ADENOMA SEBACEUM—A HISTOPATHOLOGICAL REAPPRAISAL

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Summary

A histopathological reappraisal of skin biopsies from 5 cases of adenoma sebaceum showed that there is no pathognomonic microscopic appearance of adenoma sebaceum. Histopathological appearance in one case was that of pigmented naevus.

Adenoma Sebaceum (Synonyms-Tuberous sclerosis, Epiloia, Bourneville's disease, Brushfield-Wyatt's disease) is a rare syndrome characterised by the presence of cutaneous facial angiofibromata, mental deficiency and epilepsy. The term adenoma sebaceum is a misnomer because the papules are not formed due to hyperplasia of the sebaceous glands only but there is in addition consistent hyperplasia of the hair follicles and fibrous tissue without any change in the elastic tissue. Since the microscopic changes are essentially a combination of the vascular and connective tissue hyperplasia, it would, therefore, be more appropriate to name the condition as angiofibroma. The angiofibromas consisting of many small reddish papules are distributed symmetrically in the nasolabial folds, on the cheeks and chin. Large, raised brownish fibrous, asymmetrically arranged nodules on the face and scalp, may be seen along with periungual fibromas and shagreen patches.

In the past attempts were made by various authors to classify the cutaneous lesions of adenoma sebaceum into 3 types:

(1) White variety-Balzer type[3]; (2) Red soft variety-Pringle type[4]; (3) hard variety, with predominance of fibrous tissue-Hallopeau Leredde type[5].

The microscopic description as given in Pringle's original article is thinning of the epidermis which dips deeply into the dermis giving rise to an appearance of great papillation. This appearance could be due to pushing upward of the epidermis by proliferated collagen. This proliferation of collagen also leads to downward and lateral displacement of the ducts of the sebaceous glands and hair follicles. Instead of laying stress on angio-fibromatosis, Pringle[4] emphasized sebaceous gland changes. The accepted microscopic description of adenoma sebaceum during the past 82 years would have been different, had Pringle emphasized angiofbiromatous change.

Nickel and Reed[6], however, on the basis of 74 skin biopsies studied by them have shown that the microscopic picture of adenoma sebaceum comprises of fibrosis and capillary dilatation in the dermis the sebaceous glands being generally

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atrophic. In some lesions, the large and stellate shaped fibroblasts simulate glial cells. In older lesions there may be perifollicular proliferation of collagen bundles resulting in atrophic hair follicles. Elastic tissue is absent in angiofibromas. Shagreen patches, however, showed increased amount of collagen material and relative increase of elastic fibres.

Butterworth[7] has stated that the essential change in adenoma sebaceum is hyperplasia of hair follicles along with increase in collagen. If histopathologically a lesion showed above changes it was designated as Hallopeau-Leredde[5] variety. In addition to hyperplasia of the hair follicles and of the collagenous tissue if that of the sebaceous glands is also present, the lesion is designated as Balzer's type[3].

Increased vascularity in addition to above described features, makes a lesion the red, telangiectatic, Pringle type[4]. The increase in number of hair follicles in an adenoma sebaceum has been explained by Nickel on the basis of the sectioning of tissue parallel to the surface rather than to any actual pathological changes. Sehgal and Sehgal[8] reported a case of adenoma sebaceum in whom the presence of increased number of immature and mature hair follicles has been described.

We obtained skin biopsies from 5 cases of adenoma sebaceum. Their histopathological appearances are given below :—

A 14 year male: (Fig. 1 page No. 146) The histopathological examination of the skin revealed normal epidermis and multiple atrophic hair follicles showing concentricaley arranged collagen around them. The sebaceous glands were prominent and no dilated capillaries could be seen. Elastic fibres were not prominent. Foci of chronic inflammatory reaction were seen in the superficial dermis.

A 30 year female : (Fig. 2 page No. 146). The skin biopsy showed normal epidermis with totally absent sebaceous component. The hair follicles were normal and the capillaries not prominent. Some collagen bundles showed slight hyalinisation. Elastic fibres were not prominent. Groups of melanin containing melanoblasts were seen in the superficial dermis. The histopathological diagnosis was 'pigmented nevus.'

A 12 year male : (Fig. 3 page No. 146) Microscopic examination of the section showed normal epidermis and multiple atrophic hair follicles surrounded by collagen fibres. Sebaceous glands were very few and atrophic. Capillaries were neither dilated nor prominent. Some of the collagen bundles were thickened and hyalinised. Elastic fibres were not conspicuous.

A 30 year female : Histopathological examination of the section revealed hyperplasia of the covering stratified squamous epithelium with foci of chronic inflammatory cells, in the superficial dermis. Numerous hair follicles were a prominent feature, with mononuclear infiltrate around them. At some places, they were atrophic and not surrounded by collagen fibres. Sebaceous glands were hypertrophic and very prominent. No abnormality was seen in the capillaries. Collagen bundles were dense and hyalinised in the deeper dermis. Elastic fibres were not prominent.

A 25 year female : Microscopic examination of the skin biopsy showed normal epidermis and multiple pilosebaceous units. The hair follicles were not prominent, and capillaries did not show any change. The sebaceous glands were hypertrophied. The collagen bundles

were thickened and hyalinised. Elastic fibres were not prominent.

Comments

Our histopathological findings show that there is no pathognomonic microscopic appearance in adenoma sebaceum. The sections from one of the cases showed prominent sebaceous glands, whereas in others they were less conspicuous and rather atrophic. The hair follicles were compressed by hyalinised collagen fibres.

Our findings are in agreement with those of Nickel and Reed[6] who have stated that there is no typical feature of adenoma sebaceous from the histological point of view, and each case must be judged on its own merit keeping in mind the variations.

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False

Aquagenic urticaria is a distinct and rare form of urticaria which occurs on contact with water. It presents as an itching eruption of small papules very similar to those of cholinergic urticaria. Shelly and Rawnsley in 1964 described three patients who had aquagenic urticaria. This form of urticaria has been reported to be produced after water ski-ing, sweating after exercise, hot baths, contact with distilled water, tap water, etc.

Before this rare type of aquagenic urticaria can be diagnosed, the usual tests for the other patterns of physical urticaria must be carried out. It is probably correct to reserve the term aquagenic urticaria for the very rare cases which show the typical wheals on the application of a compress of water applied for 30-40 minutes preferably kept at 36°C throughout and in which the tests for other patterns of physical urticaria are negative. It has been suggested that the water acts with a component of sebum to produce a histamine liberator which is absorbed with subsequent discharge of histamine from the perifollicular mast cells.

Reference: Warin RP and Champion RH: Urticaria, W. B. Saunders Company Ltd, London, 1974, p. 143.