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EROSION OF THE SKULL IN A CASE WITH A VARIX OF THE SUPERIOR SAGITTAL SINUS¹

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G. C. T. was the inventor of the braided wire picture cord. For a number of years he was manager of a large manufacturing concern until, at the age of fifty-one, he had a paralytic stroke. This resulted in a left-sided hemiplegia, which persisted until his death nineteen years later. He was unable to continue his work, and in November, 1916, at the age of seventy, began to show some mental impairment. He was sent to this hospital, May 24, 1917, and at entrance showed marked memory defect and confusion. Physical examination showed tortuous vessels about the forehead, and a lump the size of a guinea egg, bluish in color and pulsating to palpation, at the junction of the frontal and parietal sutures. The heart sounds were regular and of good quality. The blood pressure was 110—80. There was some evidence of peripheral arteriosclerosis. There was a left-sided hemiplegia involving face and limbs. The muscles of the left arm and leg were in a state of contraction. Neurological examination showed pupils that reacted sluggishly to light and accommodation. The hearing was impaired and there was suggestion of a speech defect. The touch, pain, temperature, tactile discrimination, and stereognostic senses were impaired on the left side. The superficial reflexes were present. No clonus or Babinski. The patellar reflex was active on the right side, absent on the left. Loss of motility of facial muscles on the left side. There was a fine tremor of the extended fingers. The urine, blood ex-

¹ Contribution No. 68, Danvers State Hospital Papers. Presented for publication August 1, 1918.

aminations, and Wassermann tests were negative. For years he had suffered from severe pains in the head, left side of face, and left side of body.

The staff considered the patient's condition, postapoplectic, with some question as to whether the case showed a definite psychosis. Without any apparent illness, except general weakness, and inactivity, the patient died.

Autopsy No. 2047. Pupils are unequal, the right measuring 5 mm., the left 4 mm. The muscles of the left arm and leg are atrophic. Heart is hypertrophied. Coronaries and larger vessels show little sclerosis. There is a stone obliterating the cystic duct. A large perforation is present in the right tympanic membrane. Other organs are not remarkable. There is no lump present on the head now, but in the region of the bregma there are two irregular depressions in the calvarium that may be felt through the scalp. The thumb and index finger fit snugly into these and there is irregular exostosis of bone about them. When the thickened scalp is removed, the eroded areas are plainly seen beneath the intact periosteum. In one of these eroded areas, there is a dark cruor clot. The erosions and the findings within the brain are seen in the accompanying photographs.

Discussion.—It is a well-known fact that aneurysms of the aorta may erode ribs or even bodies of vertebræ by the pressure exerted and by the constant hammering that takes place when the force of the heart beat is transmitted to these aneurysms; but so far as we are able to find, no case has ever been reported of erosion of the skull due to an increase in intracranial pressure with a varix of any sinus. Adami (1) says that a familiar instance of bone atrophy from pressure is the depression in the bones of the calvarium due to the Pacchionian bodies. He also states that hydrocephalus and intracranial growths lead to atrophy of the calvarium, but does not cite any cases.

In the first place, we are dealing with a somewhat different condition than arterial aneurysmal erosions. It is difficult to imagine the same amount of pressure and pulsation present in a cavity like the cranium. Bergmann (2) has shown, in a woman with a defective skull, that the brain expands within its cavity synchronously with each cardiac systole. Furthermore, that it expands with expiration due to the pressure in the right side of the heart damming the blood back into the sinuses; while the contrary takes place in inspiration. The brain is unlike the kidney in the latter respect because of the absence of efficient valves in the cranial and vertebral veins, and the continuity of these veins with the right auricle and vena cava. Cramer (3) first recorded the venous pressure and



FIG. 1. Shows the inner surface of the calvarium with the dura attached by firm, thick adhesions that are torn away with difficulty. In the region of the bregma can be seen the varix in the superior sagittal sinus. This varix is 2 cm. in diameter and extends outward through the dura and internal table, to the external table, where it has caused irregular erosion of bone. It will be seen that the sinus opens directly into this, and the varix is lined throughout with the ordinary endothelial coat. When the dura is removed the primary erosion, 2.4 cm. in diameter, is found extending through the internal table into the diploë. At the anterior and posterior edges are seen diverticuli which lead off into the diploic space. Through this opening can be seen the irregularly eroded outer table. The spaces for the meningeal vessels are deeply grooved, and on the left there is a minute erosion through the inner table. In the frontal region, especially on the left side, there are irregular exostoses. There is slight erosion in the region of the internal occipital protuberance. The calvarium as a whole is thinner than usual.



