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Segmental Pigmentation Disorder

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Abstract. Pigmentary changes with a dermatomal distribution are described in 30 children. The disorder, up to now unrecognized as an entity, is apparently caused by an embryological determination. It is more obvious in children with darker skin, and seems to fade very slowly over the years. In our experience the incidence is 0.35%. No relation to other anomalies could be established.

Various pigmentary disorders are known to be associated with lesions of the nervous system.

Segmental pigmentation disorder (SPD) is a rather common hypo- or hyperpigmented lesion, having a dermatomal distribution or being sharply delineated along the midline and thus related to a neurogenic distribution.

Occasional case reports of this condition have appeared, but it has not been thoroughly discussed. The purpose of this communication is to report on our experience with this entity.

MATERIAL AND RESULTS

Thirty cases corresponding to the description of SPD were seen in our pediatric dermatology clinic between 1977 and 1981, giving an incidence of 0.35%.

The children were referred because of various dermatological lesions, but only a few of them were related to this specific disorder. In no case was attention drawn by the referring physician to the neurological distribution.

The ages of our patients ranged between 2 months and 17 years. There were 16 males and 14 females.

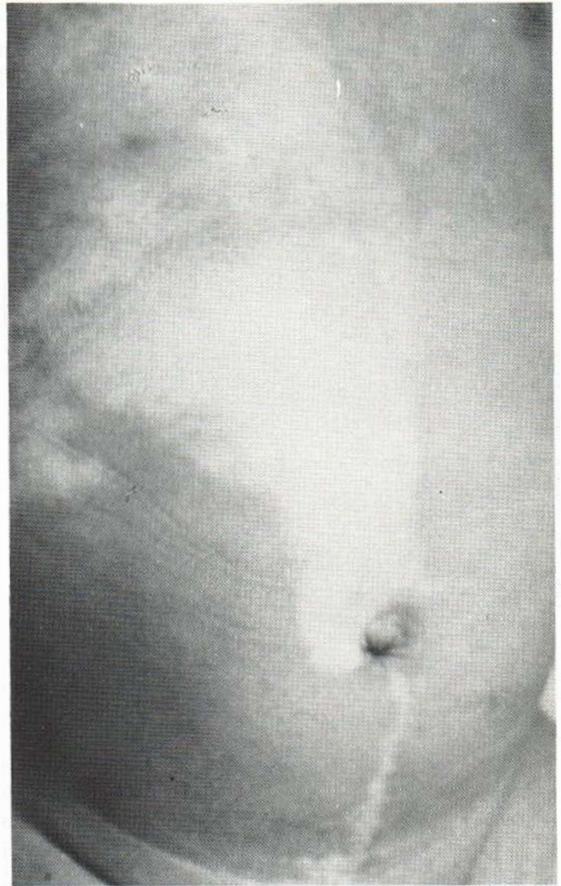


Fig. 1. Depigmentation on abdomen wall with a sharp midline demarcation.

All the children examined were Jewish. The majority (21 out of 30) were of oriental origin (who tend to have a darker skin complexion). The parents of 3 children were of mixed origin, and only 6 children had parents of Ashkenazi origin.

In 23 the lesion was noticed by the parents before the age of 1 year, in 25 before the age of 5 years. In most cases the parents did not seem to be concerned and only 5 children had been brought previously to medical attention because of this pigmentation disorder.

Skin characteristics: 15 cases presented with hyperpigmentation, 15 with hypopigmentation. The distribution varied as follows: over the back (9), the chest (12), and the abdomen (16), with a sharply demarcated border over the midline in the linea alba, but not along its whole length. The lateral border was not sharply delineated. In 7 cases the pigmentation disorder had a dermatomal distribution over an upper extremity (3) or lower extremity (4), in 2 cases over the face. Occasionally it was difficult to determine whether the disorder was hypopigmentation of one side or hyperpigmentation on the neighbouring area.

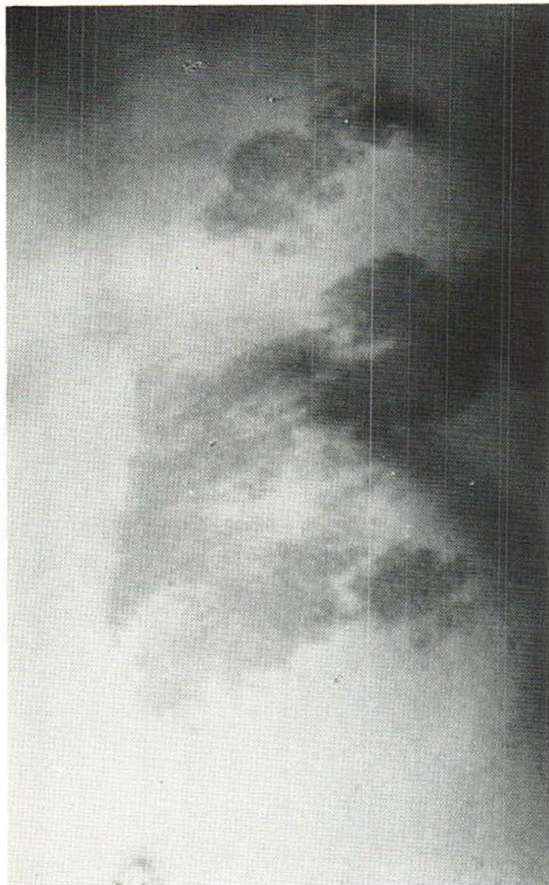


Fig. 2. Hyperpigmentation on left chest wall with a clear midline border.

