Naevi congenitales pigmentosi gigantei cutis

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SUMMARY

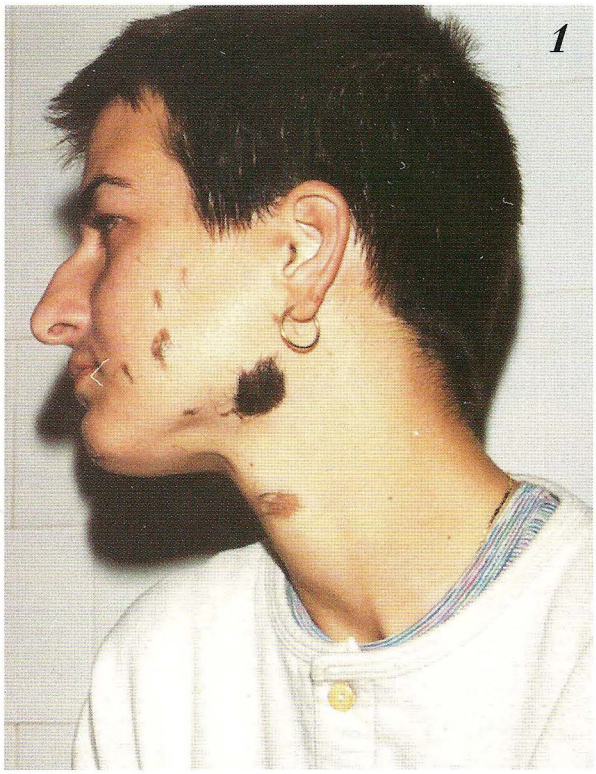
Authors present the case of a 21-year-old female patient with the characteristics of the congenital pigmented hairy nevi.
There were numerous changes in the form of the giant nevi, characterized by the pigmented, hairy appearance. All observed changes were present from birth and had dermatomical distribution on the skin of the trunk, upper and lower extremities and the face. According to the Wallace's rule of nine, total skin surface covered by the changes was about 20%. The patient did not observe changes in the color, size or distribution of the nevi during her life.
Coexisting changes in the brain structures, which may indicate neurocutaneous melanosis, induced us to perform EEG and MRI examinations but the obtained results were negative.
The extensive skin changes and the lack of evidence of malignant alteration of the nevi were the reasons we suggested further clinical surveillance and observation of the patient.

KEY WORDS

pigmented nevus, congenital, giant

Introduction

Congenital giant pigmented melanocytic hairy nevus (CPHN) of the skin (naevus congenitalis pigmentosus et pilosus giganteus) usually is present at birth, but in about 2% of the cases it may appear in the first few weeks of life. CPHN is characterized by the appearance of great pigmented skin changes that are distributed according to dermatomic areas, mainly on hips, abdomen, upper and lower extremities. These pigmented skin changes are unequally colored, being either dark brown or black. Wart-like protrusions could cover the surface of the changes. Edges of the affected skin are irregular, accompanied by smaller satellite nevi or surrounded by hair. Concomitantly with development of a child, skin surfaces covered by CPHN are becoming thicker, darker and covered by wart-like protrusions. Changes found on the head and neck may accompany meningeal melanocytosis (neurocutaneous melanosis). Skin changes are ranging in size from extremely small (less than 1.5 cm in diameter) to very large (more than 20 cm in diameter). Possibility for malignant alteration of CPHN into the malignant melanoma is estimated to be 2-31% (1).



