CHONDRODERMATITIS HELICIS

By J. J. BIBER (New York).

In recent journals of dermatology, appeared several publications about a new pathological entity called chondrodermatitis nodularis chronica helicis, or, in short, chondrodermatitis helicis which—it is thought—owing to its localization, deserves our attention.

In 1915, Winkler described eight cases of a lesion with constant localization at the upper margin of the helix. Basing on his histopathological sections, showing changes not only involving the overlying skin but also the adjacent perichondrium and cartilage, he suggested the aforesaid name for this lesion.

The common feature of all described cases was formation of a small nodular lesion at the upper margin of helix, in shape of a round or ovoid growth, white or greyish in colour, showing sometimes livid discoloration, especially when due to secondary inflammatory processes the surrounding vessels were congested. The lesion was mostly tender and the epidermis peeled off from the surface in form of scales or was covered with a crust. After a removal of the scales or the crust the centre showed an impression or formed an ulcer.

Foerster reported independently in 1918, four similar cases, calling it "*painful nodular growth of the ear*". Several years later (1925) eight other cases were reported by the same author. Just as Winkler, he found this lesion in male adults only.

Rost (1926) was the first to describe among seven observed cases, also three in females. One of the females was a nun who acquired a lesion at the anthelix and this caused Rost's suggestion that it might have been caused by chronic irritation of a starched hood.

Carol and Van Haren (1941) suggested the name "clavus helicis" as they assumed that their histological findings justified it.

Several similar cases were described by other authors (Culver, Fox, Klauder, Roxburgh, *et al.*) and discussed its pathogenesis and possible relation to malignant degeneration.

Recently there appeared an exhaustive work by Shuman and Helwig dealing with this subject from clinical and pathological view point. They reviewed 150 cases and analysed 100 of them regarding its clinical feature, possible errors in clinical diagnosis, relation to age, race, sex, and localization. They found the prevailing incidence of this lesion in white race (98 cases). Sixty cases were confined to the right ear, 33 to the left,

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and seven showed bilateral lesions. In 85 per cent. the lesions were found on the upper margin of helix, six at anthelix, two at the tragus, one case at antitragus, and in two cases the lesion was found in the concha.

Clinical Picture

A small, well-confined nodule, round or ovoid in shape, at the margin of helix (vide fig. 5). It is tender, white-greyish in colour, but may appear reddish or livid, especially when due to concomitant secondary infection the surrounding skin shows inflammatory reaction. The epidermis peels off at the top of the lesion in form of scales or is covered with a crust. After its removal, a depression or an ulcer forms.

Histological diagnosis is made according to Shuman and Helwig by finding:

(1) Nodular hyperplasia of epidermis, centrally depressed or ulcerated and topped by hyperkeratotic and parakeratotic scales.
(2) Œdema, homogenization and fibrinoid necrosis of dermal connective tissue, with formation of cystlike spaces and clefts, often communicating with the ulcerated surface.
(3) Proliferation of highly vascularized granulation tissue surrounding or partly replacing the degenerated dermis.
(4) Presence of chronic inflammatory infiltrate with different amount of lymphocytes and plasma cells.
(5) Evidence of perichondritis or degenerative changes of the underlying cartilage, which may be more or less pronounced. In that case, there is found swelling of interstitial ground substance and chondrocytes. The chondrocytes often are found sparse or disappear, leaving cystlike spaces.

Ætiological and Differential Diagnostical Considerations

The ætiological factor is a much disputed and often controversial subject as several authors express different opinions. Winkler and others assume that embryological variations in development of the male cartilage of the ear might constitute a locus minoris resistantiae, which might favour pathological conditions by exposure to physical or chemical irritations. Others, like Halter, underline the importance of nutritional factors of the marginal part of the helix. Lack of larger vessels and sufficient anastomoses together with scarcity of underlying connective tissue should favour development of pathological processes.

Ebenius and his followers suggested the predilection of the right ear in development of the lesion might be due to the fact that most of the patients slept on that side. (This seems to be hardly acceptable as in this case the anthelix as the more protruding part of the auricle would be more exposed to pressure.)

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It would be too much to discuss the controversial theories in this paper but it is most probable that not one, but several factors have to be blamed for formation of this lesion (thermal, chemical, and other physical factors).

These factors combined with malnutrition of the marginal part of helix might determine the localization. The clinical diagnosis might be difficult as it has to be differentiated from other similar lesions which have to be taken under consideration like senile keratosis, cornu cutaneum, clavus, squamous papilloma, verruca cutis, inflammatory processes, squamous carcinoma, and some dermatoses. The relation of chondrodermatitis helices to malignant degeneration has been much discussed by several authors. Ebenius examined 26 cases histologically and although unable to prove its malignancy supposed to have found pre-cancerous changes.

