

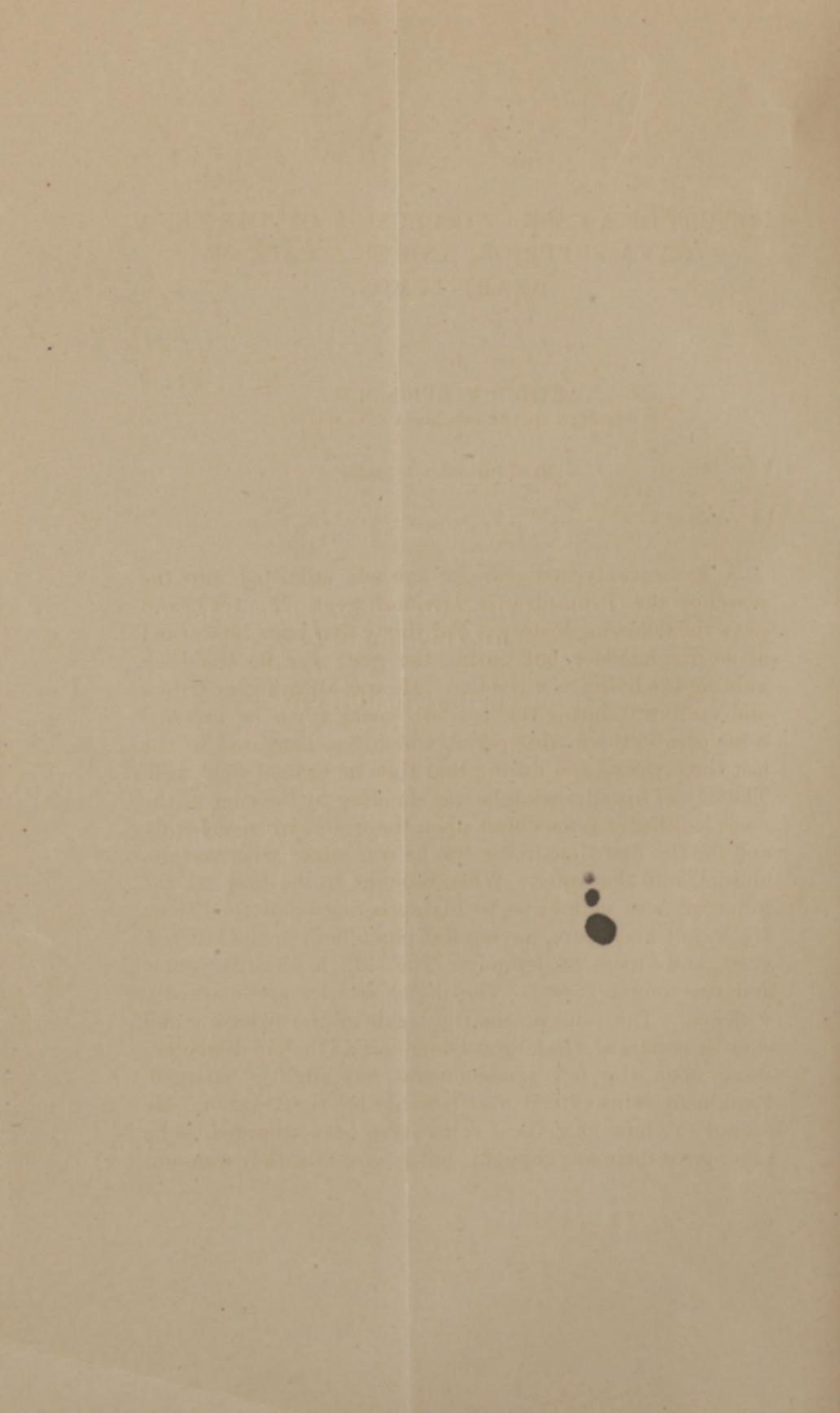
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EXTRACTED FROM THE

TRANSACTIONS OF THE COLLEGE OF PHYSICIANS OF
PHILADELPHIA,

THIRD SERIES, VOLUME VIII.





REPORT OF A CASE OF OCCLUSION OF THE VENA
CAVA SUPERIOR, AND OF A CASE OF
HEART TUMOR.

By ✓

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[Read December 3, 1884.]