FIG. 2. Shows the erosion of the external table, which is 4 cm. long and irregularly 1.5 cm. in width. There is scar-like exostosis around the edges. There are no areas of softening to be found, and no suggestion of an old cranio-tabes.



FIG. 3. Shows an opening that was made into a large degenerative cyst in the region of the first and second frontal convolutions of the right hemisphere, when the dura was torn away. The top of this cyst was adherent to the dura just beneath the varix in the superior sagittal sinus, the opening being made when the dura was torn away. The brain tissue within a radius of 4 cm. showed marked softening and a tendency to collapse when the cyst was drained. There are large varicosities in the veins over this area, especially at the tip of the frontal lobe. The veins over the convexity of both hemispheres are greatly dilated and tortuous. The pia is greatly thickened; the capillaries engorged; the sulci shallow and the convolutions indistinct. Brain atrophy is a noticeable feature, especially on the right side.

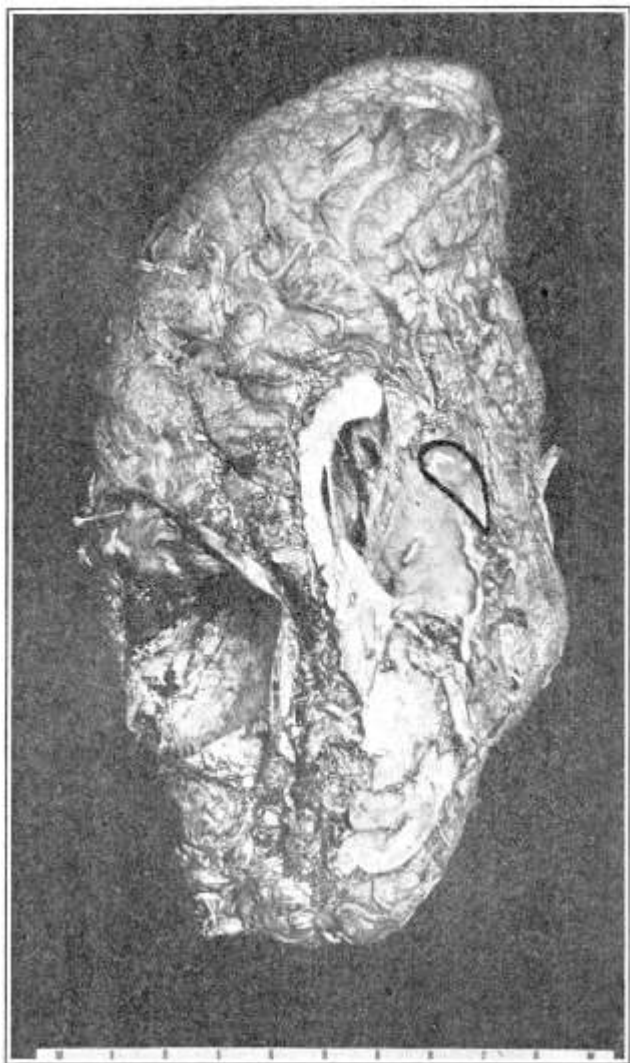


FIG. 4. Shows the right hemisphere after the cyst has been opened. The inner wall of this cyst extends to the septum made by the falx cerebri, with which it is slightly adherent. The walls are formed by a thick fibrous tissue capsule which has been cut and pinned back in order to give a better view of the cavity, which is 4 cm. in diameter. On the tip of the frontal lobe may be seen the vari-
cosities described above. The india ink line shows the outline of a degenerative cyst, the size of a small olive, in the right cerebral peduncle. This contained a small amount of straw-colored fluid.

