

## CUTIS VERTICIS GYRATA \*

### REPORT OF CASES

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Cutis verticis gyrata has been well described by various authors since the first report by Jadassohn at the Ninth German Dermatological Congress in Berne. A recent interesting report and review of the literature by Wise and Levin<sup>1</sup> describes the first example of the anomaly observed in America. As far as I know, the cases that I am presenting constitute the second and third reported in this country.

The condition is characterized by the appearance of deep furrows and convolutions in the upper posterior portions of the scalp more or less resembling cerebral convolutions. By many it is regarded as a congenital tendency to furrowing which becomes pronounced later in life. My first patient comes in this category. Some believe that the process is usually intensified by local inflammation. My second patient comes under this classification.

Through the courtesy of Professor Lesser, I had the privilege of seeing a case in Berlin in 1912, so the scalp that I am presenting here constitutes the second well developed example of the condition that I have seen.

My first patient's scalp, preserved in Kaiserling solution No. 4, is here presented for your inspection. My second patient, presenting the deformity in its early stages, refused to part with his scalp. A photograph will be published later.

As will be noted on studying the gross specimen and photograph, the appearances suggest neurofibroma. We thought (and hoped) that it was that condition but microscopic study failed to disclose the presence of nerve tissue, and there were no lesions elsewhere.

### REPORT OF CASES

*History.*—An apparently healthy Jewish girl, 15 years old, had had an oval well circumscribed sessile tumor with numerous convolutions covering most of the back of the head since birth. It had slowly grown larger, with deepening of the sulci. The growth had progressed more rapidly during the last few years. It had never been painful.

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\* Read at the Forty-Fifth Annual Meeting of the American Dermatological Association, Washington, D. C., May 2-4, 1922.

1. Cutis Verticis Gyrata—Report of a Case. Wise, Fred, and Levin, Oscar L.: Interstate M. J. **25**: No. 5, 1918.

She had always been in good health with the exception of occasional headache and nausea in the morning. She also had had measles, pertussis and parotiditis in early childhood, with no sequelae. There had been no accidents, operations or local infections, and the scalp had never been inflamed. For several years she had had occasional general headache and nausea in the morning.

Her eyes, ears, nose and throat had always been normal, and there never had been marked glandular swelling. She did not have goiter. There was no evidence of past or present disease of the respiratory systems. Her mentality was quite good.

*Examination.*—The skull was dolichocephalic in form. In the occiput and extending almost to the top of the head was an oval, sessile tumor with a

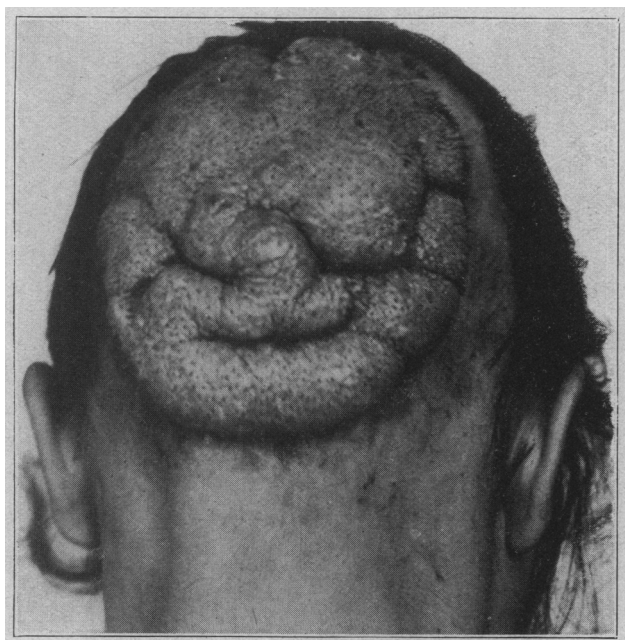


Fig. 1.—Cutis verticis gyrata in a girl, 14 years old, with a dolichocephalic type of skull. The hair was shaved for operation.

sharply defined, slightly constricted base. It was about 15 cm. in its broadest diameter and was elevated about 1.5 cm. at its highest point. It presented three main horizontal furrows and five secondary ones with convolutions like those of the cerebrum. These furrows were narrow and deep. The skin was loose and soft, and through it many firm round nodules 0.5 to 1 cm. in diameter, resembling sebaceous cysts, were plainly felt. When the scalp was shaved for the operation, the skin was seen to be normal in appearance. The hair was long, thick and black. It grew evenly over the entire tumor mass. The rest of the scalp was normal.

*Eyes:* The pupils were equal, regular and reacted to light and accommodation. The conjunctivae, eyelids and lashes were normal.

