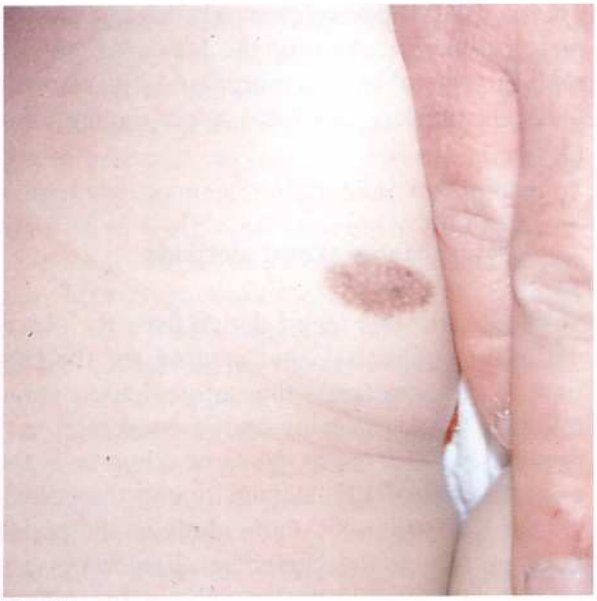

Fig. 5

Fig. 4, 5, 6: Congenital melanocytic nevus of the left buttock in a little girl at the age of 1 (Fig. 4) and 5 years (Fig. 5), when the hyperpigmented area disappeared. At the age of 5 dermoscopy (Fig. 6) showed a polymorphic appearance, due to the presence of irregularly distributed dots and globules, areas of multifocal hypopigmentation, which are devoid of hairs, focal hyperpigmentation and regressive structures level with the previously evident (Fig. 4) hyperpigmented area.

