CICATRISING ALOPECIAS

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An attempt was made to study the distinguishing clinical features, histopathological findings especially the elastic tissue stain and examination of the plucked hairs in 30 cases of cicatricial alopecia. Clinically, pseudopelade had irregular areas of atrophy with tufts of normal hairs arising from the bald scarred areas. Folliculitis decalvans had a history or presence of inflammatory papules and pustules with atrophy extending to the temporal areas. DLE showed features of cutaneous DLE at least in some places. A smooth, slightly violaceous atrophic patch with spinous lesions in other areas suggested lichen planopilaris. Morphoea presented with a sclerosed plaque which was linear in some ases. Most of the plucked hairs in cicatrising alopecias showed anagen hairs which were easily plucked, in contrast to the telogen hairs of alopecia areata. Histopathologically, accumulation of elastic fibres around the vertical fibrous column and loss of elastic tissue in the papillary dermis were diagnostic of pseudopelade. In folliculitis decalvans, fibrosis was irregular with many dilated blood vessels even in the late stage of the disease. Appearances suggestive of solar elastosis were characteristic of advanced lesions of DLE. Irregular atrophy with scanty fragmented elastic fibres were found in the scarring stage of lichen planopilaris. Morphoea showed thickened and closely packed collagen bundles and fragmented elastic fibres. On the basis of its characteristic clinical and histopathological features, not found even in the late stages of other scarring alopecias, it is suggested that pseudopelade is an autonomous disease.

Key words : Alopecias, Histopathology.

Cicatricial alopecia is the end result of a wide variety of pathological processes which cause destruction of the hair follicles and lead to permanent baldness. The destruction may be the result of a developmental defect, physical or chemical injuries, acute or chronic inflammatory changes of infective origin, or due to benign or malignant neoplasia. Most of the fore-mentioned conditions do not cause much diagnostic confusion since they can usually be recognised by the clinical features and appropriate laboratory procedures. Several other diseases, characterised by permanent cicatricial alopecia, such as pseudopelade (PP), folliculitis decalvans (FD), discoid lupus erythematosus (DLE) and lichen planopilaris (LPP), sometimes cause considerable diagnostic difficulty.

The present study is an attempt to define the clinical and histopathologic characteristics of these disorders.

Materials and Methods

Thirty cases of cicatricial alopecia collected over a period of one year, were studied for the incidence, age, sex, morphology, alopecic sites and associated diseases. Biopsies from the bald areas were studied histopathologically using H & E and Verhoeff-van Gieson's elastic tissue staining. Hairs for microscopic examination were plucked from within and around the bald areas by means of a surgical needle-holding forces, the jaws of which had been covered on the grasping surfaces with one to several layers of cellulose tape to ensure a tight closure and to prevent slipping of

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hair shafts during the pulling procedure. Four to five hairs were grasped close to the surface of the scalp and extracted with a quick, forceful pull. The proximal ends of the extracted hairs were cut and examined under distilled water.

Results

Various causes of cicatricial alopecia are shown in Table I.

Table I. Causes of cicatricial alopecia in 30 cases.

Etiological factor	Number of cases		
	Males	Females	Total
Discoid lupus erythematosus	4	7	11
Lichen planopilaris	2	3	5
Pseudopelade	3	2	5
Folliculitis decalvans	2	1	3
Localised scleroderma (Morphoea)	—	2	2
Cicatricial pemphigoid	—	1	1
Epidermolysis bullosa dystrophicus	—	1	1
Nevus sebaceus	—	1	1
Post-furuncle scarring alopecia	1	—	1
	12	18	30

There were 18 (60%) females and 12 (40%) males. Twenty (66.5%) patients were between 11-40 years of age. The youngest patient was 4-year-old and the oldest was 70 years. Father of one patient having FD, also had the same disease. Two sisters had LPP simultaneously, and the sister of another case of nevus sebaceus had a similar problem. The maximum duration of alopecia in this series was 58 years and the minimum one month.