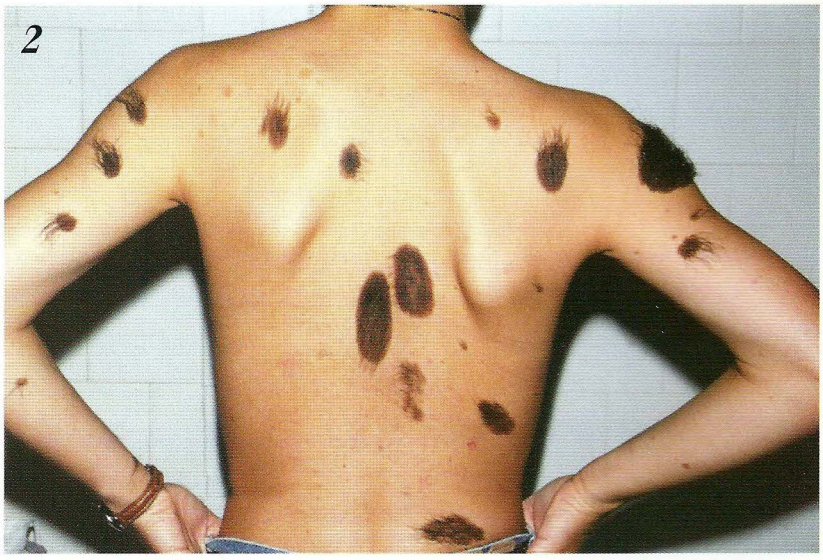
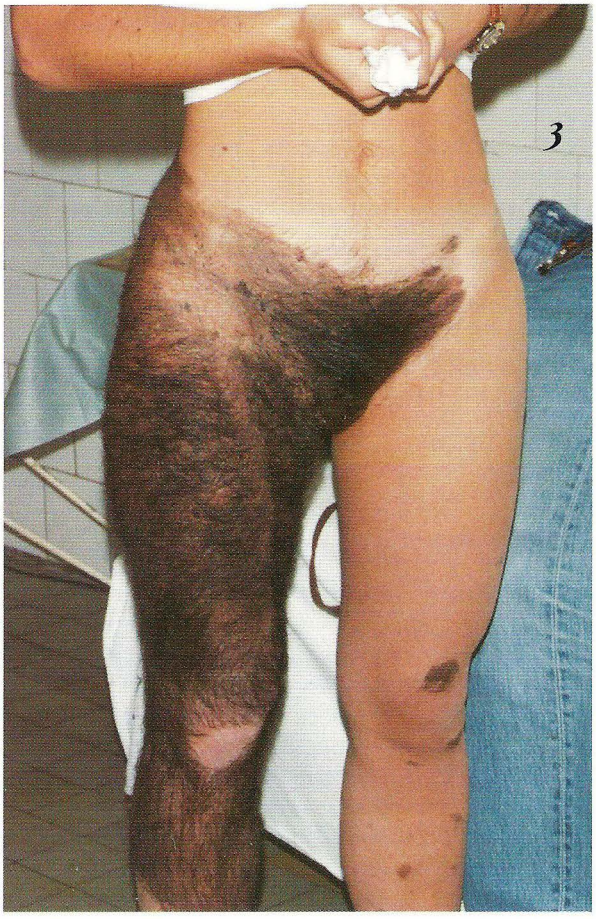


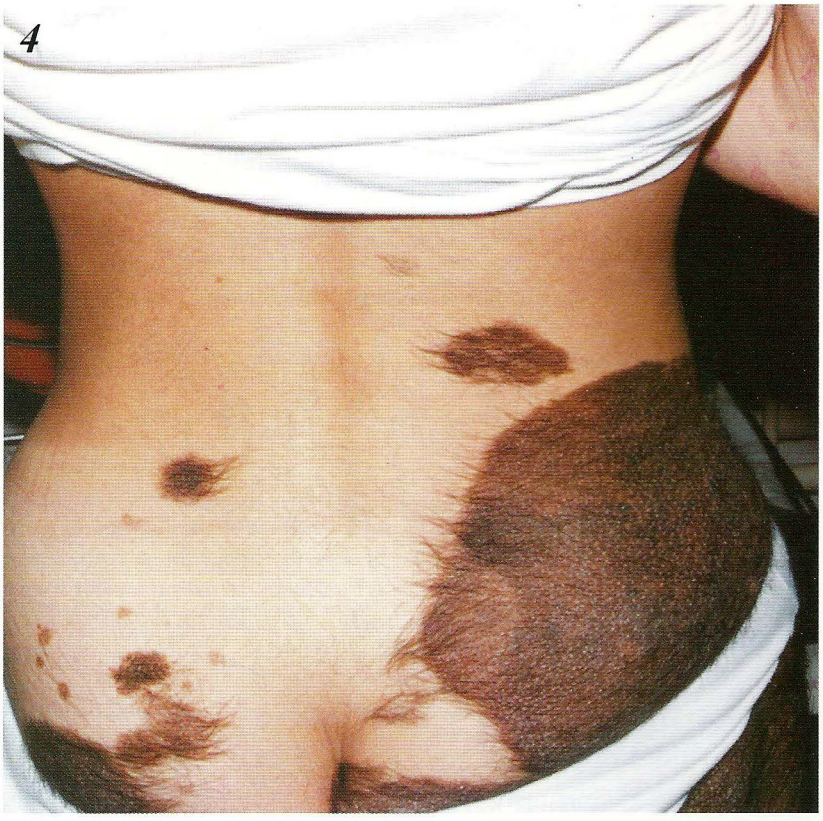
Figure 1. Small and middle sized congenital pigmented hairy nevi on the face and neck.

