

INCONTINENTIA PIGMENTI ACHROMIANS (ITO)

Lars Hellgren

From the Department of Dermatology, University of Gothenburg, Gothenburg, Sweden

Abstract. A case of incontinentia pigmenti achromians syndrome with associated abnormalities occurring in a 21-year-old woman is reported. Since birth she has had whorling macular, hypopigmented areas on the skin of the lateral, anterior, and posterior aspects of the trunk. Histopathological picture: the melanocytes in the depigmented areas are slightly reduced in number. Some of the associated abnormalities that she exhibits, such as coarse curly hair on her scalp, and pigmented spots in her left iris, have never before been described. Other of her symptoms—thick lips and myopia—have been reported earlier. The patient's mother, 15-year-old sister and 9-year-old brother showed, from birth, brown hyperpigmented maculae on the left halves of their bodies.

Key words: Incontinentia pigmenti achromians (Ito); Hypomelanosis (Ito); Associated abnormalities; Family history

Ito was the first to describe a case of incontinentia pigmenti achromians, in 1952. Since then, more than 20 additional cases of the disease have been reported in the literature. A carefully performed review of these cases and their associated symptoms

is found in (2). Associated symptoms, often severe and frequently multiple, of the hair, eyes, musculoskeletal system, teeth, CNS, blood vessels, sweat glands, and kidneys have been reported (2).

In the present paper, another case of hypomelanosis of Ito is described, and which adds new associated symptoms to the clinical picture.

CASE REPORT

A 21-year-old Swedish woman, who since birth has exhibited whorling, macular, hypopigmented areas on the anterior and lateral left part of the trunk and especially sharp outlines on the abdomen and proximal thoracic region (Fig. 1). Her hair on the left side of the scalp is slow growing, much coarser and more curly than the hair on her right side (Figs. 2, 3). In her left iris, she has a brown pigmented, pin-head sized spot (Fig. 4). Her lips are thick and she is myopic (Fig. 5). Histological examination of the hypopigmented areas showed a slight decrease in the number of melanocytes in epidermis and shortage of melanophores in the upper corium. The normally pigmented areas were entirely normal and showed no features such as those for example seen in

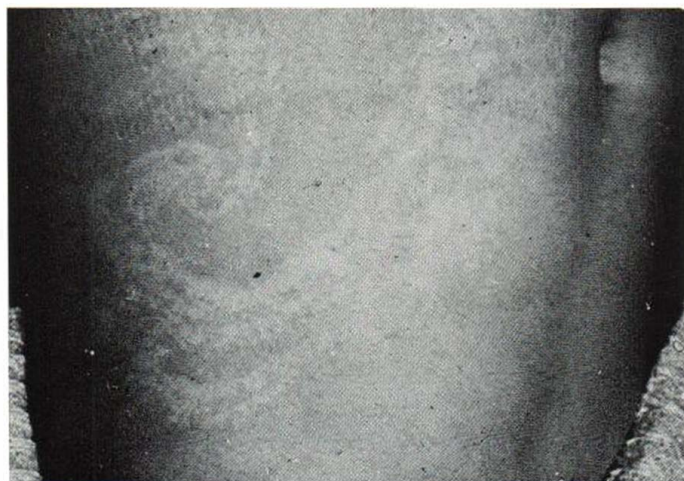


Fig. 1. Whorling hypopigmented pattern on the left side of abdomen.

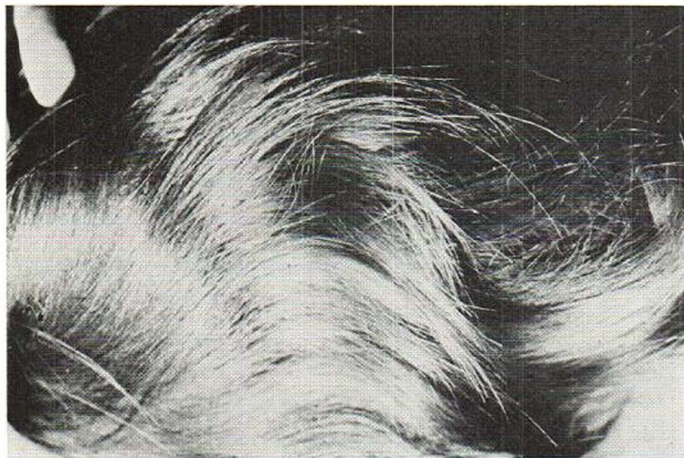


Fig. 2. Coarse and curly hair on the left side of the scalp.

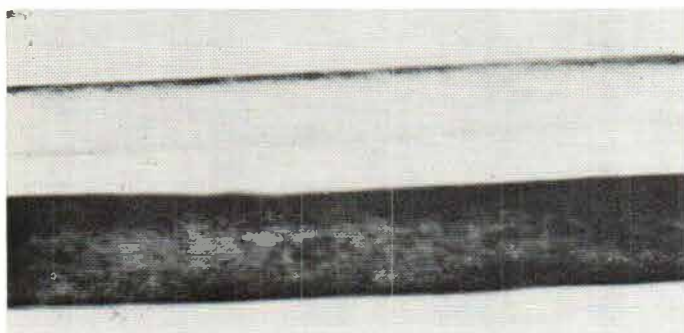


Fig. 3. Terminal hair from the right and the left side of the scalp.

