BASILAR IMPRESSION (PLATYBASIA)

A Bizarre Developmental Anomaly of the Occipital Bone and Upper Cervical Spine with Striking and Misleading Neurologic Manifestations*

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Basilar impression, sometimes spoken of as platybasia, is a deformity of the occipital bone and upper end of the cervical spine resulting from anomalous development. It may produce marked pressure effects upon the medulla and adjacent parts of the central nervous system. These pressure effects may simulate syringomyelia, disseminated sclerosis, or progressive spastic paralysis so closely as to be indistinguishable from them. It is, therefore, of paramount importance that every practicing neurologist and radiologist be thoroughly familiar with the clinical manifestations and roentgen findings, especially as surgical reconstruction of the foramen magnum and upper spinal canal may prove of marked benefit to the patient (*vide infra*, Case I).

In spite of the fact that the anatomic changes are bizarre and, once they are appreciated, striking, the condition is easily overlooked. Our own experience suggests that typical examples could be gleaned from the current x-ray film files of many large medical centers, by reviewing all cases of syringomyelia, multiple sclerosis, and progressive spastic paralysis.

The morphologic changes shown by roentgenograms give the impression of softening of the base of the skull and moulding through the force of gravity. It is as though the weight of the head has caused the ears to approach the shoulders, while the cervical spine, refusing to be shortened, has pushed the floor of the posterior fossa upward into the brain space. The resultant changes in relationships and shapes of such parts as the clivus, petrous portions of temporal bones, and foramen magnum are shown by the roentgenograms (Figs. 1 and 2) and drawing (Fig. 3).

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Varying degrees of fusion or assimilation of atlas into occiput are met with, as might be expected from a consideration of the embryology of these parts. In some cases the atlas has lost its identity completely. In others it is fused with the margin of the foramen magnum on one side or the other, or the ventral part of the vertebra is fused with the ventral margin of the foramen while the neural arch is defective.

Clinically these morphologic changes are less apparent than upon the roentgenograms, but the shortness of the neck may be noticeable or even marked, the movements of the head upon the neck are limited and the head is often carried at a peculiar angle.

The effects of the deformity upon the nervous system may be slight or profound. There is a marked tendency for these nervous manifestations to be progressive. In a certain number of cases they ultimately become the cause of death in the third, fourth, or fifth decade, though the patient may not have exhibited any nervous manifestations during the first few years of life. This has led a number of observers to conclude that the deformity itself is increasing, and must therefore be on an osteomalacic or rachitic basis rather than on the basis of a developmental anomaly. Dr. Richard U. Light has called my attention to the possibility of a better explanation, namely, that when skeletal growth takes its characteristic and normal spurt in the years just preceding puberty, the spinal cord has already attained its normal adult length. The increase in the length of the spine may produce a certain amount of traction on the cord and in cases of "basilar impression" deleterious degrees of pressure of the odontoid process against the medulla may begin at this period of skeletal growth.

The American literature is singularly lacking in reference to "Basilar Impression" though a few references have been made to a somewhat related type of anomalous development, at a lower level in the cervical spine, the so-called Klippel-Feil syndrome.[3, 5, 11]

The morphologic changes have been covered with characteristic thoroughness in the Old World literature and there are many good case reports of nervous manifestations with clinical neurologic diagnoses of "syringomyelia," etc. Ebenius[1] cities Rokitansky as mentioning the deformity in 1844, but Virchow[12] was the first to submit a more detailed description of the skull changes (1876). In 1880, Grawitz[2] contributed careful anatomic descriptions of six skulls with basilar impression. Virchow and Grawitz held that while anomalous

development was etiologic in some cases, others were caused by rickets, syphilis, etc.

Because of his careful neurologic observations, correlated with post-mortem study, the article by Homén[4] (1901) is important. He noted local pressure effects of the odontoid process upon the medulla, a striking feature of at least two of our cases (*vide infra*).

