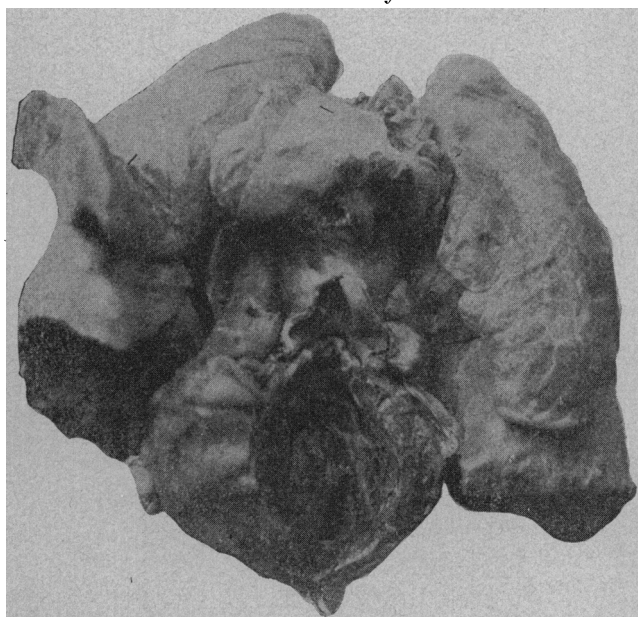


have contributed to the sclerotic tendency, together with the important evidence of a congenital heart lesion; all of these by themselves inconclusive signs, we believe to be sufficient to warrant the diagnosis of latent hereditary syphilis, with consequent degeneration of the arterial walls and aneurism. The father's spurious epilepsy and his suggestive venereal history render the conclusion practically certain.

It would appear that the case under discussion is the only instance noted in the literature of death by rupture of an aneurism of the thoracic aorta at such an early age. We find a number of instances of aneurism of the thoracic aorta, of the cerebral arteries, also of the abdominal aorta, usually discovered at the autopsy. One of the youngest subjects that has come to our notice suffering from aneurismal involvement is a child (mentioned in the Reports of the City Inspector of New York for the period 1856 to 1864) between 2 and 5 years (not of the arch), one between 5 and 10 years of the arch, two between 10 and 15 (neither of the aorta), and an increasing number as the age rises.

Le Boutillier<sup>1</sup> has also recently collected a series of



Ruptured aneurism of aortic arch in child of four years.

cases of aneurism in early life, and himself reports one of the thoracic aorta in a child of 9 years, still living. In his bibliography there are noted seven cases of thoracic aneurism in children under 10 years of age, all dying of some independent pathologic condition, or still alive at the time of the report. The youngest of these cases was 2 years of age, the death being due to hemorrhagic smallpox, not to the aneurism.

In most instances these aneurisms are the result of hereditary syphilitic change in the vessel walls, perhaps rarely to tuberculous or rheumatic arteritis, far less frequently (probably never in the case of the aortic arch) to thrombosis and other causes. In Le Boutillier's case, as in that under discussion, there would appear to be a possibility that the paroxysmal effort in whooping cough was a contributing causal influence.

While the case reported in this paper is undoubtedly an instance of a most rare condition, it is none the less of practical interest in its bearing on the diagnosis of intrathoracic conditions in very young children.

1. Amer. Jour. Med. Sci., May, 1906.

## NEUROFIBROMA OF THE ORBIT: KRÖNLEIN OPERATION.\*

WALTER R. PARKEP, B.S., M.D.

Professor of Ophthalmology in the University of Michigan.  
ANN ARBOR, MICH.

Cases of neurofibromata (pseudo-neuromata, solitary neuromata) of the orbit are extremely rare, there being but two cases on record. Of these, one reported by Tersch<sup>1</sup> involved the lachrymal branch, and one reported by Marchetti<sup>2</sup> the infraorbital branch of the fifth nerve.

The ophthalmic division of the fifth nerve has been involved in multiple neuromata, and Bietti<sup>3</sup> has described amputation neuromata of the ciliary nerves after optic-ciliary-neurotomy. Houel<sup>4</sup> has described small tumors of the third, fourth, nasal and frontal branches of the fifth nerve, found postmortem, which had never caused clinical symptoms.

