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[*From Transactions of American Ophthalmological Society*, 1891.]

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Strictly speaking, the term neuroma should be limited to tumors composed of new-formed nerve fibres, and, while in certain instances true new formation of nerve tissue does occur, the majority of these growths are false neuromata, *i. e.*, fibromata and myxomata of the connective tissue of the nerve, unaccompanied by multiplication of its nerve fibres. Indeed, the latter usually are compressed and atrophied by the surrounding tissue. Vernueil* gave the name "plexiform neuroma," and Paul Bruns† "Rankenneurom," or "Neuroma Cirsoideum" to a morbid process of this character when it appears in the form of a more or less convoluted mass, somewhat corded beneath the skin, and affecting an entire nerve territory. In a few instances this variety of the growth has been situated in the eyelid and surrounding region, notably the temporal area. The researches of Von Recklinghausen have shown that multiple fibrous tumors of the skin often contain nerve fibres, or, to speak more accurately, are developed from the fibrous sheath of small cutaneous nerves, and thence involve the connective tissue structure of the vessels, sweat glands, and hair follicles, and are, in fact, neurofibromata. Tumors of this nature may invade the fibrous sheaths of the different tubular structures contained in the skin and sometimes reach a great size, until finally the appearance of elephantiasis is developed, which, to distinguish it from the acquired variety of this type of hypertrophy, is designated by some authors as "congenital elephantiasis." Under such circumstances the tumors may not be circumscribed, but an overgrowth of the skin takes place, giving rise to an extensive hypertrophy.

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* *Archives Générales de Medicine*, 1886, Vol. II., V serie, T 18.

† *Virchow's Archiv.*, Vol. L, 1870, p. 80.

To the cases of so-called plexiform neuroma of the eyelid already upon record I desire to add another case of congenital tumor of this region, which presented itself in the form of considerable thickening of the eyelid, causing an extensive ptosis and a morbid growth in the temporal region, clinically giving rise to the appearance of a fibro-fatty tumor, or to that condition which has received the name elephantiasis, and which in rare instances has appeared as a congenital affection in the upper lid. Examination of the slides and of the photo-micrographs will, however, demonstrate that in addition to hypertrophy of the glandular structures in the skin and of the connective tissue, the most marked pathological lesions are large masses, which in cross section appear as concentric whorls of soft fibrous tissue, containing in their center more or less degenerated nerve fibres.

The history of the case is as follows: Edward Hawkins, aged 20, was admitted to the Philadelphia Hospital September 2, 1890, and came under the care of my colleague, Dr. G. M. Gould, on account of the condition which is represented in the accompanying photograph, namely, a large, irregular tumor of the right temporal region, ptosis, and thickening of the upper lid and the tissue between it and the margin of the brow. When I took charge of the wards I obtained the following history: The tumor was present at birth, and, indeed, had not materially changed in its appearances except by its expansion with the corresponding growth of the tissues in which it was situated. The distance from the edge of the brow to the lower margin of the swollen eyelid was 7.5 cm.; the longest transverse diameter, *i. e.*, from commissure to commissure, 9 cm., and the thickness varied from 1 to 1.5 cm. The temple tumor extended from the outer commissure to the ear, and from the margin of the hair to the zygoma. The skin over the area of the growth was slightly brown in color, a pigmentation which had developed since the birth of the patient. To the palpitating fingers the underlying mass gave the impression of a somewhat lobulated, slightly corded growth. There was no pain and no inconvenience, save that caused by the ptosis. Both eyes were normal in structure and vision, and there were no congenital faults

or tumors elsewhere in the body, neither had a similar growth been present in any member of the patient's family or ancestors. The palpebral conjunctiva of the affected lid presented the appearances of a chronic blenorrhœa, and across the lid, on its conjunctival surface, there were three furrows, bounded by corresponding ridges, and several transverse divisions, so that the whole presented a somewhat lobulated appearance. There was

FIG. 1.

NEURO-FIBROMA OF THE RIGHT UPPER EYE-LID AND ADJACENT TEMPORAL REGION.

moderately free secretion of muco-pus. Some attempts to reduce the size of the tumor by local blood-letting had met with indifferent success. The first operation was done January 6, 1891, and consisted in removing a semi-lunar flap from the lid, dissecting out all of the affected tissue which it was possible, and afterwards approximating the edges by means of sutures. After the operation the long transverse diameter of the lid measured 5 cm. The second operation was made January 20, 1891, and consisted in an attempt to overcome the ectropion which had resulted from the first operation. The outer commissure

was slit for a distance of 5.5 cm., and from the end of the incision a second one was carried forward and backward for 5 cm., a parallelogram of skin and subcutaneous tissue, together with the underlying tumorous mass, was dissected away from the temple, and the edges were approximated. Twenty millimeters of the thickened outer edge of the upper lid were cut off, the lower lid pared correspondingly, and the two united with sutures as in the

FIG. 2.