Schüller[10] reported the first case diagnosed during life (by roentgen examination, of course). He thoroughly understood the effects of the deformity upon the nervous system and presented clinical examples of (1) paralysis and irritation of cranial nerves (vagus, hypoglossal, etc.); (2) compression of cerebellum against the tentorium, with even a block of the aqueduct of Sylvius; (3) local pressure effects upon the medulla by the odontoid process and the stenotic foramen magnum.

A few words should be said about the technic of the roentgen examination. The conventional lateral projection is diagnostic, especially if one pays close attention to the relationships of the clivus, atlas, and odontoid process of the axis. By drawing a "base line" from the dorsal lip of the foramen magnum to the dorsal margin of the hard palate any measurable degree of basilar impression becomes apparent (see normal comparison, Fig. 1). It is well to see that the lateral projections include all of the cervical vertebrae because of the frequency with which fusions of other segments lower down in the spine are encountered.

The lateral projections do not give any comprehensive outline of the foramen magnum. The usual "occipital projections" (central ray of x-ray beam directed in mid-sagittal plane of head, from "hair line" to nape of neck) may be difficult or impossible on account of the short neck and limitation of movements of the head upon the neck. We have, therefore, developed a technic which might be termed a fronto-vertex projection. Essentially the reverse of the old "occipital projection," the central ray of the roentgenographic beam projects the shadow of the dorsal rim of the foramen magnum through the middle of the frontal bone, in the neighborhood of the hair line, or even higher, in the region of the coronal suture (Figs. 2 and 4). This view has the theoretical disadvantage of placing the desired detail (foramen magnum) at a great distance from the roentgen film. But in most roentgen departments, the size of the x-ray tube focal spot has now become so small as to render this factor of negligible importance, and we have found it desirable to substitute this

"fronto-vertex" projection for the older "occipital projection" in a great many of our skull examinations, because of the comprehensive outlines of the occipital bone and foramen magnum which it gives us.

Case Reports

Case I. E. F., male, 20 years, college student, admitted March 24, 1937. *Chief Complaint*: Weakness in legs; loss of sensation in certain skin areas.

History of Present Illness: Five years ago, when the patient was 15 years old, he was carrying a canoe on his head, supported by a tonk strap, when a slight obstacle in the portage trail forced a quick jerk of his neck muscles to readjust the weight of the canoe. Following this episode he noted slight stiffness in his neck. Others noticed that he began to carry his head tilted slightly toward the right. Osteopathic treatments, designed to rectify the torticollis, failed to do so, but seemed to increase the range of joint motion. No further symptoms developed until February, 1936, when he exhibited unexplained tremor of both hands. In March he noted frequency of urination and some difficulty in controlling his sphincters. In April he noted spontaneous muscular reactions of the Babinski type in the left foot; these disappeared after about 10 days of medication with sedatives. Shortly after this he discovered, by scalding his right shoulder under a hot shower, that certain skin areas were anesthetic. At about this time he engaged in spring football practice and was thrown violently to the ground, on one particular occasion, alighting on his buttocks. This produced tingling sensations in both lower extremities and in the right arm and shoulder, with a sensation of coldness in the right hand, and transitory weakness in the legs. He was able to walk, but felt "top heavy." There were no sphincteric disturbances. After 15 minutes of rest the tingling and weakness disappeared. In May, about 4 weeks after the football injury, he developed a carbuncle on the right shoulder, with pain referred to the back and into the neck and with twitching of muscles under his right shoulder blade. At this time he noted spontaneous muscular reactions of the Babinski type in both feet, and his back "felt terrifically sore." He could still walk, but his legs felt stiff. Weakness developed in the right arm and hand, becoming progressively worse until, on May 26, 1936, he could not hold the ordinary utensils for eating. Diagnoses of syringomyelia and intramedullary tumor were considered at that time, but following lumbar puncture the neurologic manifestations all improved. In July, 1937, lipiodol was introduced into the cisterna magna and passed without delay into the caudal sac. (About 30 hours after the introduction of the lipiodol, he experienced a violent diffuse pain reaction, burning in character, lasting a few days.)