A MAN seventy-two years of age was admitted into the ward of the Pennsylvania Hospital Sept. 18, 1883, and gave the following history: For thirty five years he worked as an iron-moulder, but during the past year he has been gaining his living as a peddler. He was always very strong and well until during the last two years, when he has had some pain in the lumbar region which has increased in the last three weeks, and during that time he has not slept well. The day of his admission he was standing by the edge of the river looking at some object upon the water, when suddenly and for the first time in his life he was seized with vertigo, and fell into the water. When brought to the hospital the following was noticed as to his appearance and condition: He is tall and spare, having lost much flesh in the last few years, and stoops moderately. The skin is slightly yellow and the tongue furred. The radial arteries are markedly stiffened. The veins on the right side of the abdomen and chest are enlarged, the largest being half an inch in diameter; those upon the left are comparatively slightly enlarged. Prominent veins extend also toward the right axilla. He cannot say how long these veins have been enlarged, as he never gave them any thought, but is sure that they were not

always so. Examination of the heart reveals a murmur which precedes the impulse and the carotid beat. There is slight visible impulse in the epigastrium. At the base of the heart, the sounds are almost inaudible, but they can be heard beneath the sternum; no accentuation of the second sound can be distinguished at the base, although at the apex the second sound is loudly accentuated. The hepatic dulness in the nipple line begins at the sixth rib, and extends slightly below the costal arch. The radial pulse is irritable, but the beat is not increased when the arm is elevated.

September 21. It was noted, the cardiac impulse is in the fifth interspace two inches to the left of the sternum and is rather feeble; there is somewhat more forcible impulse beneath the lower end of the sternum and some visible throbbing of the abdominal aorta. Percussion one inch to the left of the sternum shows the cardiac dulness to begin in the third interspace, and there is flatness at the fourth rib level. Transverse percussion at the fourth rib level shows dulness, beginning at midsternum and extending to the left about four inches. There is distinct impulse in the second interspace to the left of the sternum, and this appears to precede the apex impulse; there is also impulse in the second right interspace, which also precedes the apex beat. There is dulness on percussion in the first interspace to the right of the sternum and in the third interspace. The percussion resonance at the corresponding areas on the left side is natural. There seems to be impaired resonance upon percussion over the upper part of the sternum. The presystolic murmur at the apex is distinct. At the pulmonary area the sounds are very faint but distinct, and the same is the case at the aortic area. At the ensiform cartilage both sounds are distinct, but the first is rather thumping, and is accompanied by a faint blowing murmur which is synchronous with the beat of the carotid artery. There is no undue pulsation of the arteries of the neck, and the veins are full, but do not pulsate. There is a systolic murmur audible in the carotids which is increased by pressure; this is also dis-

tinct in the subclavians. There is marked systolic murmur in the femorals and in the abdominal aorta as low down as the umbilicus. Lung examination shows slightly impaired percussion resonance at the right apex, while the note is full at the left. The respiratory sounds are slightly more feeble at the right than at the left apex. Posteriorly the percussion note is short, but not markedly dull. The respiratory sounds are unusually feeble.

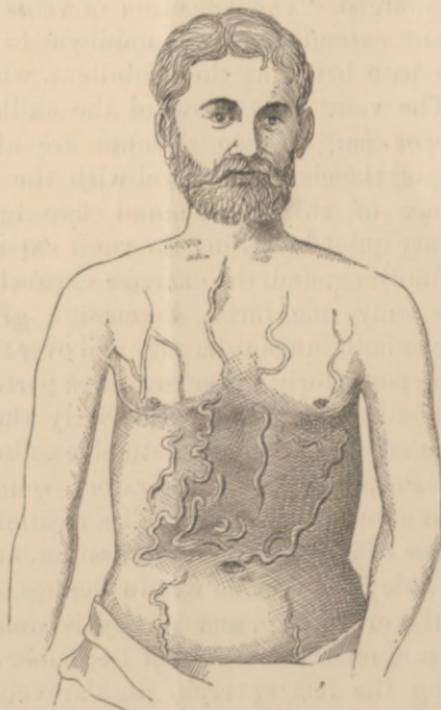
23*d.* The radial pulse is delayed. Pressure upon the enlarged veins at the lower third of the sternum causes the portion of them below the spot pressed upon to become less prominent and full, and produces a sensation of fulness over the eyes. The veins in the right iliac region tend to be enlarged.