FROM A PHOTOGRAPH AFTER THE THIRD OPERATION. THE ARTIST IN REDRAWING HAS SOMEWHAT EXAGGERATED THE EFFECT.

operation for tarsorrhaphy. This operation relieved the ectropion, and when the patient looked directly forwards the lid fissure was separated about one centimetre. A third operation was done in April, 1890, which consisted in Panas's operation for ptosis slightly modified, owing to the thickened tissues. This yielded a fair result, elevated the lid, and gave the boy a reasonable palpebral opening.

The tumorous masses which had been dissected from the lid, and to less degree from the temple (the mother positively declined to allow the whole mass from the temple to be removed)

were placed in Mueller's fluid, carefully hardened, and sectioned for the microscope, as were also the pieces of skin which had been removed from the temple and from the eyelid. The following are the most important microscopic lesions:

Integument of the Eyelid. The horny layer of the epidermis is thickened, uneven, its flat cells being somewhat discolored; both the granular layer and the rete mucosum are thicker and more prominent than is normal in the skin of the eyelid. As is natural to the part, the papillae of the pars papillaris are not well marked in this layer, and the pars reticularis of the corium is not readily separated; the entire corium appears to be composed of a loose fibrous connective tissue containing numerous wavy bundles, free corpuscles, and elastic fibres. Prominent features in this portion of the microscopic field are the hypertrophy of the sebiparous glands and the wide dilatation of the lymphatic spaces which sometimes assume large areas lined by distinct, flat, endothelial cells, in close relation to wide-mouthed, thin-walled veins (Fig. 2, Pl. I). Here and there in some sections racemose and tubular glands with hypertrophied walls are present. In the deeper portion of a section made directly through the integument, the first portions of the true tumor appear in the form of the strands presently to be described, which are the characteristic feature of the growth.

Main Body of the Growth. This is composed of strands which pass through a framework of loose connective tissue containing fat cells and large blood-vessels, lymphatics, and muscle fibres. The strands themselves, when seen in cross section, are composed of concentric whorls of loose fibrous connective tissue containing many nuclei and in their center medullated nerve fibres, sometimes intact and sometimes partially destroyed by fatty degeneration. Figure 1, Pl. I represents the general outline of a cross section of several of the strands seen under a low magnifying power, while these other sections exhibit various appearances of the center of the whorls under a higher magnifying power. In order to positively demonstrate the presence of medullated nerve fibres, which, however, are readily seen in many of the sections stained by a carmine nuclear dye, the method of Weigert was employed, and Figures 3 and 4, Pl. II

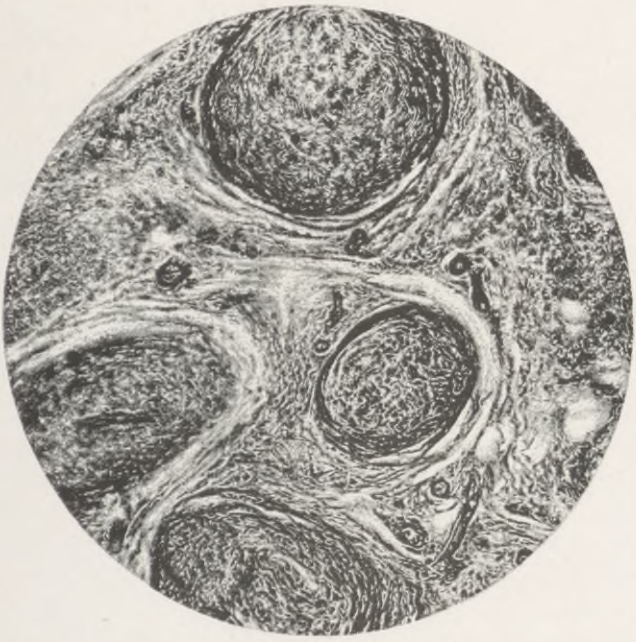

FIGURE 1. Section of the tumor, showing the concentric whorls.
[× 125.]

FIGURE 2. Vertical section of the skin of the eyelid, showing hypertrophied gland and dilated lymph spaces.