Neurologic Examination: On March 25, 1937, the positive findings included the following: Bilateral hyperactivity of patellar and Achilles

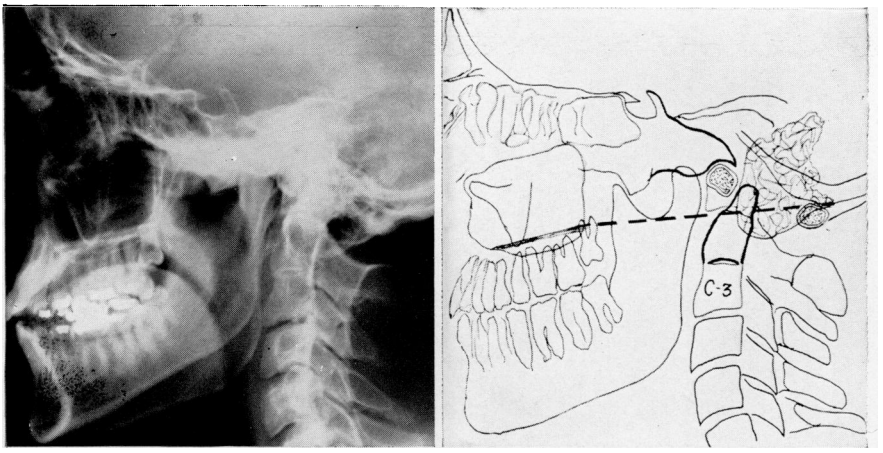

FIG. 1 a-b. *Left*: Right lateral projection of upper cervical spine and base of skull in a typical case of platybasia (Case I). *Right*: Tracing from same. Note the cephalic bulge at the clivus, the displacement of odontoid process of axis and most of the atlas to a level cephalad of the indicated base line (from hard palate to dorsal margin of foramen magnum). Anomalous fusion of bodies of C2 and C3 is also shown.

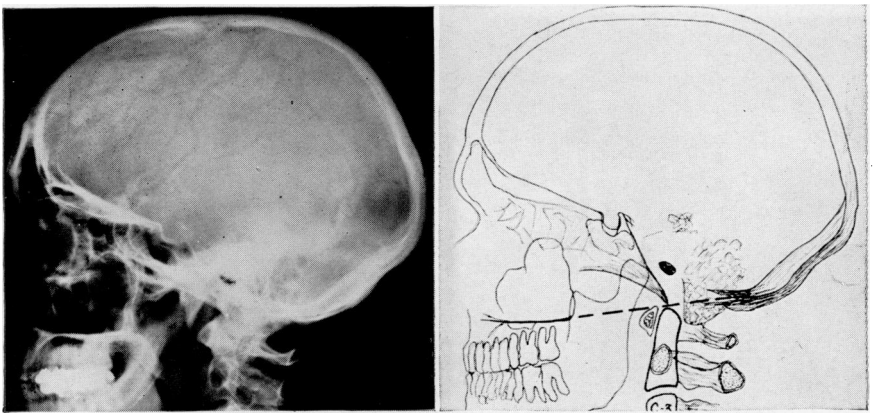

FIG. 1 c-d. *Left*: Lateral roentgenogram of normal, for comparison. *Right*: Tracing from same. Note that all parts of atlas and axis lie caudad of the "base line."