The extreme rarity of neurofibromata is shown by the following statement of Tersch: "A solitary neuroma is altogether a rare tumor and its location in the orbit



Fig. 1.—Photograph of patient before operation.

appears at least a curiosity;" and of Marchetti, who, after eliminating the plexiform neuromata as described by Billroth and others, says: "The exposition of the case shows its importance; a tumor rare as a pathologic type and unique in consideration of the place of its development. In fact, in the literature which I have been able to consult I have found no example of tumors developing through the infraorbital nerve."

A short review of the two published cases will be given in this paper, and a report added of a tumor of similar nature removed by me. The whole will be treated from a clinical standpoint, referring those who are interested in the detailed pathologic findings to the reports of Tersch and Marchetti.

MARCHETTI'S CASE.—*History*.—Male, aged 30. At the age of 5 the mother first noticed on margin of lower lid a nodule

\*Read in the Section on Ophthalmology of the American Medical Association at the Fifty-eighth Annual Session, held at Atlantic City, June, 1907.

1. Arch. f. Ophth., 1902.

2. Ann. di Ottalmologia, 1904, xxxiii.

3. Arch. f. Ophth., 1900.

4. Lagrange: Etudes sur les tumeurs de l'oeil.

of reddish color with smooth surface. At the end of one year it had acquired the size of a cherry, producing lagophthalmus. At the age of 14, nine years later, the tumor had reached the size of a medium-sized pear, the larger part hanging down on the cheek. There was no pain, but considerable tenderness on pressure. Other than the drawing down of the lid, the eye was apparently normal. There was a history of frequent hemorrhage following a slight abrasion. The tumor gradually increased in size and extended upward and backward into the orbit with final loss of function of the eye. The skin covering the tumefaction, at first but slightly changed, now assumed the aspect of elephantiasis and secreted a considerable quantity of limpid fluid. The tumor became extremely sensitive with frequent shooting pains extending over brow and head. Intellectually, the patient was subnormal, of fairly robust appearance and otherwise healthy. Family history negative. The tumor finally assumed the size of the head of a newborn child, extending downward beyond the chin with the stem rather large, extending into the orbit. The arch and inner wall of the orbit were normal; the outer wall, however, was pressed outward one and one-half centimeters. The lower wall could not be examined. The eye, greatly changed in appearance, was crowded upward and forward. The orbit was filled with a mass which was continuous with and formed part of the tumor. It was hard, apparently of a fibrous nature and covered with thickened skin which was movable on the growth. The orbital portion was slightly movable.

*Operation.*—In May, 1900, under chloroform anesthesia, the tumor, together with the eye, was extirpated without difficulty,

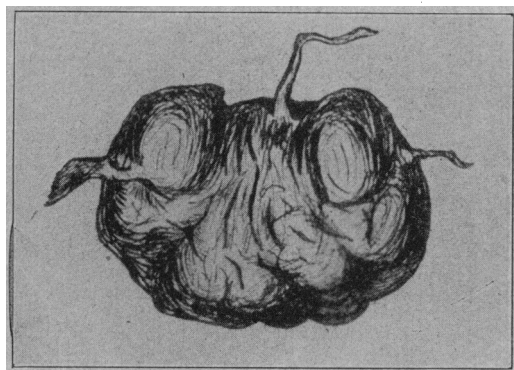


Fig. 2.—Tumor,  $1\frac{1}{2}$  times natural size.

there being no attachment to the orbital wall. Recovery was uncomplicated. The lower orbital wall was found converted into a groove in which the neck of the tumor rested.

*Macroscopic Examination.*—The tumor was found to consist of two portions well differentiated one from the other. The orbital portion about the size of a fist, egg-shaped, with well defined capsule. On the anterior surface was the eye, greatly modified. The lower part of the posterior surface was surrounded by branches of the infraorbital nerve, which were normal in appearance and size and so intimately connected with the tumor that they could not be removed from it without tearing. Attached to the lower and outer part of the orbital tumor was the mass which protruded on the cheek.

*Microscopic Examination.*—Only the orbital portion, it was found, could be considered as the tumor proper, the lower part being exclusively represented by the characteristics of elephantiasis of the skin.

After a detailed description of the tumor, together with the microscopic findings, the author made the diagnosis of false neuroma.