*Nose:* There was no obstruction or discharge.

Ears: The hearing was good. There was no discharge.

Mouth: The teeth were good and well kept. The tongue protruded normally. The tonsils were fairly large but apparently not diseased.

Neck: There were a few small palpable glands in the posterior cervical triangles. There was no goiter. There were no abnormal pulsations.

Chest: The cardiovascular and respiratory systems were normal.

Abdomen: The abdomen was flat. There were no masses or areas of tenderness. The liver and spleen were not palpable. The reflexes were normal.

Unfortunately, complement-fixation tests were not made. However, there was no personal or family history suggesting syphilis, and there were no stigmas.

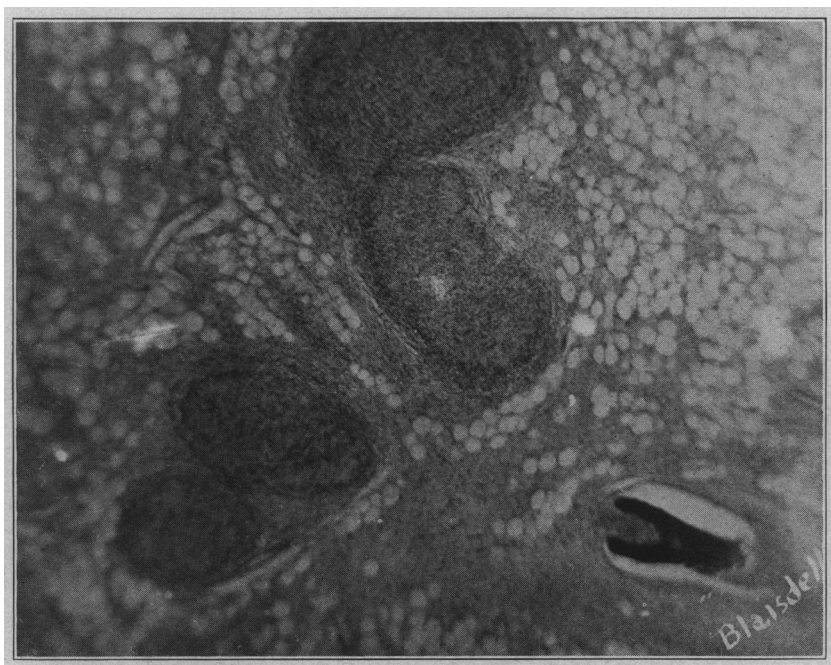


Fig. 2.—Four fibrous cords in corium. Note adipose tissue and hair follicle. Van Gieson stain.

*Treatment.*—Dr. Phillip K. Gilman, associate clinical professor of surgery, Stanford University Medical School, to whom I am indebted for the privilege of reporting the first case, removed the growth in its entirety and applied Thiersch grafts from the thigh. There were no particular difficulties connected with the procedure. The patient, after having been observed regularly for six weeks, was dismissed in "excellent condition." Since then our social service department has been unable to locate her.

*Histologic Findings.*—A piece of the growth from the summit of a convolution, including the skin, was fixed and hardened in formaldehyde solution. Sections were stained with hematoxylin and eosin, Mallory's connective tissue stain, van Gieson's stain and silver nitrate.

The tumor, preserved by the Kaiserling method No. 4, is presented for inspection. This work was done by Dr. F. E. Blaisdell, associate professor of surgery (surgical pathologist) of Stanford University.

The epidermis showed pronounced acanthosis with slight hyperkeratosis. In the upper corium the collagen showed lessened affinity for the acid stain. There were numerous young connective tissue nuclei and dense collections of pigment here and there. The blood vessels were quite numerous, dilated and their walls were not thickened. There were occasional plasma cells. Fat cells were rather numerous even in the upper regions.

The lower corium showed the chief changes. Here there were large cords or whorls of connective tissue, the bundles running in every direction. In between them were many fat cells. These structures extended down into