The clinical errors in differentiating this lesion from other pathological conditions might be only avoided by histological examination of the excised lesion and even in these cases the histopathological findings could be disputed.

In Shuman and Helwig cases, no clinical diagnosis was made in 42 cases and only 15 of the remainder 58 cases were correctly clinically diagnosed as chondrodermatitis helicis.

A correct pathological diagnosis was made in about 41 per cent. of their cases, 58 lesions were incorrectly diagnosed, 9 per cent. of the cases were diagnosed as basal or squamous cell carcinoma, and in 29 per cent. the diagnosis implied that the condition was either cancerous or pre-cancerous.

Treatment

The variety of possible errors in clinical diagnosis suggests that a radical surgical excision of the lesion is the most successful way of treatment.

Three case reports of my own observation and by courtesy of Dr. B. N. Rosenberg, Gallipolis, Ohio:

Case I. Mr. E.W., age 64.

Complaining of a tender growth at the upper margin of the auricle, which persisted for several months and caused pains during the night while sleeping on that side.

O.E. On the upper part of the right helix a small ovoid lesion, greyish-white in colour, the surface elevated, horny and keratotic. The growth is tender on pressure. June 4th, 1954, excision of the lesion under local anæsthesia, with primary suture.

The histological report by Dr. R. P. Carson, reads:

"There is an area of acanthosis and hyperkeratosis (Fig. 1) and on the central aspect of specimen the epithelium is ulcerated into the dermis. The

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dermis shows marked focal hyalin and fibrinoid changes in the collagen as well as formation of small cyst. In more peripheral areas there is fibrosis, increased vascularization and scatterings of lymphocytes. Although auricular cartilage is not included with the material, the above described features are consistent with those which are commonly seen in perichrondritis or chondrodermatitis helicis (Fig. 4).

Diagnosis: keratosis, consistent with perichondritis or chondrodermatitis helicis."

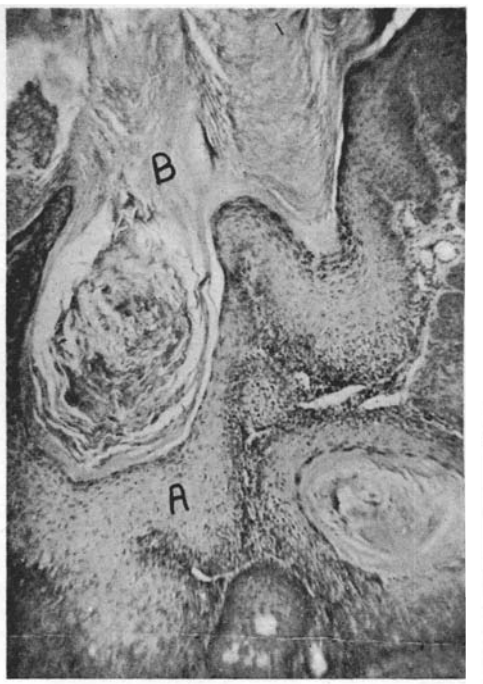


FIG. 1.
(*a*) The epidermis is the site of nodular and circumscribed acanthosis.
(*b*) The stratum corneum is thickened by laminated and heaped up hyperkeratosis.
(Case E.W.)

CASE II. Mr. C.S., age 58.

The patient visited the hospital on December 12th, 1953, to see about a small roughened area on the right ear on the upper part of helix. It has been first regarded to be senile keratosis and he has been advised to keep it softened with vaseline and also advised if it broke open to return to the hospital.

He returned on April 6th, 1954, complaining about tenderness, especially as he slept on the right side. In local anæsthesia, the lesion has been excised with the adjacent cartilage and the wound edges sutured after approximation.

Histological report by Dr. R. P. Carson reads:

"There is a small piece of auricular cartilage in lowermost aspect of specimen. This shows fragmentation and degeneration at its borders. Surrounding it and

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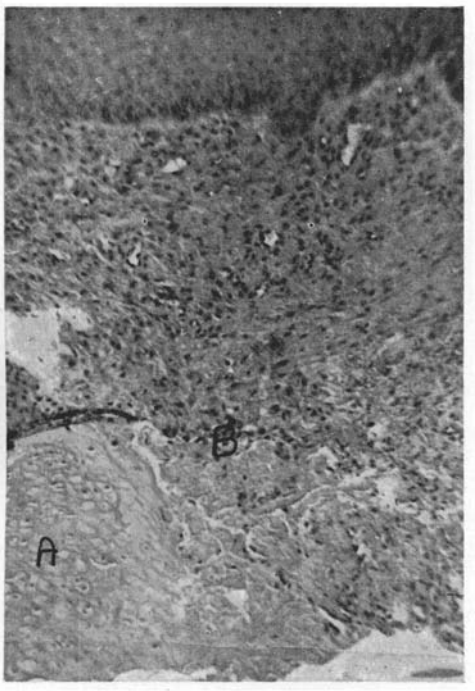