28*th.* When the patient is in the erect position, the veins on the left side of the chest between the nipple and the sternum are enlarged. The collection of veins is somewhat cone-shaped, and extends from the umbilicus to the ensiform cartilage, the base being at the umbilicus, which it partly surrounds. The veins going toward the axillæ and those upon the upper part of the sternum are also enlarged. Measurement of the chest on a level with the nipple shows a circumference of thirty-three and five-eighths inches. During ordinary quiet breathing the chest expansion is only one-eighth of an inch, and the extreme expansion on forced inspiration is only one inch. Percussion gives a sound almost dull over both lungs anteriorly and over the upper part of the left lung posteriorly; over the lower portions the note is fuller, but still very short. Anteriorly the respiratory murmur is exceedingly feeble on both sides, almost wanting on the left. Posteriorly the respiratory sounds are very feeble, more so above than below. The treatment consisted at various times of iron, iodide of potassium, and strychnia. During the whole time that he was in the hospital he complained bitterly of vertigo, and finally became very much disheartened, and on the morning of December 12th cut his throat, severing the left external jugular vein. After he

had lost a great deal of blood, the vein was ligated and he lingered until December 20th, when he died.

The clinical history was taken by Dr. Charles Baum, and I am indebted to Dr. Da Costa for the opportunity of continuing the history of the case after I gave up the charge of the ward. The post-mortem examination was made and the note of it furnished by my colleague Dr. Longstreth, who is pathologist to the hospital. It was the Resident Physician, Dr. W. J. Taylor, who first drew attention to the facts that pressure upon the enlarged external veins caused a sensation of fulness in the head and suffusion of the eyes, and that the blood current was downwards in these veins, and it was from these symptoms principally that I was enabled to make the diagnosis.

FIG. 1.



Autopsy.—The first step in the examination was to inject the venous system. An opening was made on the right side of the neck, and the large vein was raised up in the middle cervical portion. The injection-matter used was gelatine colored with carmine. The mass flowed with comparative ease, and was very soon seen flowing and distending the superficial enlarged venous branches over the front of the thorax, at its lateral parts, and at and around the umbilicus.

Allowing time for the warm mass to harden, the examination was continued by dividing the ribs and clavicles in the line of the nipples, and separating the whole mass of the anterior abdominal muscles from the skin. Raising up this anterior mask of the thorax and abdomen, the abdominal aorta and the inferior vena cava were dissected free; the stomach, liver, and diaphragm were freed from their posterior attachments. In the neck, all the vessels and muscles, together with the trachea and œsophagus, were raised up. In the thorax the conjoined organs were cut free from the vertebræ. Thus, the whole mass of organs from the thorax and abdomen were removed *en bloc*, and their dissection was proceeded with from the posterior aspect.

In the left pleural cavity, placed superficially just beneath the membrane, was seen a large venous trunk about one-half an inch in diameter, situated on the bodies of vertebræ and extending from the arch of the aorta to the attachment of the diaphragm below. This vessel was the left azygos; it was on the left side of the aorta. In dividing the tissue of the posterior mediastinum, the veins of communication between the spinal and intercostal vessels were found unusually large, from $\frac{1}{3}$ to $\frac{1}{2}$ inch in diameter, and were thus in condition to carry a large amount of blood from the upper part of the trunk and the head downwards to be emptied into the abdominal vena cava. On removing the organs, the injecting material was found to have filled the veins to the arms and neck on each side, the spinal and intercostal vessels in the thoracic portion, numerous large veins in the dia-

phragm, converging directly towards the inferior vena cava, and also the abdominal portion of that vessel.

In dissecting the neck, numerous large veins (several one-quarter inch in diameter), were found passing behind the trachea, and they served as a communication from the right to the left side. They commenced at about the level of the episternal notch and descended into the thorax passing towards the left, and terminated in the beginning portion of the left innominate or transverse vein. The left innominate vein at this part was very much enlarged, and as the vessel advanced towards the right side to join the vena cava, it was found to be converted into a flattened fibrous cord, and this portion was pretty tightly adherent to the surrounding tissues and the branches from the arch of the aorta lying in contact with it.