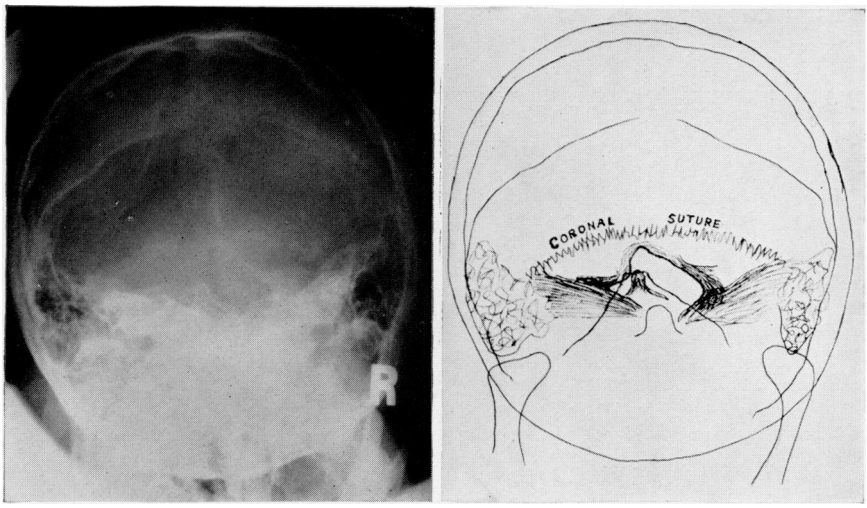

FIG. 2 a-b. *Left*: Fronto-vertex projection of skull in platybasia (Case I). Central ray of x-ray beam enters skull at dorsal margin of foramen magnum and projects the shadow of this part upon the film through the frontal bone near the coronal suture. *Right*: Tracing from same. Note the deformity and stenosis of the foramen magnum. This patient's symptomatology and neurologic findings were characteristic of syringomyelia.

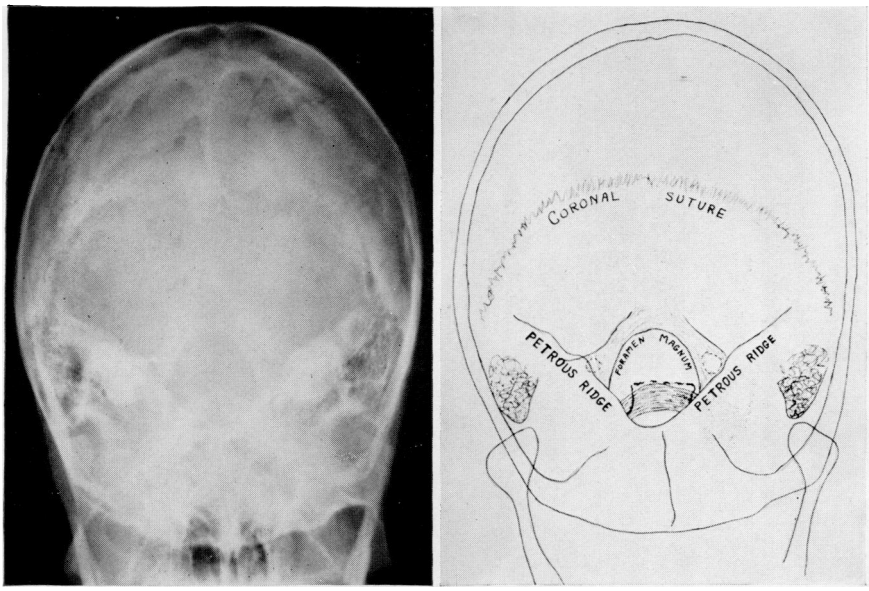

FIG. 2 c-d. *Left*: Fronto-vertex projection of the skull in a normal patient. *Right*: Tracing from same. Even though the ventral inclination is not as marked in this study as in the one above (shadow of coronal suture higher on film and shadow of dorsal margin of foramen magnum lower), the foramen magnum is better visualized than in the case of platybasia. Note the dorsum sellae and dorsal part of atlas, framed in the foramen magnum.