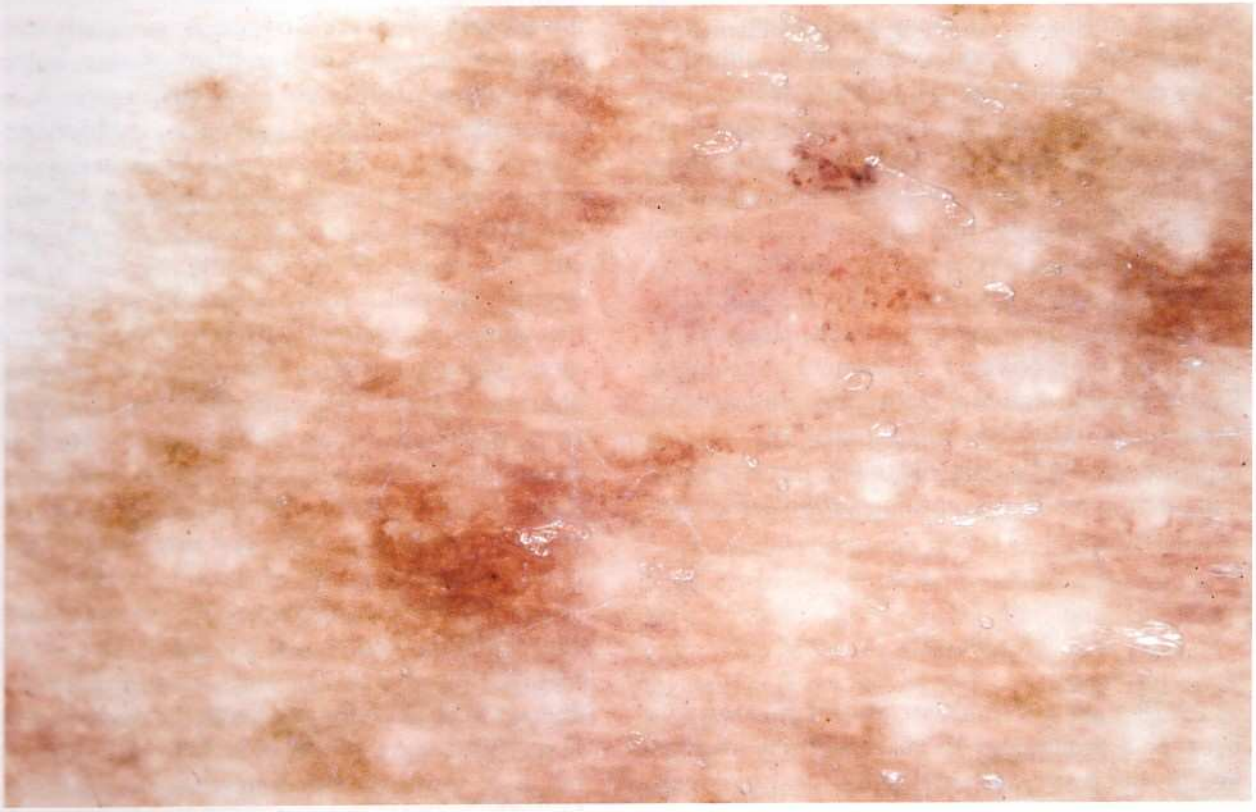

Fig. 6

criteria of CMN, which wrongly suggest a malignant behavior. Among the latter we should mainly remember polymorphism, but also the clear-cut borders and focal hyperpigmentation (3, 6).

Material and methods

The data of this report derive from the observation of congenital nevi coming for the first time to the outpatient clinic of pediatric dermatology of Bari university and of congenital nevi previously observed in the same clinic or in the neonatal unit of Bari university and then recalled for a control visit. Particularly in the period between March and November 2004 were visited 72 subjects, 32 males, whose age ranged between 3 days and 40 years, with an average age of 8.2 years, who comprehensively presented 78 congenital melanocytic nevi. The diagnosis of CMN was made thanks to the history of being present at birth and to the conventional objective datum of its size ->4mm in the first year, >1cm in the prepubertal age, >2cm in the postpubertal age-. Also the nevi >2cm in the first year were considered congenital, although being not present at birth by history taking. The nevi were first clinically observed and then they underwent dermoscopy with 10x magnification (Dermogenius, Rodenstock Prazisiosoptik, Munich, Germany) after interposing a drop of oil for dermoscopy (Heine, Germany). The dermoscopic images were acquired with an objective Dermaphot (Heine Optotechnik, Herrsching, Germany) on a Nikon F 70 camera and with a Kodak Ektachrome 100 ASA film. Slides were then scanned with a scanner for slides (SprintScan 4000, Polaroid). The dermoscopic data were classified according to the criteria of Agenziano et Al. (1).

Results

13/78 cases, 7 males, whose age ranged between 1 month and 13 years, with an average age of 4.5 years, presented multifocal hypopigmentation (MFHypo) without evident hairs inside (Fig. 6); 18/78 cases, 9 males, whose age ranged between 2.5 and 27 years, with an average age of 10 years, presented MFHypo centered by hypertrophic hairs (HyperH) (Fig. 3); 4/78 cases, 1 male, whose age ranged between 3 days and 16 years, with an average age of 6 years, presented HyperH without MFHypo and, finally, 43/78 cases did not present MFHypo nor HyperH. In 24 of these 43 cases the hairs could not be present because we were dealing with plantar (4 cases) or vulvar (1 case) region or because the global pattern was homogeneous or because there were gross exophytic papillar structures (3 cases).

Comment

We would like to underline first the significant changes met with by congenital nevi with years (Fig. 4, 5). Two nevi, one giant and the other one small, completely regressed. The most significant changes involved the color and the hair component of the nevus. With regard to the dermoscopic characteristics of the congenital melanocytic nevi, the first one immediately evident is a marked polymorphism of size, color, with mixing of different local findings such as net of pigment, dots and globules, cobblestone-like structures, sometimes striae, irregular hyperpigmented areas, hypopigmented areas, exophytic papillar structures, horny pseudocysts etc.. Moreover, CMN, especially thin CMN, frequently showed a hair component, which was, however, already present from a clinical point of view. The latter is much more frequent and thus characteristic of the congenital melanocytic nevi, although it can be rarely observed in acquired nevi. We should distinguish two conditions as follows: CMN of the scalp, which is one of the few sites with terminal hairs in the newborn, and CMN of other sites.