Figure 2. Numerous small and middle sized congenital pigmented hairy nevi on the back and upper extremities.

Figure 3. Congenital pigmented hairy nevus (CPHN) in the same patient covering completely the right lower extremity and the pubic region.

Figure 4. Congenital pigmented hairy nevus (CPHN) in our patient covering right gluteal area.





Case description

A female patient, 21-year-old student, came to the Clinic for Dermatovenereology in Novi Sad because of the appearance of lichenoid papules. The changes were shiny, smooth, distributed on both wrists. In addition to these findings the patient complained only about pruritus. After clinical examination the diagnosis lichen ruber planus was made. During the examination numerous large and pigmented changes distributed on the face, neck, abdomen, upper and lower extremities were observed. The total surface covered by the 75 lesions amounted according to the Wallace's rule of nine to approximately 20%. The lesions were round. Some had a rough papillomatous surface covered by thick dark-brown hair, 0.3 to 4 cm long. Lesions were sharply delineated, unequally pigmented and scattered. Diameters of single lesions were from 0.5 to 2 cm, while some were of giant size, one covering the entire right leg, extending as far as the gluteal and pubic area.

Laboratory and other findings

Blood cell count: normal; sedimentation rate: normal; MRI: normal findings on leptomeninges and other brain and spinal tissue structures; EEG: normal.

Discussion

Frequency of the appearance of CPHN with diameter larger than 20 cm, in a pediatric patient, is estimated to be 1: 4 150. Female patients are predominantly affected by these skin changes. In the 74% of diseased patients there is concomitant appearance of satellite nevi, and in 31% of the cases changes on the mucosal membranes were observed. Pigmented skin lesions were covered with hair in about 95% of the cases (3). CPHN localized on extremities usually reduces their growth, therefore causing cosmetic, as well as functional disturbances (2). In our patient, besides CPHN localized on the right lower extremity, satellite nevi on the skin were present as well. Pigmented changes on the mucosal surfaces were absent, as well as the signs of the reduction in growth of the affected extremity.

Data about possible malignant alteration of CPHN into the malignant melanoma are different, but they range from 6-12% (4).

Cytogenetical researches performed by Vollenweider et al. (5) proved that CPHN, together with malignant melanoma, belong to the clinical manifestation of "ring chromosome 7 syndrome" (6). These findings obtained by Vollenweider could be a good basis for the explanation of malignant alteration of CPHN. Our patient had no clinical signs of malignant alteration, and due to technical problems we were not able to perform further cytogenetical research.

According to the statements from the literature (2,7, 8,9) about possible appearance of the intracranial melanosis in patients with CPHN, EEG and MRI testing were performed in our patient. These tests excluded the presence of melanoma of leptomeninges, as well as of other brain structures. Results proved that our patient did not show signs of either neurocutaneous melanosis, or of melanoma of leptomeninges.

Literature data also mention simultaneous appearance of CPHN with angiolipoma of the skin (10) and the appearance of CPHN in the Shokeir's syndrome (congenital universal alopecia, epilepsy, mental retardation and pyorrhea) (11). A case characterized by so-called "swimming suit distribution" of the CPHN, that appeared together with subcutaneous retrolumbal ependymoma (two concomitant neuroectodermal tissue defects) was mentioned (12). Our patient did not demonstrate any sign of other congenital neuroectodermal defect or tumor.

Taking into consideration great possibility for malignant alteration of CPHN into malignant melanoma, therapy should be directed toward early prophylactic surgical measures during childhood (13). Surgical procedure in patients with CPHN is usually individual, differing from case to case (14). Lifelong surveillance of the patient in order to detect the early signs of the malignant alteration, enables grafting of the normal skin or excision, as a part of the therapeutic procedure (14). Since our patient does not have any visible clinical signs of malignant alteration, we recommend further clinical surveillance of the skin changes in order to detect possible malignant transformation.

Conclusion

Due to the very high risk for malignant alteration of CPHN into malignant melanoma, we present this case for further education of younger colleagues who may, frequently, be faced in their everyday routine with tumors of melanocytic origin

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