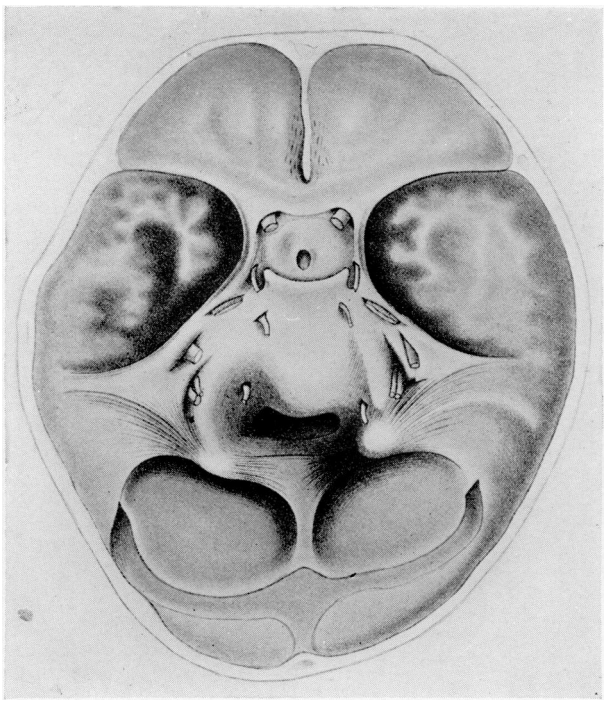

FIG. 3. Drawing (from Homén) of interior of skull after removal of brain in a case of "basilar impression." The dura has not been removed. The cephalic bulge of the clivus and the stenosis of the foramen magnum are very apparent and typical.

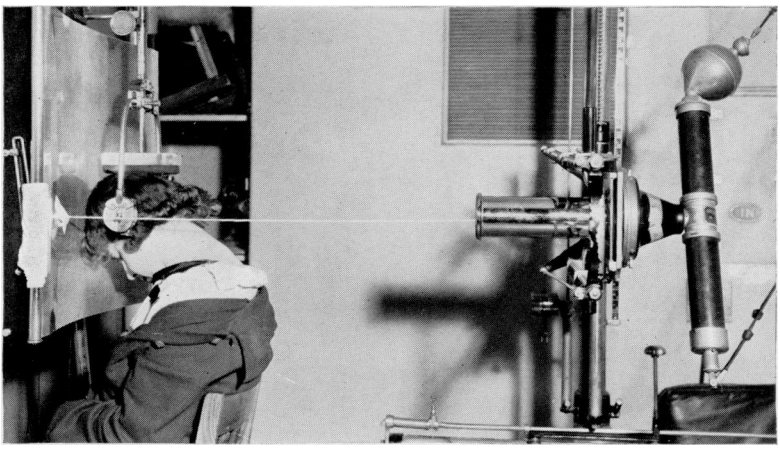

FIG. 4. Photograph to show the arrangement of patient, x-ray tube, and Potter-Bucky diaphragm for the "fronto-vertex projection" (compare Fig. 2). The arrangement of the apparatus and the positioning of the patient are far easier than in the case of the opposite direction of projection, the so-called "occipital projection." Since this technic is designed with particular reference to delineation of the occipital bone and since this part is at a distance from the film, the x-ray tube focal spot must be of the smallest and the target-film distance must be ample. We have found this type of roentgen examination very informative.

reflexes with ankle clonus and positive Babinski; slight motor impairment right triceps and deltoid; slight atrophy of right thigh with motor impairment, especially in flexors; tactile sensation not impaired but bilateral hyperesthetic zone for pain and temperature at junction of 2nd cervical dermatomeres and 3rd divisions of trigeminals; similar hyperesthetic zones for pain and temperature at 6th thoracic dermatomere on the left and 7th thoracic on the right; pain sense completely absent from 8th to 10th thoracic with the "smooth skin" of a "vasomotor level"; pain sense present but decreased below 10th thoracic, more on the left; tingling sensations over lateral aspects of both upper extremities when head is flexed to the limit.

