

ERYTHEMA DYSCHROMICUM PERSTANS (ASHY DERMATOSIS)

Report of Two Cases from Scandinavia

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Abstract. Two patients with a widespread macular ashy-coloured eruption are described. They fit clinically with the erythema dyschromicum perstans (ashy dermatosis), previously only reported from Central America (about 150 cases) and Britain (1 case). Cases with similar clinical and histological features were found in the literature under various names or presented as cases for diagnosis. The condition seems to be an entity, and it may be more common than is generally realized. Increased knowledge of the characteristic clinical picture and the use of a single name is important.

During the past 2 years we have investigated 2 patients with acquired melanotic patches on the trunk, which clinically and histopathologically closely resemble erythema dyschromicum perstans (ashy dermatosis). This syndrome has only been reported from Central America and neighbouring parts of North and South America (1, 4, 7, 12). Recently, however, a similar patient was reported from Britain (13). This dermatosis is characterized by eruptions of ashy-grey macules, usually on the trunk, resistant to therapy, and with a marked chronicity (1, 4, 7, 12). In active lesions the histological picture is distinct—but not specific—including a hydropic degeneration of the basal cell layer and melanophages in the upper dermis. The etiology is unknown. The following seem to be the first cases from Northern Europe.

CASE REPORTS

Case 1

A 29-year-old Caucasian man. No pigment disturbances in relatives. April-October 1970 he worked on board a ship visiting India and Africa. He had always been healthy until December 1971, when he noticed slightly itching, grey patches on the trunk. He had not observed pre-existent erythema or other eruptions. For 2-3 months the number of patches gradually increased. Since March

1972 there has been no change in number or colour of the patches. He has not been on topical or systemic drugs. Lice infestation was not found.

Physical examination in March 1972 was essentially normal except for the cutaneous changes. Ashy-grey macules were seen symmetrically on the trunk (Fig. 1). They all had a smooth surface, were not infiltrated, and there were no gross signs of skin atrophy (Fig. 2), nor were the margins of the patches erythematous or infiltrated. Rubbing produced no abnormal skin reaction. Mucous membranes, hair, nails and teeth lacked pathological pigmentation.

Laboratory findings: Complete blood cell count normal. ESR 2 mm/hr. Liver function tests, serum creatinine, and immune electrophoresis of serum, normal. Antinuclear factor in serum, absent. Wassermann test (on several occasions), negative. Treponema Pallidum Immobilization test, negative. The urine analysis was normal. X-ray of the lungs: nothing noteworthy.

Histopathology: The specimens were stained with haematoxylin-eosin and according to van Gieson and Masson. Epidermis of normal thickness. Basally in the epidermis there is increased melanin pigmentation and in places liquefactive degeneration of the basal cell layer. In the upper corium there are numerous pigment-laden macrophages and lymphocytic infiltrations around the blood vessels (Fig. 3). No increase in mast cells.

Course. When the patient was re-examined in December 1972 he was still in good health. The pigmented patches were unchanged in number and colour.

Case 2

A Caucasian girl, born in Sweden in 1961. Neither of her parents has any cutaneous pigmentary disturbances. She has always been a healthy child. In August 1971, slightly itching, small red spots appeared on her trunk and the proximal parts of the arms and legs. After 14 days, the red colour disappeared leaving blue-grey pigmented macules which no longer itched. During the next months these spots grew somewhat. The patient has never been on systemic or local remedies. She has not been outside Sweden.

Physical examination in February 1972 was normal except for the cutaneous changes. On the trunk and prox-

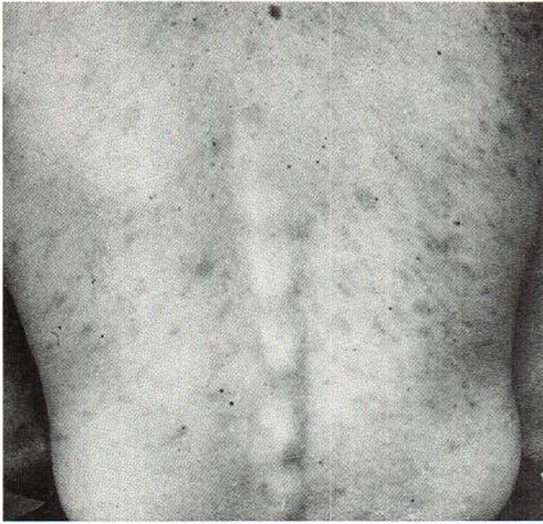


Fig. 1. Distribution of pigmented macules on the back (case 1).