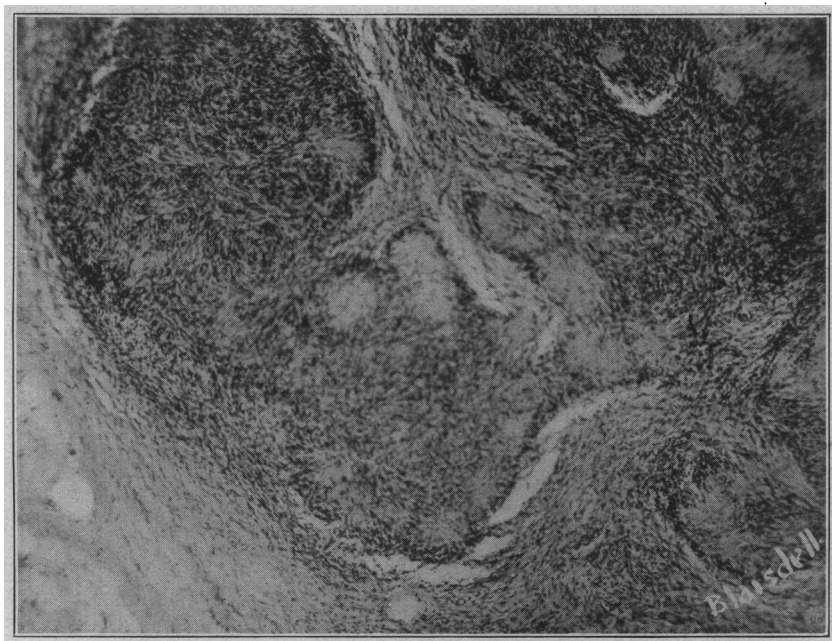


Fig. 3.—Fibrous cords under higher magnification. Hematoxylin and eosin stain.

the subcutaneous tissue where they were numerous. Many of these bundles were packed closely together.

The collagen in places took the acid dye intensely. The numerous sebaceous glands were quite large. The hair follicles were normal. Occasionally there was seen dense perifollicular infiltration of small round cells which extended the full length of the hair. Some apparently independent collections of these cells in serial sections belonged to the perifollicular deposit. No polymorphonuclear leukocytes were seen. The sweat glands were normal. No nerve tissue was seen.

*Comment.*—The general appearances and certain special features suggested a congenital connective tissue growth involving the skin. The signs of inflammation here and there were apparently incidental results

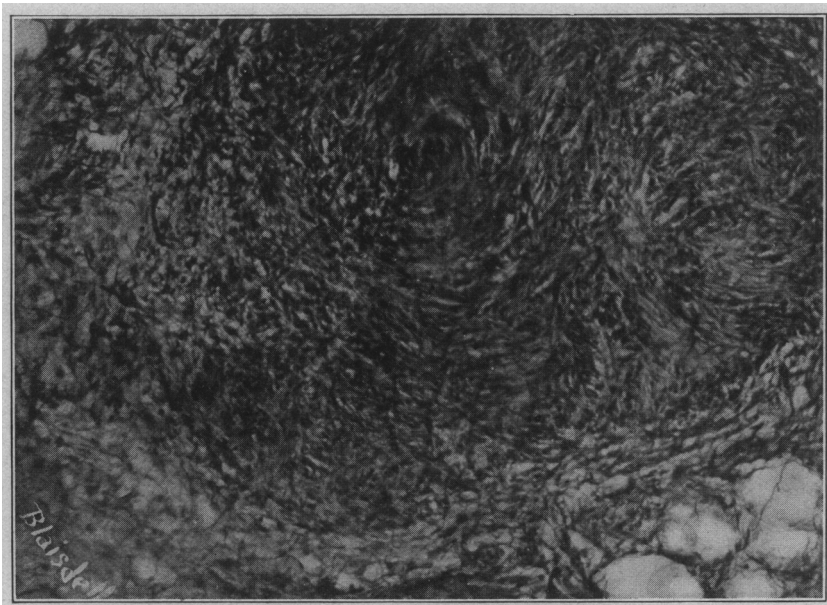


Fig. 4.—Fibrous cord under higher magnification. Note whorling and interlacing of fibers. Van Gieson stain.