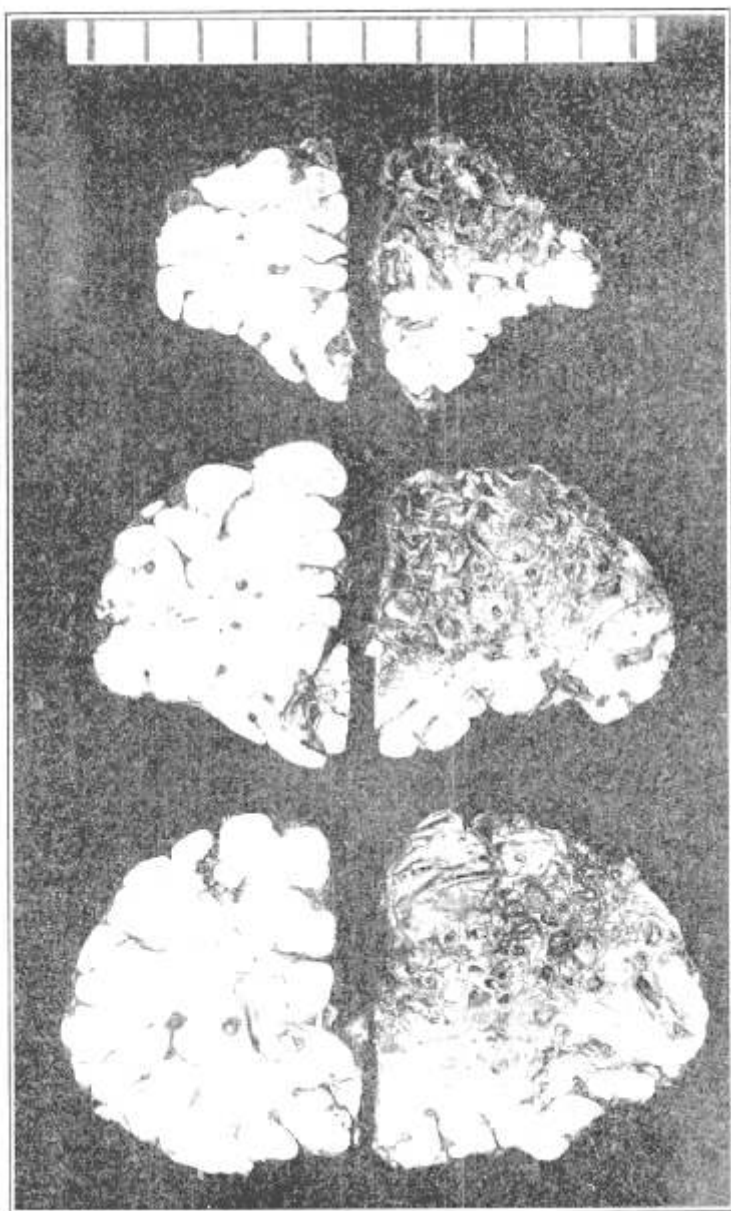


FIG. 5. Shows vertical sections through the frontal lobes, anterior to the cyst. The extensive degeneration of the right hemisphere with the brain substance replaced by a network of dilated vessels is evident.

showed the transmission of the pulse into the transverse sinus. Gaertner and Wagner (4) confirmed this observation. It would seem that the experimental work of Leonard Hill (5) had settled for all time the question of the pressure and pulsations within the cerebral sinuses. By an ingenious method he made simultaneous records of (a) the arterial pressure in the central end of the caroid; (b) the general venous pressure in the right auricle; (c) the cerebral venous pressure in the torcular Herophili; and (d) the cerebrospinal fluid pressure by trephining the atlas. This served to establish the fact that pulsations are transmitted from arteries to the cerebrospinal fluid, the cranial veins and sinuses. He also showed that the brain within its cavity may be likened to a limb or a kidney in the oncometer. The arterial stroke is transmitted to the vessels at the base of the brain and then to the cerebrospinal fluid and venous sinuses. Any increased cerebral tension was found to give proportionate increase in the pulsation within the sinus.