Level with the scalp, congenital nevus at birth can be well evident, barely visible or exclusively manifesting as a tuft of hairs of different color and texture as compared with the surrounding hair. With time the congenital melanocytic nevus of the scalp gets barely visible or invisible. When CMN is barely visible, one can only

see a tuft of hairs of different color and the different hairs can be darker or white. The latter usually appear later on and are probably expression of regression.

In CMN of other sites the hair component, namely the presence of terminal hairs, which are thicker and usually darker than the vellous hairs of the surrounding normal skin, is rarely evident at birth. It usually gets evident after the second year of life and significantly worsens the esthetical damage. Anyway, even when not evident at birth, the hair component is surely already determined at this age. This is why, when we remove partially by "curettage" or dermabrasion the congenital nevus, its hair component gets evident on the whole area previously affected by the nevus after a variable period of time, usually after 1 or two years.

The hair component is not present only in CMN, being also evident in Becker's nevus. In the latter the hair component gets more evident at puberty. From the dermoscopic point of view, Becker's nevus can show characteristics similar to CMN such as hypertrophic hairs and multifocal hypopigmentation. Becker's nevus can also show global characteristics similar to CMN such as a net of pigment (Fig. 8). However, Becker's nevus does not show the polymorphism of CMN, because hyperpigmented areas, globules, cobblestone-like structures, and focal hyperpigmentation of the net usually lack. This is not surprising, given the clinical monomorphism of Becker's nevus.

CMN can be more easily differentiated by hyperpigmented nevus, namely that nevus underreported in the relevant literature and with different names, which is the opposite of the hypopigmented nevus. In the hyperpigmented nevus there are not nevus cells, nor elongated epidermal crests and increased epidermal melanocytes. There is only increased melanic pigment within the epidermis. From a clinical point of view, its color is lighter than that one of melanocytic nevus and similar to that one of "café-au-lait" spots, with which is often mistaken, leading to the wrong suspicion of neurofibromatosis. It can be sometimes hardly differentiated from congenital melanocytic nevus in the first months of life, when some CMN show a light pigmentation. In these cases the dermoscopic observation can be useful, putting in evidence in CMN a net of pigment or dots, which usually lack in hyperpigmented nevus. In some cases of hyperpigmented nevus we showed a homogeneous hyperpigmentation, which was lighter of that one of CMN, but enough to give greater evidence to the normal rhomboid outline of the skin. Moreover, both hypertrophic hairs and multifocal hypopigmentation lack in hyperpigmented nevus, making the differential diagnosis from CMN and Becker's nevus easier.

Another dermoscopic finding is characteristic of CMN, namely a multifocal hypopigmentation. This finding, which is probably related to the hair component, was sporadically mentioned by other Authors (1, 7, 8). The hypopigmented areas are regularly distributed in congenital nevi, mainly in those ones barely infiltrated, in which the net of pigment and dots prevail. Their number, their regular distribution and mainly the presence in their center of hypertrophic hairs, immediately raises a relation with the hair component. This report was aimed at verifying the possible existence of a relationship between the multifocal hypopigmentation and the hair component. Its results showed that MFHypo with or without HyperH was present in 31/78 cases. However, in 24/43 cases the hairs lacked because we were dealing with the plantar (4 cases) or vulvar (1 case) region or because the global pattern was homogeneous (16 cases) or there were gross esophytic papillar structures (3 cases). In these cases both hypertrophic and multifocal hypopigmented areas lacked, confirming their relationship. Out of 31 cases with MFHypo and with or without HyperH, 18 showed a characteristic global pattern due to the presence of gross multifocal hypopigmented areas, which were regularly distributed on the entire nevus surface. These areas were centered by hypertrophic hairs (Fig. 3). In 13/31 cases the multifocal hypopigmented areas were evident, but hair structures were not visible (Fig. 6). Only in 4/78 congenital nevi there were hypertrophic nevi without hypopigmented areas.