Sunlight aggravated the disease in 7 (23%) cases of DLE. No other aggravating factors were recorded.

Vertex was involved either alone or in combination with other sites in 17 (56.6%) cases, frontal area in 12 (40%) cases and occipital region alone or in combination, in 10 (33%) cases.

Conspicuous follicular plugging at the margin of the bald area was found in 5 DLE cases. Erythema and adherent scales were present in 9 out of 11 DLE patients. Hyperpigmentation was seen in 2 cases and one case showed telangiectasia. Margins of the alopecia patches in FD cases were erythematous, infiltrated and elevated in 2 cases, and follicular papulopustules were seen in one. Slight follicular hyperkeratosis was present in one case of LPP and one case of PP, whereas conspicuous follicular plugging was seen in 3 cases of LPP. Nine (30%) cases showed prominent follicular plugging at the periphery of the bald areas. Atrophy of the alopecic areas was observed in 22 (73%) cases. In 16 patients, the bald area was smooth and shiny. Other changes were mottled hyperpigmentation in 6 cases, hypopigmentation and depression in 5 cases and sclerosis in both cases of scleroderma. Alopecic site was verrucous and papillomatous in case of nevus sebaceus.

The plucked hairs were in the anagen phase in 27 (90%) cases (Table II), of which 17 were

Table II. Microscopic appearance of the plucked hairs from the margin in different types of alopecia.

	Anagen hair		Telogen hair	
	Normal	Abnormal	Normal	Abnormal
Pseudopelade	2	2	1	0
Folliculitis decalvans	1	2	0	0
Lichen planopilaris	4	1	0	0
Discoid lupus erythematosus	5	4	2	0
Morphoea	1	1	0	0
Cicatricial pemphigoid	1	0	0	0
Epidermolysis bullosa dystrophicus	1	0	0	0
Nevus sebaceus	1	0	0	0
Post-furuncle scarring alopecia	1	0	0	0
Total	17	10	3	—

normal and 10 were abnormal (Fig.1). In 19 (63%) cases, only mild force was required to pluck the hairs from the margin of alopecic sites. Out of these, 7 cases were of DLE, 4 cases of PP, 3 cases of LPP, 3 cases of FD and 2 cases of scleroderma.

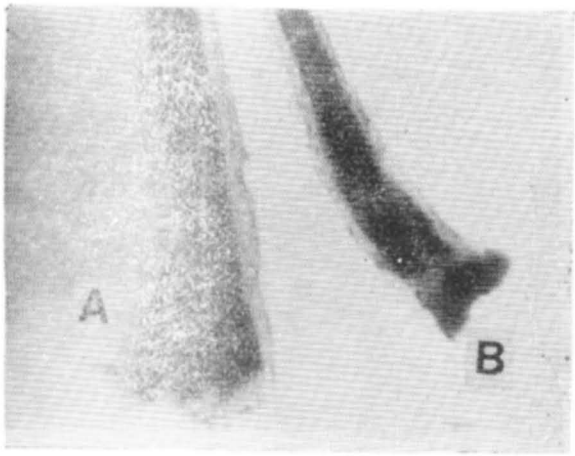

Fig. 1. Plucked hairs showing normal anagen hair (A) and abnormal anagen hair with dystrophic bulb (B).

Histopathology

In most (55%-72%) of the DLE cases, the epidermal changes consisted of hyperkeratosis with funnel-shaped keratotic plugging and hydropic degeneration of basal cells. Atrophy of the stratum malpighii was seen in 6 (54%) cases. The infiltrate in the dermis was patchy, mainly in the vicinity of the atrophic hair follicles and blood vessels. Lymphocyte was the predominant cell, although a few polymorphs were also seen. Fibrosis of the dermis was the prominent feature in all the cases. Dilatation of blood vessels in upper dermis was seen in 7 (63%) cases and of lymph vessels in 5 cases. In three cases, atrophy of the sweat glands and arrectores pilorum was noted. On staining for elastic fibres, the elastic tissue was found to be increased, thickened, curled and tangled in 9 (81%) cases (Fig. 2) and separated from the epidermis by a narrow band of normal collagen. In the remaining two cases, the elastic fibres were destroyed and fragmented.