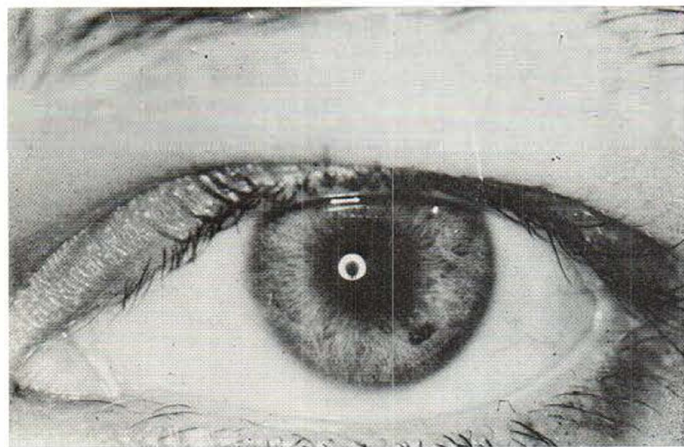


Fig. 4. Pin head sized brown pigmented spot in the left iris.

incontinentia pigmenti. Except for the reported symptom the patient is quite healthy and the routine blood and urine laboratory tests are normal. EEG normal. Her family is healthy but her mother has a 5×5 cm large, hyperpigmented brown macula on the left side of her neck, and one 3×3 cm large on her left side; her 15-year-old sister has a 4×4 cm brown pigmented spot on her left thigh; her 9-year-old brother has a 3×3 cm brown hyperpigmented spot on his left thigh (Fig. 6).

COMMENT

This is a typical case of incontinentia pigmenti achromians beginning early in life and persisting for many years. A number of reports have made additions to the clinical picture. Thus for example the thick lips exhibited by this woman have been described earlier (4) and also the myopia (1), but

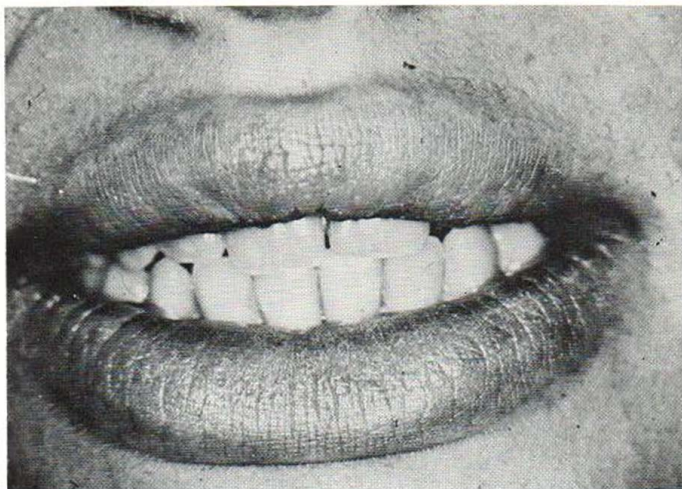


Fig. 5. Macrocheilia.



Fig. 6. Pigmented brown area on the left part of the neck of the patient's mother.

not the pigmented spot in the iris as found in the present patient. Hair symptoms such as diffuse alopecia and facial hypertrichosis have earlier been described (5) but not such coarse and curly hair, unilaterally, as in the present case.

The family history is curious. The patient's mother, sister and brother, all had pigmented spots of the same type on the left side on their bodies; none on the right. This finding is identical with the lesions of the patient, who had all skin symptoms and associated abnormalities (except thick lips and myopia) on the left side of her body only. Familial cases of hypomelanosis Ito have been described (3, 4) and that the disease should be autosomal dominant inherited has been proposed. In the present case there was no familial aggregation of hypomelanosis Ito but the whole family, except the father, had these hyperpigmented spots instead of hypopigmentation, and no other symptoms. Asymmetric hyperpigmented areas of this type are found in incontinentia pigmenti. It has been discussed whether hypomelanosis of Ito should be considered a variant of this disease (2). A mother with incontinentia pigmenti achromians and a daughter with incontinentia pigmenti have been described (1). The coincidental occurrence in a mother and a child of two such rare conditions is exceedingly unlikely. Histological investigations of the pigmented spots of the family members have not been performed.

REFERENCES

1. Hamada, T., Saito, T., Sugai, T. & Morita, Y.: Incontinentia pigmenti achromians (Ito). *Arch Dermatol* 96: 673, 1967.
2. Jelinek, J. E., Bart, R. S. & Schiff, G. M.: Hypomelanosis of Ito ("Incontinentia Pigmenti Achromians"). *Arch Dermatol* 107: 596, 1973.

3. Masumizu, T.: Incontinentia pigmenti achromians (Ito). Jap J Dermatol Urol 8: 214, 1954.
 4. Pinol, A. J., Romagurera, C. & Mascaro, J. M.: Incontinentia pigmenti achromians (Ito). Med Cutanea (Spain) 3: 287, 1968.
 5. Rubin, M. B.: Incontinentia pigmenti achromians. Arch Dermatol 105: 424, 1972.
- Received October 18, 1974*
L. Hellgren, M.D.
Department of Dermatology
Sahlgrenska sjukhuset
S-413 45 Gothenburg
Sweden