*TERSCH'S CASE.*—*History.*—Female, aged 43. Three years previously the patient noticed that the unaffected eye was deeper in and higher up than its fellow. A diagnosis of retrobulbar tumor was made and operation advised, but this the patient refused. The condition remained the same for two and one-half years, when the tumor began to grow rapidly. The patient then consented to an operation.

*Examination.*—Behind the upper lid of the right eye could be

felt a resistant tumor, which rested on the bulb like a cap, extending from the trochlea to the lachrymal gland without being attached to either. The finger could be easily forced between the tumor and the eyeball. The tumor pushed the eyeball down and out, so that the upper limbus of the cornea was on a level with the lower pupillary border of the other eye. The exophthalmus was 12 mm. The movement of the eyeball except upward was free. Left eye normal. Fundi normal. Examination revealed no other tumor or swollen lymph glands. Patient was not examined for disturbances of sensibility in the fifth nerve areas, but had no pain or parasthesias.

*Operation.*—The operator, Professor Fuchs, made an incision parallel to the eyebrow, and, entering the orbit, removed a rather large tumor, apparently a lipoma, which seemed to be sharply circumscribed. Removal was not difficult, the growth being connected with the surrounding tissue merely by a few thin strands. No other tumor could be palpated. The wound healed readily. There resulted a slight ptosis. Exophthalmos disappeared and the eyeball receded. After six months the patient appeared normal, the two eyes similar in appearance, and no further tumor formation noted.

*Examination of Tumor.*—The tumor appeared egg-shaped, 3.5x2.75x2.5 cm. in size, of soft consistency, yellow color, lobulated upper surface. A distinct capsule could not be seen

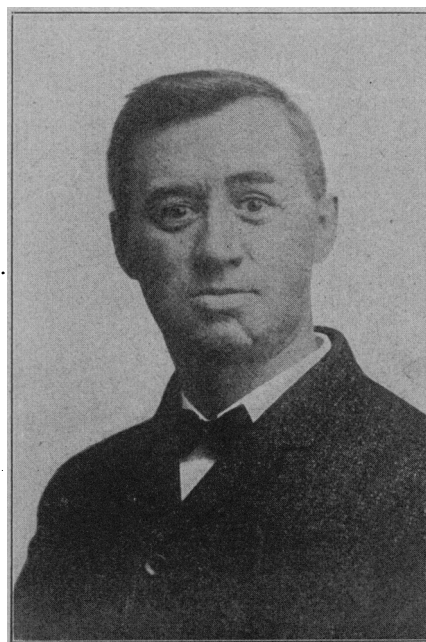


Fig. 3.—Photograph of patient six months after operation.

macroscopically. On account of its consistency, the tumor was diagnosed a lipoma. At one pole of the tumor a nerve was found entering the growth.

According to the pathologic and histologic findings, there was no doubt that the growth was a so-called pseudo-neuroma (neurofibroma), which is called in the literature a solitary neuroma. It was difficult to decide from which nerve the new growth sprang; but the author, by exclusion, concluded that it was a neuroma probably of the lachrymal branch of the fifth nerve. So much for the two cases on record.

*THE AUTHOR'S CASE.*—*Patient.*—Male, aged 28, referred to the ophthalmologic clinic from the oto-laryngologic clinic of the University of Michigan, in November, 1906.

*History.*—The patient gave a history of exophthalmus of the left eye, gradually increasing during the preceding twelve years. He had been subject to attacks of acute mania during the past six years. There was present no pain nor discomfort in the eyes except from double vision, which was especially noticeable if looking upward. No fundus changes. Vision about 20/30. The patient being illiterate and mentally substandard, accurate results could not be obtained.

**Examination.**—The skin of the upper lid as well as that surrounding the orbit was normal. Deep in the orbit could be felt a resistant tumor which seemed to surround the bulb, but was found not to be attached to it. It was difficult to determine before the patient was anesthetized whether the tumor was movable or not. No well-defined demarcation between the tumor and the superior orbital wall could be felt. The exophthalmos was marked, outward about 12 mm. and downward so that the upper margin of the cornea was on a level with the lower pupillary margin. The movements of the globe, with the exception of upward, were normal. Right eye normal. Fundi normal. Patient was of medium stature and except for his mental condition, apparently healthy. The lymphatic glands were not enlarged, and with the exception of a small nodule on the brow, there were no other tumors of the skin or other parts of the body. Examination of the sensibility disturbance of the trigeminus branch was unsatisfactory. A nodule, about the size of a cherry, was situated on the brow just above the supraorbital notch. With the idea that there might



Fig. 4.—Photomicrograph of tumor showing entrance of nerve (x24).

be some relation between this nodule and the orbital tumor, it was determined to remove the nodule and submit it to examination. When removed, it had every appearance of a lipoma. A preliminary report from the pathologic department was fibroma, possible neurofibroma. Deep palpation while the patient was under ether revealed the fact that the orbital tumor was movable.