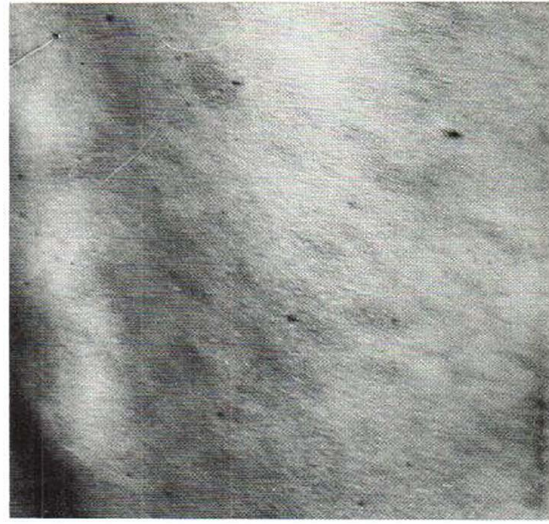


Fig. 2. Close-up of lesions on the back (case 1). Some black naevi pigmentosi also present.

imal parts of the extremities there are about 50 grey macules up to 3–4 cm in diameter. Most of them are elliptical in shape and follow the lines of cleavage. No erythematous border could be seen. No itching. There were no signs of lice infestation. Mucous membranes, hair, nails and teeth were not pathologically pigmented.

Laboratory examination: Complete blood cell count, normal. ESR 10 mm/hr. Liver function tests, serum creatinine and immune electrophoresis of serum, normal. Antinuclear factor in serum, absent. Repeated Wasserman tests negative. Treponema Pallidum Immobilization test, negative. Urine analysis, normal.

Histopathology: The specimens were stained with haematoxylin, and according to van Gieson and Masson. The epidermis shows a slight acanthosis. The basal

cell layer exhibits hydropic degeneration. In the upper parts of the corium there are diffuse oedema, dilated capillaries and marked infiltration of lymphocytes and histiocytes. Around the vessels there are numerous melanophages. There is no increase in tissue mast cells in slides fixed in basic lead-acetate and stained with toluidine blue.

Course. During 1972 we gained the impression that there has been a slight increase of small macules. The colour has not changed.

DISCUSSION

The clinical pictures of the 2 cases have much in common with that of ashy dermatosis as de-

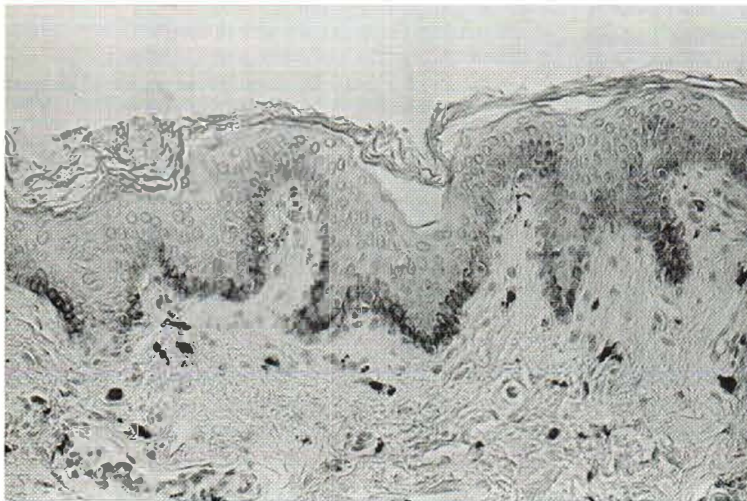


Fig. 3. Biopsy at low power to demonstrate distribution of pigment (case 1).

scribed in the literature (1, 4, 7, 12). The typical colour of well delineated, widespread macules on the trunk, the transient erythematous stage (case 2), the chronicity, the rarity of symptoms and the absence of internal manifestations are characteristic. The histopathological findings of hydropic degeneration of the basal cell layer of the epidermis, perivascular infiltrates of lymphocytes and histiocytes and especially the abundance of melanin-laden macrophages are also in accordance with the reports of Ramirez from Latin America (7). However, biopsies from inactive lesions may merely show pigmentary incontinence into the corium (4, 11).

There are some discrepancies, however. We have the impression that a proportion of the American cases show fewer, larger and less regular cutaneous macules, sometimes also with a reddened border. Some authors even describe an elevated border in progressing lesions (1, 4) which, however, was absent in Ramirez' cases. We believe, however, that the similarities between our two cases and the American cases of ashy dermatosis are so great that they probably represent the same disease. The case of Verbov & Borrie of a 6-year-old girl (13) seems to be identical with ours. These authors are also of the opinion that their patient has ashy dermatosis.

The differential diagnosis of acquired hyperpigmented macular eruptions has recently been discussed in detail by Ramirez (7) and Knox et al. (4). We shall therefore only discuss some points relevant to our cases. Both the distribution of the patches and the often elliptical configuration following the lines of cleavage recalls the pattern seen in pityriasis rosea. Pigmentation is rare following pityriasis rosea, fades quicker and has histological discrepancies. In post-lesional hyperpigmentation of this type there is as a rule only an epidermal melanin increase, because the basal cell layer is intact (5).

Fixed drug eruptions may be followed by circumscribed pigmented plaques which may be difficult to separate histologically from ashy dermatosis, because of the damage to the basal membrane during the initial stage resulting in pigmentary incontinence (8). These eruptions, however, usually have a different configuration and distribution, but it is always necessary to exclude a drug aetiology. Histologically a similar picture can also be seen after lichen ruber planus, lupus

erythematosus and dermatomyositis as well as in incontinentia pigmenti (Bloch-Sulzberger). The clinical signs are quite different, however, and these diseases should not cause any difficulties.