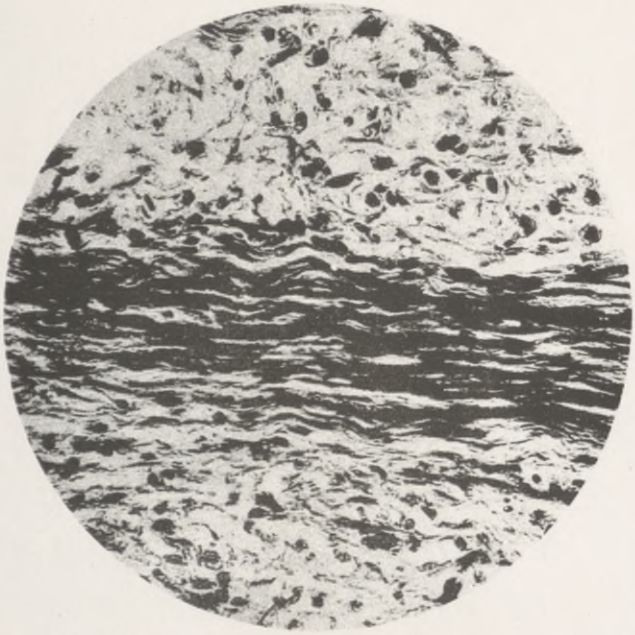

FIGURE 3. An oval whorl, showing a band of medullated nerve fibres passing through its centre. [× 250, Weigert's stain.]

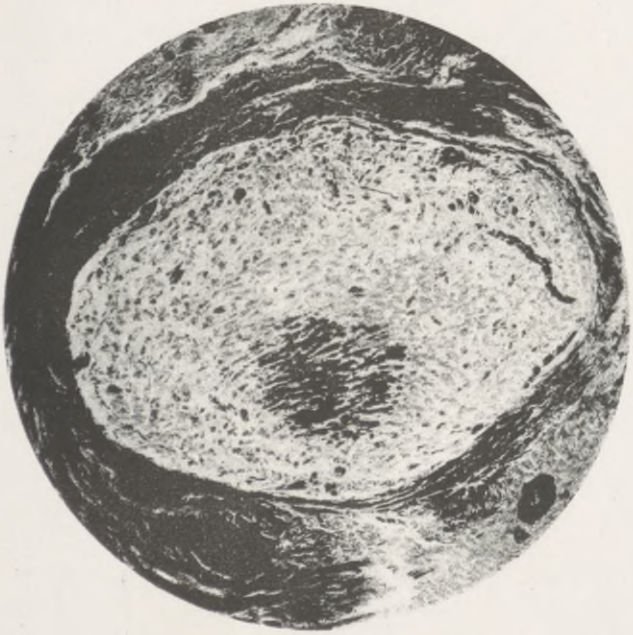

FIGURE 4. Small, oval whorl, showing in its centre a patch of medullated nerve fibres. [× 125, Weigert's stain.]

represent the various appearances elicited by this method of staining. In Figure 3 the oval whorl is seen to contain directly in its center a band of medullated nerve fibres, while on either side the remains of the loose connective tissue, nuclei, and here and there some myxomatous change are visible. Additional sections represent, on the one hand, an oval whorl, and on the other an irregular-shaped whorl with nerve fibres in their centers, while in Figure 4 a small round whorl is depicted with the nerve fibres cut transversely and placed somewhat laterally. All of the photo-micrographs were taken with apochromatic objectives, 16 mm., 8 mm., and 4 mm. of Zeiss, on orthochromatic gelatine dry plates, by sunlight illumination rendered monochromatic. The photo-micrographic illustrations and the microscopic slides were prepared by Dr. William M. Gray of the Army Medical Museum.

The literature of plexiform neuroma is not an extensive one. Marchand* has collected twelve cases, and Cartaz† six cases, and in five, perhaps six, of these eighteen cases the growth has been situated in the region of the upper eyelid and temporal area. Since the paper of Marchand and Cartaz no very elaborate work on the subject has been done, although here and there scattered reports appear, not, however, with reference to a situation in the eyelid. Interesting observations will be found in the papers of Christol,‡ Garel,§ and especially in the research of Lacroix and Bonnaud,‖ the latter observers having made a careful histological examination of a plexiform neuroma situated in the back in the neighborhood of the scapula, and presenting the rare characteristic of being largely composed of amyelinic nerve fibres. The development of plexiform neuroma belongs in the majority of instances to fœtal life. Some hereditary tendency has been shown, but not with great definiteness. When this has existed it seems to have concerned that form of the tumor which partakes more of the nature of an elephantiasis than of a neuroma. When these tumors occur in the region with which we are at present interested, they appear in the form of a some-

* *Virchow's Archiv.*, Bd. LXX, page 36.
† *Archives Générales de Medicine*, August, 1876.
‡ *Gaz. Hebdomadaire*, 1870.
§ *Lyon Médical*, 1887.
‖ *Archives de Médicine Expérimentale et d'Anatomie Pathologique*, II., 1890, p. 411.