A probe passed downwards within the right jugular was stopped by an obstruction at a point where the right innominate vein joined the fibrous cord in which the left innominate terminated. From the posterior wall of the right innominate, the probe entered the mouth of a moderate sized vessel which passed downwards into the thorax; its termination and communications could not be traced; it was supposed to communicate with the right azygos vein, but this vessel could not be found.

On removing the heart the pericardium was found normal. The heart was large; its weight was not taken, as a considerable portion of its auricles were left in connection with the specimen. Both the right and left ventricular walls were thicker than normal.

The mitral orifice was narrowed at its auricular aspect, but its outlet in the ventricle was of normal size. The anterior segment was rendered rigid by the deposit in it of calcareous matter. This deposit extended from near the free border, centrally through the leaflet, and terminated within one of the cusps of the aortic valve. The mitral orifice was thus not only partially obstructed, but the auricular aspect of the leaflet was roughened by the calcareous nodular deposit.

The aortic cusps were all three materially affected; one was rendered rigid through one-half its depth by the calcareous deposit extending from the mitral valve, another cusp showed a slightly projecting ridge of deposit on its ventricular aspect along the line of junction with the opposing cusp, of firmly organized fibrous material, while the third cusp was thickened, less pliable than normal, but smooth.

The other orifices and valves showed nothing especial to note.

The opening of the inferior vena cava appeared larger than normal, perhaps half as large again. The opening of the superior vena cava was found obliterated. A very fine probe could be forced in about an inch, by tearing tissues at the normal site of the cava. It was found, therefore, that the obstruction existing above in the innominate vessels was complete and extended to the auricle.

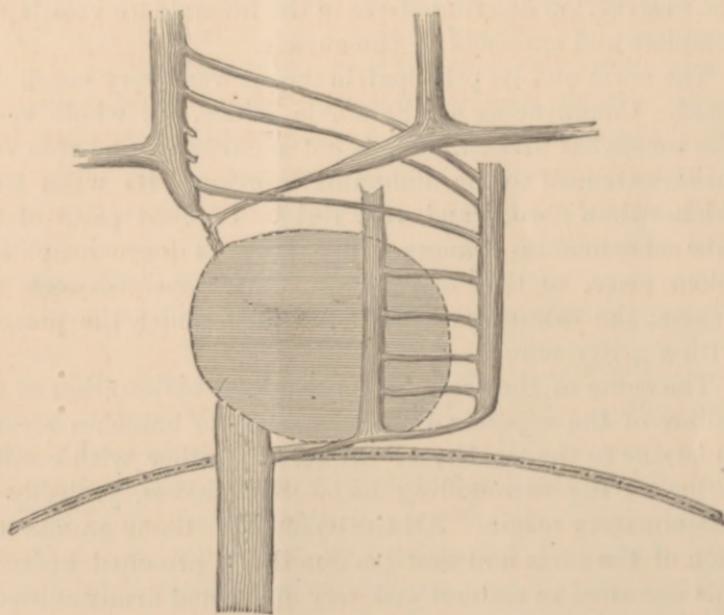
The aorta and its principal branches were very much diseased. Commencing at the aortic orifice, the whole vessel was somewhat dilated, though not aneurismal, and this condition extended to the abdominal portion. Its walls were thicker than normal and very rigid. In most parts of the tube atheromatous changes with calcareous degeneration had taken place, so that in squeezing the vessel between the fingers, the calcareous deposits cracked under the pressure with a gritty sound.

The cause of the compression and final obliteration of the calibre of the superior vena cava with its branches seemed to be due to the rigidity of the aorta together with condensation of the surrounding connective tissues, probably of inflammatory origin. All the connective tissue around the arch of the aorta and first portion of its principal branches was increased in amount and very dense, and firmly adherent to the under surface of the sternum as well as posteriorly to the bodies of the vertebræ. It was in this tissue that the obliterated venous trunks were found, and to which the

tissue adhered so firmly that it was difficult to separate the two in dissection.

The venous blood from the head, upper extremities, etc., found its way to the heart by means of the greatly enlarged veins lying on and around the spinal column posteriorly, particularly the left azygos, as well as anteriorly through the enlarged internal mammary vessels, and the superficial thoracic and abdominal branches. Most of the blood evidently came downwards to the level of the diaphragm and thence found its way directly into the inferior vena cava. Other portions of the blood descended from the superficial portions of the thorax into the abdominal vessels and thence by the lumbar branches to the abdominal vena cava.