**Diagnosis.**—The following diagnostic points concerning the orbital tumor were obtained: 1. A non-malignant growth as revealed by the history and pathologic report. 2. The displacement of the globe outward and downward indicated that the tumor was situated outside and above the muscle cone. 3. It was movable.

**Operation.**—It was decided to attempt the removal of the orbital tumor after the method of Krönlein. A curved incision was made beginning over the temple, extending along the outer margin of the orbit, dividing the periosteum, and backward along the upper edge of the zygomatic arch. The periosteum lining the inner side of the outer wall of the orbit was resected, together with the orbital contents. The lateral wall of the orbit was cut through with a chisel in two places, above,

in the suture between the great wing of the sphenoid and the malar bone, and below in a horizontal plane passing outward in a line directly above the insertion of the zygomatic arch. This piece of bone with its muscle and cutaneous attachments was forced backward, giving free access to the orbit. The periosteum was divided in a horizontal direction and the tumor then extirpated without difficulty. After removal, the periosteum was sutured with catgut, the bone and soft parts replaced, and the skin sutured with silk. Union was perfect and recovery uninterrupted. The excursion of the eye became normal and the vision remained the same as before the operation.

**Macroscopic Examination.**—The tumor was somewhat kidney-shaped and measured 35x25x9 mm. The surface was nodular and had the appearance of a lipoma. Protruding from the tumor were three nerves, which a careful examination seemed to prove to be the frontal externally, the supratrochlear below, and the supra-orbital nasally. When held before the light, the larger nerve was seen to divide into two branches, and the course of each could be traced through the mass. The tumor apparently had its origin in the sheath of the frontal branch of the ophthalmic at the junction of the supratrochlear, and supra-orbital nerves.

Following is the report from the pathologic laboratory, for which I am indebted to Professor A. S. Warthin:

**Microscopic Examination.**—Tumor consists of fibrous connective tissue loosely arranged in moderately coarse fibrils with large areas of a more finely fibrillar connective tissue, resembling edematous connective tissue. The spaces between the fine fibrillæ are large and contain a finely granular precipitate. The appearance is that of an edematous fibroma. Running through the center of the mass of connective tissue are nerve fibrils, in part connected as in a nerve trunk, but for the greater part separated by connective tissue. These nerve fibrils show no increase in size, and appear normal. The connective tissue hyperplasia is limited chiefly to the connective tissue of the pari- and epi-neurim, that of the endo-neurim being affected to a less degree. The growth is poorly supplied with vessels, the veins showing a chronic congestion.

**Diagnosis.**—Edematous neurofibroma. (Fibromatosis of nerve trunk).

Considering the three cases as a class, there are some peculiar similarities. Two of the cases were sub-standard mentally, suggesting the possibility of similar intracranial nodules. In two the tumors were diagnosed macroscopically lipomata. In all three the pathologic findings were the same, although in one there was elephantiasis of the lid.

As regards the method of operating, I believe that if the exact condition present had been known the tumor could have been removed from above, as was done in the Tersch case. No procedure, however, could have been more satisfactory than the Krönlein operation, and it offers better opportunity to meet any complications that may be present.

[FOR THE DISCUSSION, SEE THE DEPARTMENT OF SECTION DISCUSSIONS IN THIS ISSUE.]

## SUPRARENAL HEMORRHAGE; AN UNUSUAL CAUSE OF SUDDEN DEATH.\*

J. F. MUNSON, M.D.

SONYEA, N. Y.

I am indebted to Dr. B. F. Andrews, interne at the Colony, for the history and clinical notes in the following case:

**Patient.**—G. D., male, aged 25, a native of New York and by occupation a farmer, was admitted to the Craig Colony in 1901, at the age of 19 years.

**Family History.**—Mother was hysterical both before and

\*From the Laboratory of the Craig Colony for Epileptics.