FIG. 2.
(*a*) The auricular cartilage is degenerated, with swelling of ground substance and loss of orientation at its edges.
(*b*) In the surrounding connective tissue there is degeneration and fraying of collagen fibres with the formation of cyst-like spaces. Increased vascularity and chronic inflammation is present (Case C.S.)

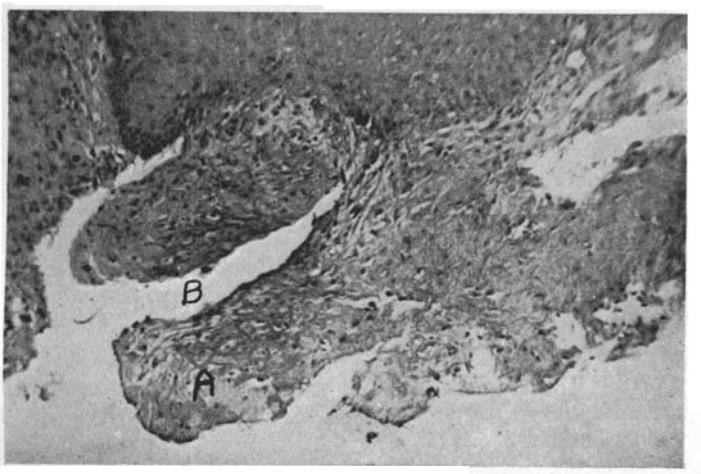


FIG. 3.
(*a*) Degenerated auricular cartilage blends into dermal collagen. In the latter, the fibres show disruption, fusing and homogenization.
(*b*) A cyst-like space layered by fibrinoid material communicates with the cartilage (Case C.S.)

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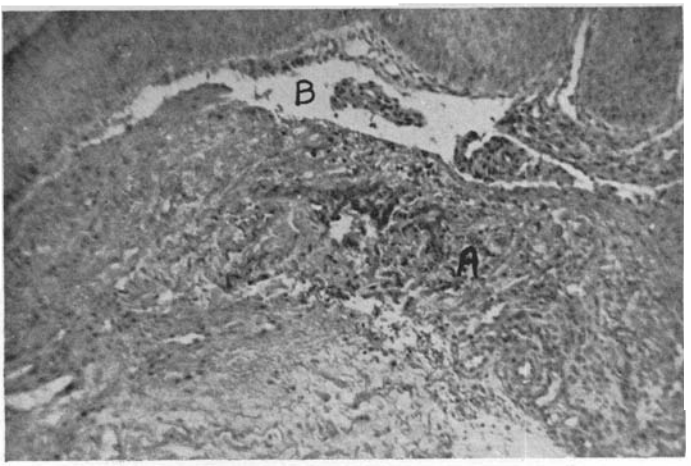


FIG. 4.

(*a*) The dermal collagen shows fragmentation of collagen and elastic fibres, fibrinoid necrosis, vascularization and chronic inflammation.
(*b*) Cyst-like spaces are evident and have extended to the ulcerated surface in another area. (Case E.W.)

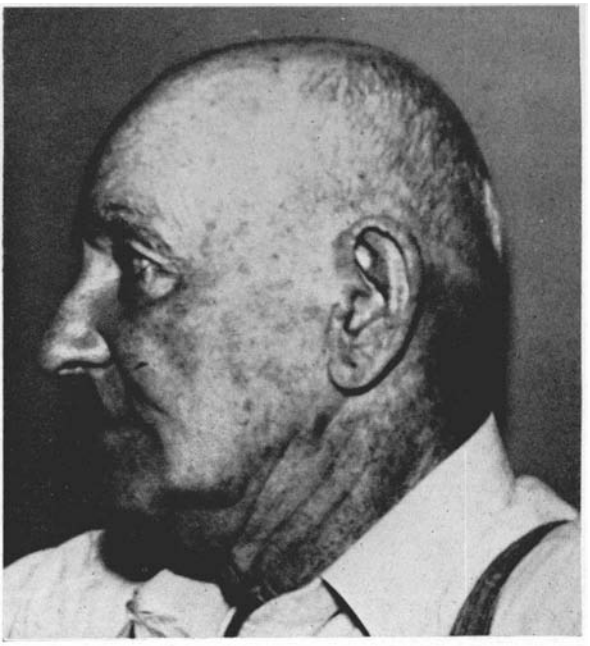


FIG. 5.