FIG. 2.



Rough diagram showing collateral circulation.

The examination of the brain showed a notable fulness of the vessels of both the external and the internal membranes.

Small patches of opacity were found on the arachnoid in some places, along the line of the sulci over the convexity. Some of the convolutions showed in places slight degrees of atrophy, and the furrows between them were widened, and at these parts there had accumulated a small amount of clear subarachnoid fluid.

The brain substance showed no especial alterations. The vessels of the circle of Willis presented themselves as India-rubber-like tubes, patulous and rigidly elastic. No calcareous degeneration of their tunics was found. The same conditions were visible in the beginning portions of the principal branches leading from the circle.

This case was a rare one, and of more than usual interest for several reasons. In the first place, there were unusual opportunities for studying the conditions, both *ante* and *post mortem*, and the physical signs were such that the diagnosis was made during life. The enlargement of the veins of the surface led to the suspicion that one of the cavæ was occluded, and it was decided that it must be the cava superior, because when the large external veins were emptied for the space of an inch or two with two fingers, the blood evidently flowed much more rapidly and forcibly downward when the finger nearest the head was first removed than it did upward when the lower finger was first removed, and this was the case as much when the patient lay in bed as when he stood, thus showing that the current in the enlarged external veins was downwards. Further reason for thinking that the return circulation from the head found its way back to the heart partly by the external veins, was that if they were compressed there was at once produced visible congestion of the veins of the face and suffusion of the eyes and a sensation of fulness in the

head. The presystolic murmur in the heart was very marked and unmistakable, and at all times present while the patient was under observation, and abundant cause for its presence was found in the calcareous deposit with stiffening and roughness of the anterior flap of the mitral valve. The systolic murmur at the base of the heart and in the arteries was caused by the stiffening and roughness of the aortic valve flaps and of the thoracic and abdominal aorta. It was suspected during life that the arteries of the brain would be found to be stiff, and this was the case; still it is very likely that the vertigo with which the man suffered, and of which he complained more than of any other symptom, may have been partly due to the obstruction to the return circulation from the brain caused by the blood having to find its way through much smaller and more tortuous routes than the natural ones, and this view would seem to receive further confirmation from the fact that there was found some flattening and shrinking of the brain convolutions. The cause of the dulness on percussion over the upper part of the right side of the chest anteriorly and the upper part of the sternum, which was so distinctly made out, was not understood during life, but the post mortem showed abundant cause for it in the very great amount of hard fibrous tissue around the ascending aorta and veins in that region, and here apparently was the seat of origin of the whole pathological process. It seems most likely that the changes which finally caused a complete occlusion of the whole of the vena cava superior and the larger portion of both the innominate veins had its origin in a periarteritis, which must have begun

around the ascending or transverse portion of the aorta. This must have proceeded gradually, with a constant increase and condensation of the tissues around the aorta and veins, until at last there was produced the large amount of hard fibrous tissue which the post-mortem examination showed to exist in that region.

A man, twenty-six years of age, was admitted to the ward July 21, 1880. No history of previous disease was obtained, except that he had had frequent attacks of ague, and eighteen months ago had typhoid fever. One year ago he had a chancre, but it was followed by no secondary symptoms. In March (four months ago) he was upset from a boat, and after being two hours in the water was taken out unconscious. Since that time he has noticed that he has had more or less shortness of breath, but felt fairly well and was able to be about until one week ago, when his left arm became paralyzed, and the next day the leg became weak so that he walked with difficulty. Upon admission he was suffering very much from dyspnœa, and his breathing was very rapid, being forty-eight per minute. The pulse was frequent and weak. In walking he dragged the left leg, and the arm on the same side hung powerless, and was much swollen and œdematous, both feet and legs being also œdematous. The face was drawn to the right, the muscles of the left side being flaccid, but the tongue was protruded without deflection to either side. The speech was rather mumbling. The urine was normal.

July 22. A more careful examination of the heart revealed a faint mitral systolic murmur, and that the heart was hypertrophied. There were numerous coarse moist rales audible over the lungs, and there was evidence of some pleuritic effusion.

23d. Condition much the same, but he is more stupid and has frequent attacks of dyspnœa.

25th. He is unconscious, and passes his urine in bed. The heart murmur is more distinct.