The perifollicular multifocal hypopigmentation reproduces in a more marked way the outline of the normal perifollicular skin, which is

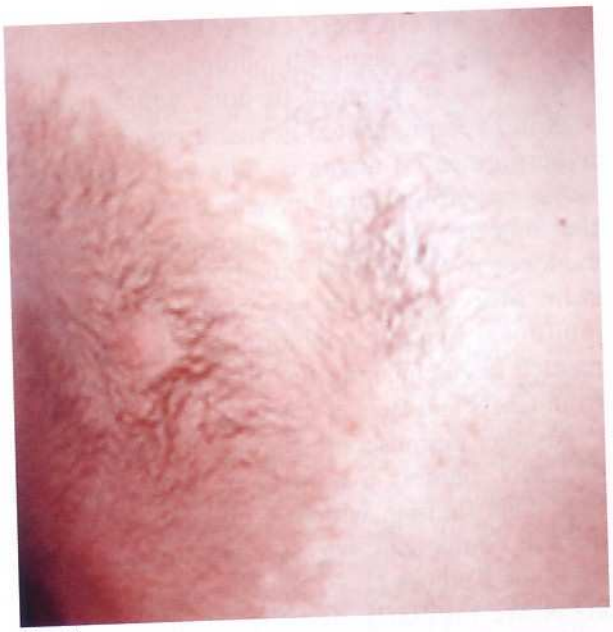

Fig. 7

Fig. 8

Fig. 7, 8: Becker's nevus of the right hemithorax (Fig. 7) in a 15-year-old boy. Dermoscopy (Fig. 8) showed a typical net of pigment, multifocal hypopigmented areas and hypertrophic hairs.

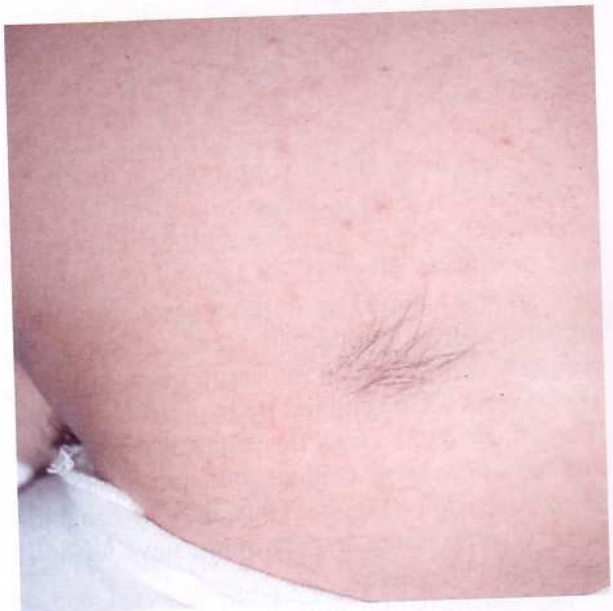

Fig. 9

Fig. 10

Fig. 9, 10: Hairy, non melanocytic nevus (Fig. 9) in a 2-year-old boy. Dermoscopy (Fig. 10) showed hypertrophic hairs coming out from hyperpigmented areas.