(Case 3) shows a small horny nodule of hyperkeratosis at the outer margin of helix typical of chondrodermatitis helicis.

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between the cartilage and the epidermis, there is vascularization of the connective tissue, active fibrosis and small areas of hyalin and fibrinoid degeneration of collagen. (Figs. 2 and 3.) The epidermis shows marked localized acanthosis and it is capped by a thick layer of hyperkeratosis and parakeratosis.

Diagnosis: chondrodermatitis helicis."

CASE III. Mr. T.W. McC., age 81.

Complaining of tenderness of both ears which prevent him from sleeping as movements to each side are painful.

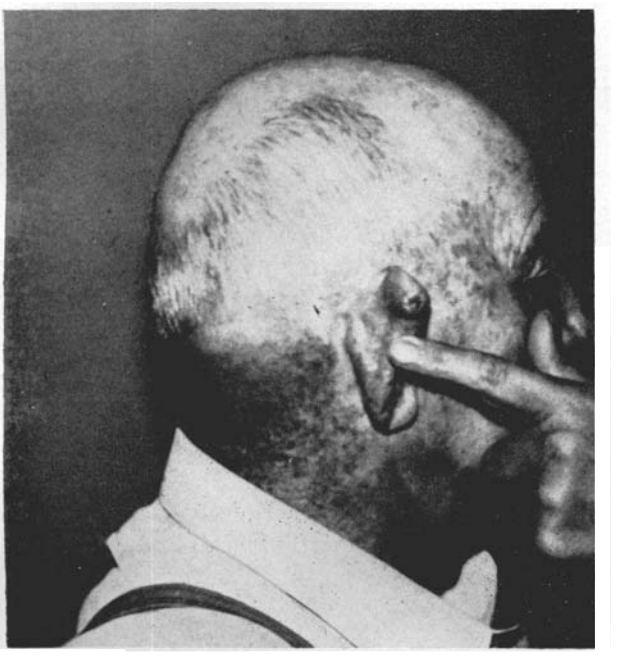


FIG. 6.

The other ear of the same case shows at the identical place a large, hard and ovoid nodule with central umbilicated ulceration suggesting neoplastic infiltration.

O.E. At its outer margin, the left auricle shows a white-greyish nodule, covered on the surface with hyperkeratotic horny scales, which are tender upon application of pressure. At the identical place the right auricle shows a lesion surrounded by infiltrated and elevated margins, centrally exulcerated, ovoid in shape, of a size of a sixpence. It progresses along the posterior surface of the helix. (Figs. 5 and 6.)

Clinical diagnosis: chondrodermatitis nodularis helicis of the left auricle and malignant degeneration of similar lesion on the right auricle. Radical excision of both lesions was performed through the whole thickness of the ear with plastic repair on the right side, on August 4th, 1954.

Histological report (Dr. Carson) reads:

"From the left ear: irregular and rather extensive hyperkeratosis, as well

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as an irregular acanthosis of the surface epithelium. In some places, this epithelium tends to be atrophic, shows plexiform extensions of its rete pegs. The underlying dermis shows fragmentation of the collagen fibres, increased vascularity and patchy infiltrations by lymphocytes and plasma cells. In one of the specimens, the auricular cartilages is included. It shows degeneration and fragmentation at its borders, as well as fragmentation and degeneration of surrounding collagen.

Diagnosis: chondrodermatitis helicis."

The specimen from the right ear reveals a rather extensive, but well differentiated squamous cell carcinoma. The surface is focally ulcerated. The auricular cartilage is not involved. Diagnosis: squamous cell carcinoma, grade II.

This case is also interesting in this respect that it might support the opinion of some authors considering the chondrodermatitis nodularis helicis as a precancerous stage.

Summary

In brief, the past publications about chondrodermatitis nodularis helicis have been reviewed. This has been recognized as a pathological entity. The subject has been discussed regarding its clinical and pathological feature, differential diagnosis with some similar lesions and possible errors. The necessity of securing the diagnosis by histological section has been emphasized in order to detect possibility of malignant degeneration.

Report of three cases has been given with histological pictures showing the main features of this pathological entity.

I wish to express my thanks to Dr. R. P. Carson, pathologist, Holzer Hospital, Gallipolis, Ohio, for the help I received from him in obtaining of the pictures and interpretation of pathological findings. I am also indebted to him for the references to the recent literature.

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