The lesion was recognized in most cases already in infancy. Only 10 of the subjects were over 5 years of age. In 6 cases some allergy or atopic dermatitis were present, but no association between atopy and SPD could be established.

In 7 cases a gradual decrease in the intensity of the affection was observed during follow-up.

There is insufficient evidence of a genetic or familial factor in SPD, although 2 of our subjects are siblings.

Histology performed in one typical case showed no particular changes in epidermis or dermis, except for increased melanin in the basal layer.

DISCUSSION

The fact that both melanocyte and nerve cell derive from a common origin can explain why certain skin diseases with pigmentary changes are associated with nervous system disorders. Neurofibromatosis and incontinentia pigmenti with hyperpigmented lesions, or tuberous sclerosis and incontinentia pigmenti achromians with hypopigmented lesions, occur with a neurological deficit. Whereas these diseases are genetically determined, in SPD there is an embryological determination that influences pigment migration even years after birth.

The implantation of melanocytes in the skin takes place in early gestation, between the 8th and 14th week of pregnancy, following migration from the neural crest (5, 10).

No definite congenital neurological defects were encountered accompanying the SPD phenomenon. It seems obvious, therefore, that by the time of

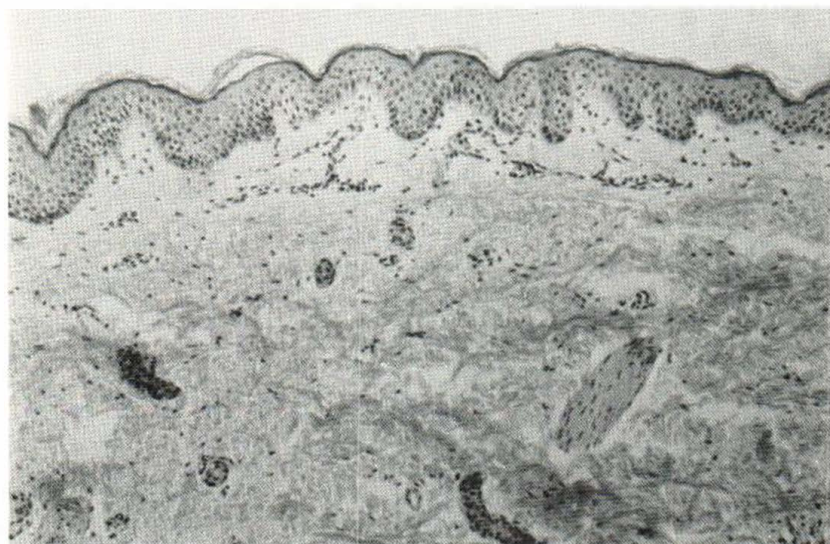


Fig. 3. Histology of a hyperpigmented lesion: normal epidermis and dermis with increased melanin in the basal layer HE stain, 100 \times .

pigment migration, the differentiation of cells in the neural crest is completed and no nerve tissue is involved.

The cause of SPD appearance is so far unknown. Lerner and others (3, 6) suggested the existence of an enzymatic stimulant resulting from a neurogenic provocation, that regulates the pigment production or destruction. Such a stimulant may explain the appearance of vitiligo with a dermatomal distribution or a zosteriform lentiginous nevus (9). If such a neurogenic enzymatic stimulant is really active—the SPD too might be affected by it. An even less plausible explanation is the occurrence of human chimaera (2) as a cause for the relatively common SPD. When presenting in the midline, the pigmentation disorder is characterized by a sharply demarcated borderline. This is due to a medio-lateral pigment migration, namely from the back midline to both sides toward the abdominal midline in the linea alba region, not traversing the linea. A defect in pigment distribution will result in excess or lack of pigment, usually on one side only. Such defects have been described elsewhere (1, 4, 7, 8) as sporadic descriptions not as a defined pathological entity. Such a state of SPD as presented here, has been overlooked, possibly because the child affected was developing nicely and was not bothered by the disorder which appears to fade away within a few years.

We may assume that alertness to this disorder in the future will show that it is quite common; with a better knowledge of SPD we might discover the pattern of its natural course. With a large series of cases, it will be possible to establish, without any doubt, that SPD is not related to any neurological or other pathology.

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Purpura with a Linear Epidermo-dermal Deposition of IgA

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Abstract. A patient suffering from purpura is reported, having persistent linear deposition of IgA along the basement membrane zone in both lesional and healthy skin. The ordinary histopathology showed the picture of purpura pigmentosa with perivascular lymphocytes, polymorphonuclear cells and extravasated erythrocytes. No blisters have been observed.

Non-thrombocytopenic purpura is a heterogeneous entity with or without simultaneous systemic disease. Circulating IgA complexes in blood have been found in Henoch-Schönlein purpura (7), along with IgA deposits in cutaneous blood vessel walls and mesangium (1). In both polymorphonuclear vasculitis and lymphocytic perivascularitis there can be immunoglobulins (IgG, IgM, IgA) in the vessel walls (9). As far as we know, there are no reports of immunoglobulin deposits in the epidermal-dermal junction in purpura. A case report is presented with purpura and persistent linear IgA deposition on the basement membrane.

CASE REPORT

A 60-year-old woman developed large purpuric lesions predominantly on her legs and arms and to a lesser ex-