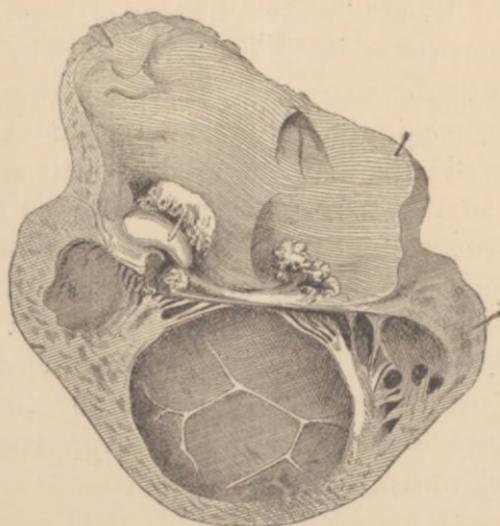
26th. Is sinking, and died July 27th.

Post-mortem examination showed the pleural sacs to contain slightly over a quart of clear serum and that the heart was much enlarged. The fluid and enlarged heart occupied so much space in the thorax that the lungs were much compressed; they felt firm to the finger, but were crepitant throughout, although they were not more than half the natural size. The heart was both dilated and hypertrophied. The right side showed nothing of interest, the valves being normal. The left ventricle and auricle were both much dilated and hypertrophied; this condition was very marked in the auricle. The mitral opening was narrowed, the valve flaps being stretched across it. When looked at from the auricle there was seen a tumor springing from the posterior flap of the valve and projecting into the auricular cavity, which was about three-quarters of an inch to an inch in diameter, and about half to three-quarters of an inch in height. This mass was made up of two different layers, the lower smooth and rounded and the upper rough and uneven, and there were numerous little shreds like rootlets hanging from its surface. There was also on the anterior flap of the valve a spot which was much roughened, and had minute vegetations attached to it. This was probably due to ulceration from friction against the tumor on the other cusp. The aorta was narrow and the valve flaps much thickened, so that the valve was incompetent. The liver was about natural in size, but the capsule was much thickened. The spleen also was natural in size, but rather firm and its capsule thickened. The kidneys appeared natural. The brain presented considerable softening of its right side; this was of the lower parietal lobule and the upper adjoining part of the temporo-sphenoidal lobe; there did not appear to be any involvement of the motor tract. Upon examination of the middle cerebral artery it was found to divide primarily into three branches. The first of these was patulous, but the two posterior ones were blocked with emboli.

Dr. Simes sent me the following report of the results of an examination of the tumor:—

“I have made a microscopical examination of the growth and find it to consist histologically of cells, fibrous tissue, and granular débris. The cells are such as are found in embryonal connective tissue, and appear to be more especially confined to the peripheral portion of the tumor. The fibrous tissue is of the ordinary kind, consisting of delicate fibrils and cells. The granular débris is apparently a metamorphosis of the cells and fibres. I was unable to see any vascular structures. These histological characters, in connection with the macroscopical appearances and clinical history of the case, would lead one to consider the new formation as syphilitic in nature—a gumma.”

FIG. 3.



The conditions presented by this patient when he came to the hospital and his history at once suggested that the cerebral symptoms were due to embolus. The distinct murmur heard at the apex of the

heart, together with the signs of hypertrophy and the general symptoms pointing toward rapidly increasing heart failure, were abundantly sufficient to warrant the belief that a fragment of vegetation had been washed from the left side of the heart into some one of the cranial bloodvessels. It struck me also that the origin of the heart disease might be syphilitic. To this suspicion I was led by the history, and because only the year before I had seen a case with a somewhat analogous condition which I reported to the College.¹ The two cases, although in some respects parallel, presented points of marked difference, both in the symptoms presented, in the histories, and in the different modes of death. In the first case there could be obtained no history of syphilis, either initial lesion or secondaries, but the man had been out of health for two years; there was, however, advanced disease of the kidney, and uræmic convulsion had occurred; the man finally died with a very slow pulse and in a certain sense rather suddenly. In the case reported to-night, the history of chancre occurring a year before death was clear, but, as in the first case, no secondary signs had appeared; the man thought himself well until about four months before his death, which was at last caused by the cerebral embolus, and he died with the usual rapid and irregular pulse of mitral disease.

¹ Transactions of the College of Physicians of Philadelphia, 3d series, vol. v., 1881.