what lobulated tumor, which gives the impression to the palpating finger that the tumor is composed of fat and connective tissue, with perhaps occasionally an ill-defined sense of fluctuation. It may and has been mistaken for congenital lipoma, encephalocele, hydromeningocele, cystic tumor, and cavernous angioma. In fact, it would be practically impossible in any of the cases to have stated with certainty that the tumor was composed either in part or entirely of nerve fibres. Pain, as is usual with neuromata, was not a marked feature in any case, although tender spots have sometimes been found. As Lacroix and Bonnaud very properly insist, the diagnosis of a growth of this character can rest alone upon a careful histological study. In a number of such careful examinations it has been shown that the interstitial tissue between the cords or strands which form the characteristic gross appearance of the tumor, is composed of a fasciculated connective tissue of loose construction. The fasciculi often arrange themselves in the neighborhood of the cords in a concentric manner, and around these, and separated from the cords themselves, are well-defined lymph spaces in which an endothelial lining could be detected. In one case the tissue of the tumor had a myxomatous character, and one observer has seen in the tumor a new connective tissue. Often in the interstitial substance well-formed sections of fat cells are found. Vessels are frequent. The cords, which are the essential part of the tumor, for the most part exhibit a clear peripheral zone which is composed of concentrically-arranged fibrillar connective tissue. Inward from this follow loose connective tissue strands accompanied by many nuclei. Usually in the center, but sometimes irregularly-placed, medullated nerve fibres may be found in all stages of degeneration, but sometimes well preserved. Non-medullated nerve fibres have also been demonstrated, and, as has just been quoted, in one instance the growth was largely composed of them. Through the cords more or less rich vessel formation is present, and in the largest cords the nerve fibres may be poorly developed, or fail altogether. The skin which covers the tumor generally is in a condition of distinct hypertrophy, which especially concerns the connective tissue of the cutis. In some cases hair follicles and sweat glands are hypertrophied, and the epidermis often shows pigmentation in the deeper layers

of the rete Malpighii. Various opinions in regard to other fine details have been recorded by different authors. Czerny and Winiwarter picture an increase of the cells of the vessel walls and the capillaries, arteries, and veins, which is also present in the subcutaneous tissue and the skin, and according to the latter observer, help in the new formation of the connective tissue. A similar development has been supposed to be present in the vessels of the muscles. Cell proliferation of the sheath of the nerve fibres and of the sarcolemma of the muscle fibres is present, and also contributes to the connective tissue formation. Czerny and Winiwarter promulgate the hypothesis that the whole affection is a disease of the trophic nerves. Whether there is really a development of new-formed nerve fibres is a question.

This *résumé* of the microscopic lesions found in so-called plexiform neuroma has been quoted in large part from Marchand's monograph, and it will be seen that it accords with the histological characters of the tumor to which I call your attention to-day, and which hence must be recognized as belonging to this group of cases. The association of the disease with fibroma and elephantiasis congenita has been referred to, and Bruns calls attention to the commingling of circumscribed elephantiasis mollis congenita and neuroma, in their relation to an elephantiasis proper, which consists of an alteration of the subcutaneous connective tissue, or one in which there is enormous development of the vessels (elephantiasis teleangiectodes). Therefore, a congenital elephantiasis with excessive development of the nerve tissues may be classified as an elephantiasis neuromatodes; and Bruns further recites from an observation of two clinical cases, that almost imperceptible transitions occur from simple elephantiasis to elephantiasis neuromatodes, or neuroma elephantiasticum. A brief description of the most important cases of so-called plexiform neuroma of the eyelid and adjacent temporal region which have been recorded is here appended:

CASE I. Boy, aged 6; tumor of the right upper eyelid reaching to the temple, composed of cords in the center of which was a nerve which had partly undergone fatty degeneration. (Billroth, Archiv. f. klin. Chirurg., 1863, IV., s. 547.)

CASE II. Boy, aged 18; plexiform tumor of the upper eyelid and the neighboring temporal region. When freed from fat it was seen to be composed of grayish-white, smooth cords of plexiform arrangement sometimes ending in fine nerves. The center of the cords was composed of atrophic and fatty degenerated nerves. (Billroth, Archiv. f. klin. Chirurg., Bd. XI., 169, s. 232.)

CASE III. Man, aged 28; part of the tumor, which occurred in the temporal region, reaching to and affecting the outer palpebral commissure. (Bruns, *loc. cit.*)

CASE IV. Man, aged 33; tumor of the left upper eyelid and temporal region, and at the same time general neuromatosis and multiple neuromas of both vagi. (Bruns, *loc. cit.*)

CASE V. A brother of the preceding case; neuroma of the left upper eyelid and corresponding temporal region. (Bruns, *loc. cit.*

CASE VI. Boy, aged 13; tumor of the left upper eyelid and corresponding temporal region. (Marchand, *loc. cit.*)