Now, granting that the blood pulsates within a sinus and there is a varix of this sinus due to some weakened condition of its wall, inflammatory in origin; or some congenital defect in the closure of the anterior fontanelle with consequent compensatory varicosity of the sinus to fill this defect, it would be hard to imagine pressure enough under normal conditions to erode bone.

According to different investigators the normal intracranial pressure has been recorded at various points between 50 and 260 mm. of water, which must mean that there are numerous factors affecting its normal range. Since no authorities agree on intracranial pressure, we cannot say what this pressure is normally. Now, to account for the erosion of the skull, in this case, it is necessary to assume that the intracranial pressure rose above the normal limits given. This is the greatest factor involved here, and now we shall attempt to demonstrate its cause.

In this case we do not have to look far to find the cause of the old hemiplegia and the subsequent rise in intracranial pressure. The large cyst containing dark fluid is evidently the remains of an old cerebral hemorrhage nineteen years ago. The thick fibrous capsule shows that it is of long standing, while the dark pigment on its walls, speaks for hemorrhage, rather than embolism or thrombosis. The only record that we have of the blood pressure is more in favor of thrombosis, but what the blood pressure was in his active life is now merely a conjecture.

A consideration of the anatomical arrangement and physical conditions (11) that bear upon intracranial pressure points to the

conclusion that this pressure is controlled by the venous and not the arterial pressure, which is sufficiently confirmed on the experimental side by the investigations of Bayliss and Hill (6). They have shown that a fall in arterial pressure slackens the flow of blood through the brain, while a high blood pressure accentuates it. They believe that intracranial pressure depends not upon the tension of the cerebral arteries, but follows the venous pressure.

Charcot and Bouchard (7) first showed that in many cases of cerebral hemorrhage, there are miliary aneurysms of the small atheromatous arteries in the brain substance, some one of which has burst and allowed the blood to escape. The mid-cerebral, lenticulostriate, or anterior cerebral are most commonly the ones affected, because as Mendel (8) has shown, the blood pressure is higher in these vessels than the smaller vessels of the cortex. Ford Robertson (9) states that pressure in the cerebral capillaries is always higher than in other similar vessels, and that they are provided with a special elastic coat.

Hemorrhage is much more common in men after fifty than in women or younger men, and if we may believe the old dictum of Morgagni, they occur more frequently on the right side. Hemiplegia is apt to be the most striking residual symptom, and in the late cases hemianesthesia may accompany the hemiplegia, which argues for implication of a portion of thalamic fibers. In the early stage the paralysis is flaccid, but may later show pyramidal tract, upper neuron disorder with accentuated knee jerks. This case represents a late stage or mixed syndrome, for the knee jerk was absent on that side and contracture present, suggesting an extension of the lesion into the internal capsule.

When hemorrhage occurs in the brain substance, there will be in the cranium a localized foreign body which occupies the space of a certain vascular area in which function will be lost, because the tissue with which it comes in contact will be more or less completely destroyed. Miles (10) has shown that even minor conditions causing a local pressure may cause mechanical injury to nerve cells. In consequence of the obliteration of the veins and capillaries the local cerebral tension will be raised to that of the arteries which normally feed the affected area. In the obliterated area there is stasis of blood, and in the border areas the capillaries and veins will be compressed in proportion as the increased local cerebral tension is transmitted to the brain substance. The pressure in the cerebral venous sinuses is high so long as there is compression with high cerebrospinal pressure, even though the systemic blood pressure may be

low. In consequence of this compression, the blood pressure will be raised in these capillaries and veins equal to arterial tension. Robertson (9) states that in such a condition, further expansion of the arteries and capillaries can only take place by an equivalent compression of veins, since the semifluid brain matter is incompressible. The pressure in the brain will then attempt to constrict the veins with their reservoirs of blood until the cerebral venous pressure becomes equal to the intracranial, which is now reckoned as the pressure of the brain against the veins. Now the whole circulatory system of the brain will have assimilated itself to a scheme of rigid tubes. We have in this case an example of such a process manifested by the tremendous engorgement of cerebral veins.