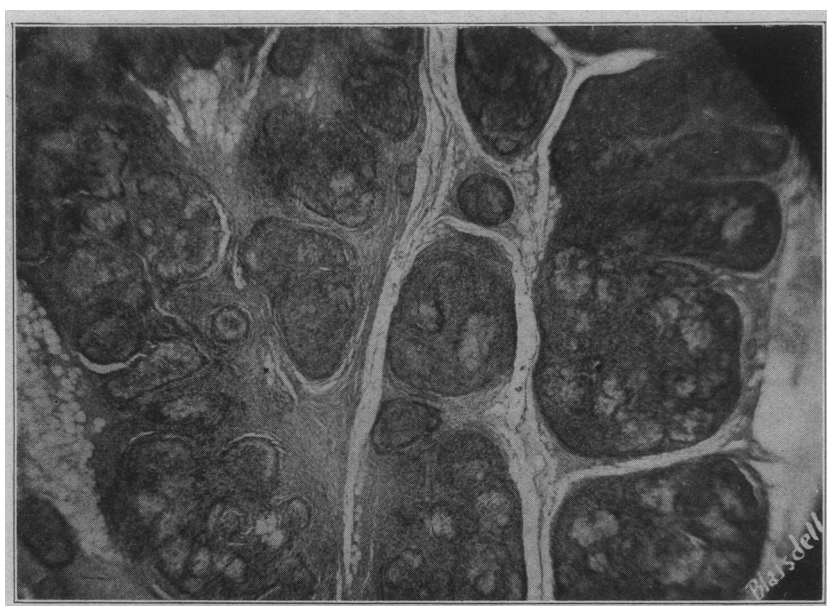


Fig. 5.—Portion of growth in subcutaneous tissue, showing the fibrous cords. Hematoxylin and eosin stain. Low magnification.

of slight traumatism. The nodules feeling like sebaceous cysts probably owed their characteristics to the arrangement of the numerous thick fibrous bundles and the uneven distribution of the fat.

*Summary.*—We have a case of cutis verticis gyrata in an otherwise normal girl, 15 years old, in good health (barring occasional headache and nausea). The condition had existed since birth and had increased in size during the development of puberty with the natural increase in activity of the pilosebaceous system. There was no history of past inflammation, as observed by various authors. The patient had a dolichocephalic type of skull, thus differing from the patients in most cases reported, in whom the brachycephalic type prevailed.

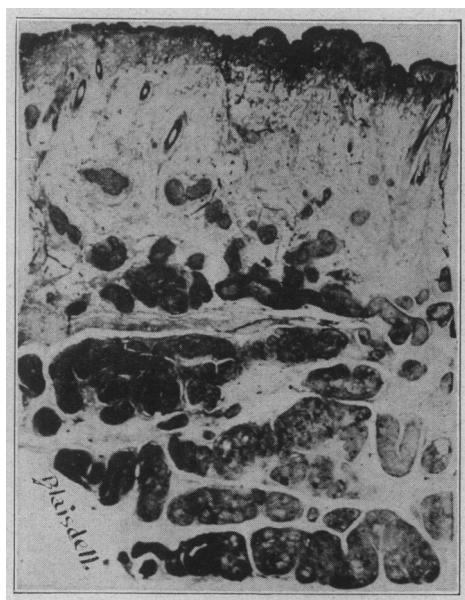


Fig. 6.—Section through entire thickness of scalp, showing growth in corium and subcutaneous tissue.

CASE 2.—I have another case under observation at the present time, in its earlier stages. The patient, a prosperous Jew, 54 years of age, obese, has a brachycephalic type of skull, a short thick neck and rather tightly drawn scalp. In the occipital region there is a deep narrow horizontal, tensely drawn, permanent furrow with a smaller curved offshoot from the same just above its right outer third. The condition has existed four or five years only. It is gradually becoming more accentuated. The hair is black, thick and normal. The patient has a seborrheal skin with typical seborrheic dermatitis in the occiput, umbilicus, groins and internatal region. The obstinate seborrheic eczema seems to be a factor in the process. I believe that the chronic inflammation there is resulting in connective tissue proliferation with consequent accentuation of the cutis verticis gyrata.

I have seen several similar but less marked cases in which the scalp was not diseased. One can see conditions approaching this in almost any crowd. A brachycephalic head with a short fat neck seems prone to develop horizontal and secondary furrows in the occiput. It is only cases showing increasing persistence of these furrows in the occiput and higher, of course, that can be regarded as early examples of cutis verticis gyrata. When chronic local folliculitis or eczema develops, naturally connective tissue proliferation occurs, and eventually the thickening of the corium and subcutaneous tissue results in the formation of this deformity.

#### DISCUSSION

DR. FRED WISE, New York: I reported with Dr. Levin what we thought was a typical case, but we were unable to perform a biopsy, which detracted from the value of the report. The second case in New York was presented by Dr. Fox two years ago.

The interesting point in Dr. Alderson's paper lies in the fact that the lesion is different from those described in the literature, the difference being that this is a decidedly circumscribed elevated growth and does not run into the scalp as all the other cases do, and I am inclined to believe that it should be interpreted as a nevus. It is a form of cutis verticis gyrata, but of nevus origin. The other cases described in the textbooks as nevus cerebelliformis are probably examples of this. The microscope reveals the facts, and the histology in those cases must be different from that described by Dr. Alderson.

Another statement occurring in the literature is that there are probably two very different types of cutis verticis gyrata, one in which there is a marked inflammatory process and the other in which no inflammatory reaction has ever been present.

DR. HARRY E. ALDERSON (closing): This summer more material from the specimen will be worked up thoroughly; sections from every part of the growth will be studied, and a complete report will be sent in later for publication.