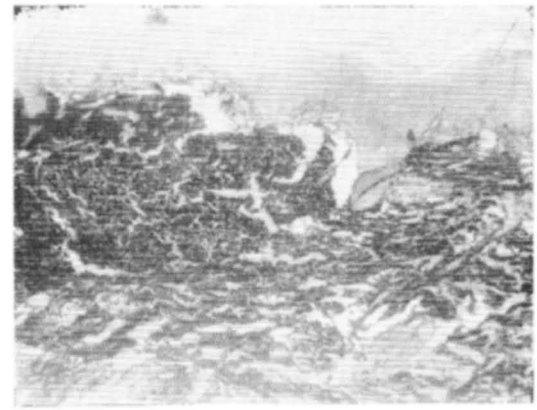

Fig. 2. Hyperplasia of elastic fibers in the upper dermis simulating solar elastosis in DLE (VVG stain x 280).

In LPP, slight atrophy and flattening of the rete pegs were seen in 3 (60%) cases. Keratotic plugging, mainly cup-shaped was present in 3 (60%) cases. Hydropic degeneration of basal cells was found in only one patient. Cellular infiltrates, predominantly lymphocytic were seen around the pilosebaceous structures in 4(80%) cases. There was slight fibrosis of the dermis in 3 cases. Pilosebaceous structures were absent in 2 (40%) and atrophic in 3 (60%) cases. No change was noted in the sweat glands, arrectores pilorum and blood vessels. Elastic fibres were very scanty and fragmented in all the cases (Fig. 3).

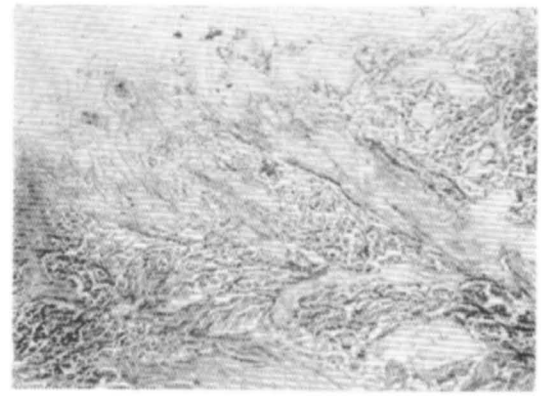

Fig. 3. Scanty and fragmented elastic fibres in the mid dermis and around the fibrous remnants of hair follicles in lichen planopilaris (VVG stain x 280).

Thinning of the epidermis with flattening of rete pegs were the prominent features in PP. The most striking feature was the total disappearance of hair follicles and sebaceous glands and extensive fibrosis of the dermis. Perpendicular columns of fibrous tissue which had replaced the follicles were seen in 4 (80%) cases. No section showed inflammatory cells in the follicular lumen. Elastic tissue stain revealed accumulation of the elastic fibres at the periphery of the fibrous remnants of atrophic follicles and areas of sub-epidermal loss of elastic fibres in 4 (80%) cases (Fig. 4).

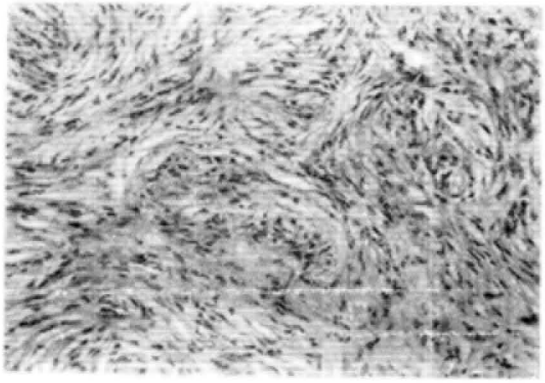

Fig. 4. Perpendicular columns of fibrosis, well outlined by elastic fibres replacing the hair follicles in pseudopelade (VVG stain x 70).