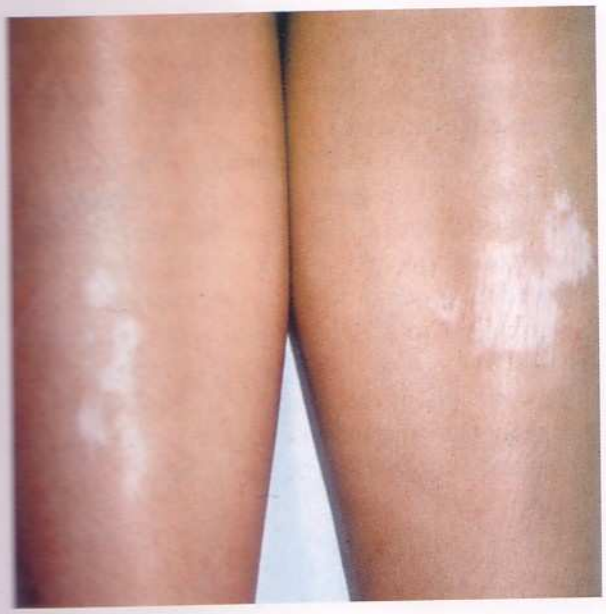

Fig. 11

Fig. 12

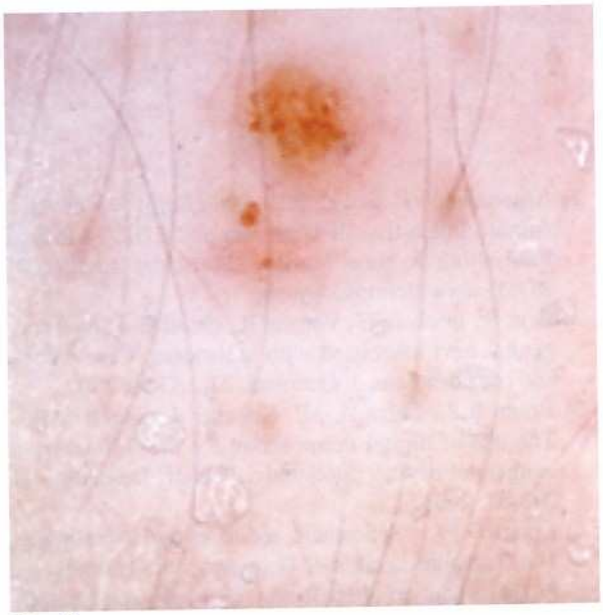

Fig. 13

Fig. 11, 12, 13: In Fig. 11 you can see systemic vitiligo with spontaneous follicular repigmentation. Dermoscopy (Fig. 12) of repigmented vitiligo showed pigmented areas around the hairs. In Fig. 13 you can see the dermoscopic examination of a halo nevus in phase of spontaneous repigmentation. You can see inflammation and vascular structures in the center, where there was the nevus. At the periphery of the regressing nevus, namely inside the white area, you can see hyperpigmented areas, which are centered by hairs.

more or less evident according to the color of the skin, being less evident in the fair complexion. The marked evidence in CMN is probably related to the presence of hair follicles thicker than in normal skin and to the hyperpigmentation of the nevus skin. On the other hand, in vitiligo (Fig. 11, 12), in the halo nevus during the repigmentation phase (Fig. 13) and in the hairy nevus (Fig. 9, 10) the only pigmented areas are around the hair follicles. Probably, this occurs in vitiligo and halo nevus because the melanocytes of the hair follicles are more resistant to the aggression of the autoimmune factors as compared with epidermal melanocytes.

The regular multifocal hypopigmentation of congenital nevi should be differentiated from the regression areas of melanoma. The latter is whiter than normal skin, is not crossed by hairs and is irregularly distributed. MFHypo should also be differentiated from very numerous horny pseudocysts. The latter are smaller, with clear-cut borders and marked refractivity.

The multifocal hypopigmentation is probably related to the presence of more thicker hair follicles. Its peripheral hyperpigmentation is probably related to piling up of melanocytes of the basal layer of the hair follicle and to the surrounding melanocytes.

We are not able to explain why in 13/31 cases multifocal hypopigmentation is not associated with visible hairs. It is possible that the hair hypertrophy becomes evident with years. This hypothesis is supported by the average age of the group without HyperH -4.5 years- as compared with the group with HyperH -10 years-.

In conclusion, the multifocal hypopigmentation is a characteristic of congenital melanocytic nevus and expresses its benign nature.

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