Pinta must be excluded. "Taches bleuâtres" in lice infestations, urticaria pigmentosa, haemochromatosis and hypermelanotic spots in endocrine disorders, i.e. Addison's disease, must also be borne in mind.

The occurrence of two cases in a short time may be a coincidence. We think, however, that this dermatosis is not as rare as might be expected from the few cases in the literature. A survey of the literature discloses several similar patients reported under various headings (2, 3, 9, 10, 14). Grekin demonstrated in 1960 to the Detroit Dermatological Society a 29-year-old white woman as a case for diagnosis (3). She exhibited brownish and blue macules on the trunk and proximal parts of the extremities, and a histopathology we consider consistent with that of ashy dermatosis. In the discussion Mishima was of the opinion that it was a typical case of a dermatosis not uncommon in Japan, *Pigmentatio maculosa multiplex idiopathica*. He had seen three similar cases since he came to the USA. Mishima describes it as "dime-sized to quarter-sized, grey or greyish brown macules that are round or oval, not sharply demarcated and are symmetrically scattered mainly over the clothed areas of the body. It occurs frequently in adolescent and young men" (3).

In Paris, 1964, Degos and Lépine demonstrated a 14-year-old girl with oval, pigmented macules (2). Initially these were slightly erythematous. The persistent spots were located on the trunk and arms. The histology is, in our opinion, consistent with that of ashy dermatosis. Shapiro demonstrated in 1967 in New York a 7-year-old girl with widely distributed small, "tan-to-brown" macules (10). The light-microscopic picture was identical with the cases described above.

In 1971 Rupec & Vakilzadeh described in detail the case of a young German girl with a pigmentary eruption that started with a transient erythematous phase (9). Histologically it was quite consistent with ashy dermatosis and these authors also found the hydropic degeneration so characteristic of the early lesions. The similarity to ashy dermatosis is discussed in the article, but the authors were of the opinion that the clinical pic-

ture was not quite like that of the American cases.

At a conference in Germany in 1972 Weidner demonstrated a 27-year-old man with a slate-grey pigmentary dermatosis quite similar to our 2 cases (14).

To summarize, we think that the clinical and histological pictures of our two cases, the American cases of ashy dermatosis, the above-mentioned cases of spotty melanoderma presented as *casus pro diagnosi* and perhaps also the pigmentation maculosa multiplex idiopathica (3) have so much in common that they may be variants of the same disease entity. A relation between the Japanese dermatosis and ashy dermatosis has also recently been suggested by Pinkus (6). The prime event seems to be an injury to the basal membrane zone. Whether there is a single aetiologic factor is not known at present. In order to accumulate more knowledge of these questions it would be profitable to report these cases under a single name, e.g. erythema dyschromicum perstans (ashy dermatosis).

REFERENCES

1. Convit, J., Kerdal-Vegas, F. & Rodriguez, J.: Erythema dyschromicum perstans. A hitherto undescribed skin disease. *J Invest Derm* 36: 457, 1961.
2. Degos, R. & Lépine, J.: Pigmentation en tache éruptives de nature indéterminée. *Bull Soc Derm Syph* 71: 296, 1964.
3. Grekin, R. H.: Case for diagnosis. *Arch Derm (Chicago)* 83: 327, 1961.
4. Knox, J. M., Dodge, B. G. & Freeman, R. G.: Erythema dyschromicum perstans. *Arch Derm (Chicago)* 97: 262, 1968.
5. Montgomery, H.: *Dermatopathology*, p. 336. Hoeber Medical Division, Harper & Row, New York, 1967.
6. Pinkus, H. & Mehregan, A. H.: *A Guide to Dermatohistopathology*, p. 328. Butterworths, London, 1969.
7. Ramirez, C. O.: The ashy dermatosis (erythema dyschromicum perstans). Epidemiological study and report of 139 cases. *Cutis* 3: 244, 1967.
8. Rook, A., Wilkinson, D. S. & Ebling, F. J. G.: *Textbook of Dermatology*, p. 1032. 2nd ed., Blackwell Scientific Publications, Oxford, 1972.
9. Rupec, M. & Vakilzadeh, F.: Eine kleinfleckige Pigmentdermatose. *Hautarzt* 22: 337, 1971.
10. Shapiro, L.: Melanoderma of unknown etiology. *Arch Derm (Chicago)* 97: 362, 1968.
11. Soter, N. A., Wand, C. & Freeman, R. G.: Ultrastructural pathology of erythema dyschromicum perstans. *J Invest Derm* 52: 155, 1969.
12. Stevenson, J. R. & Miura, M.: Erythema dyschromicum perstans (Ashy dermatosis). *Arch Derm (Chicago)* 94: 196, 1966.
13. Verbov, J. & Borric, P. F.: Case of ashy dermatosis (erythema dyschromicum perstans). *Brit J Derm* 84: 185, 1971.
14. Weidner, F.: Persistierende exanthematische Melanoderma (casus pro diagnosi). *Z Haut Geschlechtskr* 47: 898, 1972.

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