The predominant histopathologic findings in early FD were a marked perifollicular infiltrate, mainly of neutrophils but also containing lymphoid cells and plasma cells. Later, pilosebaceous structures were either absent or atrophic with a mild to moderate fibrosis of the dermis. Blood vessels were dilated in all the cases, even in the late stages (Fig. 5). Epidermis was slightly atrophic, though oedematous and acanthotic in some sections. Elastic fibres were almost absent in all the 3 cases of FD (Table III).

Morphoea cases showed slight atrophy of the epidermis. The dermal collagen appeared closely packed and thickened and stained more eosinophilic than normal. A few fragmented elastic fibres were seen in the mid dermis.

Fig. 5. Epidermal atrophy, prominent dilated blood vessels and complete obliteration of pilosebaceous structures in folliculitis decalvans (H & E x 70).

Table III. Changes in the elastic tissue in different conditions.

Elastic fibres	Number of cases				
	DLE	LPP	PP	FD	Morphoea
Absent/very scanty	—	2	1	3	—
Fragmented	2	3	—	—	1
Increased, thickened, curled and tangled	9	—	—	—	—
Accumulation at periphery of vertical fibrosis	—	—	4	—	—

Comments

It seems that the inflammatory infiltrate in all these conditions does not affect the deep portion of the follicle and the hair itself is not damaged or altered as in alopecia areta, though at times, it may be abnormal anagen hair. The hair seems to persist in anagen until weakening of the follicular sheath causes it to be lost spontaneously or by slight traction. The latter is a valuable diagnostic sign.

Histopathologically, the most prominent feature in 81% of the DLE patients was hyperplasia of the elastic tissue, very much identical to the findings in solar elastosis. Previous workers have not reported such changes in DLE of the scalp. It seems that the DLE lesions on the sun-exposed site like scalp develop basophilic degeneration of collagen, which on elastic tissue staining gives the appearance of elastic material. This further supports the views of many authors[2,3] that sunlight is one of the triggering factors in DLE.

The most striking features in the PP group were the total disappearance of pilosebaceous follicles and the presence of perpendicular columns of elastic fibres. Sub-epidermal loss of elastic fibres was also observed in 80% cases in another series.[4] The term fibrosing alopecia, first described by Pinkus,[4] is applicable to a more diffuse clinical thinning of the hair, and in which elastic fibre formation is not a feature of the cutaneous scar formation. It is distinguished from PP by the minor amount of perifollicular infiltrate and by the development of elastic fibres around the lower portion of the hair follicle.

In FD, 2 biopsies were performed, one from the centre of an atrophic smooth hairless area and a second from an area of the lesion towards the border which contained a pustule. The histopathologic findings in the central portion of the bald area seemed almost identical in all respects to those of PP, but no section showed vertical fibrous remnants of the atrophic hair follicles. In the sections containing the pustules, a marked perifollicular infiltrate was seen with a preponderance of polymorphs along with a considerable number of lymphocytes. Intrafollicular infiltrate was not seen in any section. The perifollicular infiltrate contained a good number of plasma cells, which were absent in other groups of scarring alopecias studied. Blood vessels were dilated in all the cases which was very striking.

In the present series of LPP, none of the sections showed sub-epidermal infiltrate typical of lichen planus, while hydropic degeneration of the basal cells was practically absent. These could be explained by the fact that most of our cases were in the late stages, hence basal cells were probably replaced by squamous cells.

In general, the sebaceous glands were lost, whereas sweat glands and arrectores pilorum were present in almost all the sections of DLE, PP, LPP and FD irrespective of their stage. This suggests that probably sebaceous glands are affected first in cicatricial alopecia, and thus their absence should make the dermatologist and pathologist suspicious of the diagnosis. Morphoea looked the same histopathologically as elsewhere in the body.

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