The pathological increase in intracranial pressure may be general or local, the local increase over the general being dependent upon the elasticity of the cerebral tissues, when pressure is produced by blood extravasation, tumors, etc. Hemorrhage into the brain occupies space by force; the surrounding tissues are compressed and stretched and their elasticity maintains a high local pressure. This would account for a primary rise in pressure, but a constant high pressure would need other factors for its maintenance. We have the first of these in the inflammatory reaction that is sure to follow such a hemorrhage. There is a dilatation of vessels, exudation, and finally a walling off of the foreign body, because the large hemorrhage cannot be absorbed. (We have a history of delirium in this case during such a period.)

We now notice that the varix in the superior sagittal sinus lies just above this old area of hemorrhage and inflammation, the dura beneath the varix forming the roof of the cyst which has formed in the hemorrhage area. Therefore it seems logical to conclude that in the inflammatory process the wall of the sinus suffered enough injury to produce this varicosity. Now we are ready to consider another factor which would help maintain a high local pressure. These chronic inflammatory states may easily alter the metabolism in the tissues of such an area, increasing those cell products, crystalloid in nature, which would raise the osmotic pressure, thus causing the tissues to swell and help in maintaining a high local pressure. Hill (5) first showed that the blood pulsates within these sinuses and then showed that the injection of foreign material into the cranium greatly increased the pulsations within the sinuses. So here we have created, by nature, a case which illustrates clinically, the splendid experimental work of that investigator, and in addition shows that in such a rare condition we may assimilate a case of

aortic aneurysm with erosion of ribs or vertebræ. The hemorrhage cyst with the extensive degeneration in adjoining tissues, the cysts in the peduncle and thalamus of that side, the engorged veins, the atrophic brain, the physical signs, the symptoms of a psychosis, and finally the erosion of the skull, all bear witness to the great increase in intracranial pressure which was maintained for years. If we are inclined to be sceptical about the production of the varix, and the primary erosion we might formulate a slight developmental error in the region of the anterior fontanelle with compensatory dilatation of the sinus, but there would be no other evidence of craniotabes to justify such a view, which, were it true, would not account for the extensive, irregular erosion and exostosis of the external table.

SUMMARY

1. We present a case that showed a pulsating tumor in the region of the bregma, about the size of a guinea egg, which had been present for nine years before his death. This disappeared after death. A post-apoplectic hemiplegia had been present for nineteen years. He had suffered from very severe headaches for many years.

2. Autopsy revealed a varix of the superior sagittal sinus with extensive erosion of the calvarium above the varix. There were marked changes in the brain substance, shown by atrophy, varicose veins, and degenerative cysts in the cerebral peduncle and thalamus.

3. The varix was probably formed by the inflammatory reaction that followed the cerebral hemorrhage nineteen years ago.

4. Experimental work has shown that the blood pulsates normally within the cerebral venous sinuses. Furthermore, it has been shown that foreign material injected into the cranial cavity increases the strength of the pulsations within the sinuses proportionately to the rise in intracranial pressure.

5. Hemorrhage into the brain occupies space by force. Following such a hemorrhage there is a marked rise in intracranial pressure due to the compression of veins and capillaries; the compression and stretching of brain tissue whose elasticity maintains a high local pressure. The inflammatory and metabolic changes also help in maintaining this pressure.

6. We believe that the pathological findings in the brain prove that a high intracranial pressure has been present for many years, and that the erosion of the calvarium was due to this pressure, both local and general, together with the varix in the superior sagittal sinus.

7. This case would seem to illustrate clinically the experimental work of Leonard Hill and others. It may be taken as an exaggerated example of erosions that are found in the calvarium due to the pressure of Pacchionian granules, or even assimilate a case of aortic aneurysm with erosion of ribs or vertebræ.

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