On June 25, 1937 (second admission), the progressive nature of the involvement of the nervous system was further emphasized by the following additional findings. Loss of control of bowel and bladder sphincters (present for one month); weakness of lower extremities worse than before; abdominal reflexes absent (present but unequal in March); marked increase of sensory impairment in dermatomeres C2, C3 on the right, C2 on the left.

Lumbar Puncture: Findings negative, including Queckenstedt.

Laboratory Findings: Essentially negative.

X-ray Findings: Typical deformities of basilar impression (see Figs. 1 and 2).

Operative Interference: On June 30, 1937, Dr. Temple Fay performed a low occipital craniectomy and laminectomy of cervical segments down to and including the 3rd. The dura was opened to permit inspection of the cord. There was no syringomyelia, tumor, or other intramedullary lesion. The compressive effects of the skeletal deformity were readily demonstrated, as was also the release afforded by the laminectomy and craniectomy. Its partial assimilation into the occipital bone rendered the atlas very difficult to find, especially as its neural arch was very incomplete.

Follow-up note, March, 1939: There has not been a complete return to normal, neurologically, but clinical improvement was measurable from the first postoperative day and has continued. The patient, now 22 years old, has entered medical school and is apparently under no significant handicap.

Case II. Mrs. L. T., 28 years, housewife, first admitted on December 6, 1937. *Chief Complaint*: Weakness of legs; uncertain gait; severe "drawing pain" in abdomen.

History of Present Illness: The patient was apparently well until August, 1936, when she noticed suddenly that her left leg dragged and at times very unexpectedly "gave way." About 2 months later she noticed that upon arising from a sitting position she would stagger. Shortly thereafter the severe pain in her abdomen, of a "drawing" character, became an important feature of her condition. The abdominal pain was in no way related to meals or types of food. It never occurred at night; in fact, it tended to be

relieved fairly promptly by lying down. Three years prior to the onset of her present illness, the patient had fallen on the ice, striking her head, but had developed no symptoms at the time, and gave the injury no further thought until questioned about injuries after her deformity was demonstrated by roentgenograms.

Neurologic Examination: The positive findings included the following. Nystagmus in all directions. Absent corneal reflexes. Hypalgesia in ophthalmic and maxillary divisions of 5th cranial nerve on right, less marked in mandibular division. Mild involvement of right 7th, of peripheral type. Cerebellar dyssynergia in upper and lower extremities, more on right side. Positive Romberg. Motor impairment in left upper and lower extremities. Bilateral increase of biceps and triceps reflexes with positive Hoffman, more on the left. Bilateral increase of patellar and Achilles reflexes, with ankle clonus on right. Left big toe in position of constant Babinski; mild Babinski on right. Abdominal reflexes absent on both sides. Mild impairment of muscle position sense of toes on left. Hypalgesia over entire left half of body below C6, including marked loss of temperature sense.

Lumbar Puncture: Queckenstedt positive. Air myelography revealed incomplete block at site of spinal and occipital deformity.

Laboratory work, including Serology: Essentially negative.

X-ray Findings: Characteristic deformities of basilar impression, misinterpreted as entirely traumatic until the post-mortem examination (Feb. 5, 1939) revealed the underlying anomaly.

Operative Interference: Low occipital craniectomy and laminectomy in upper cervical spine down to and including C3, operation by Dr. Temple Fay, Jan. 10, 1938. The dura was not opened.

Outcome: Patient improved markedly after the operation and remained well until October, 1938, when her symptoms returned and became progressively worse. She was re-admitted to the hospital on Jan. 20, 1939, and died on Feb. 4. At post-mortem examination the true nature of the deformity, a typical "basilar impression," was recognized for the first time, and it was obvious that the reason the laminectomy and low occipital craniectomy had failed to afford permanent relief was the continued presence of a constricting and compressive mass of calcified and greatly thickened dura. (As stated above, the dura was not opened at the time of the operation.) A profound degree of softening and pressure atrophy of the medulla oblongata was a striking feature of the post-mortem findings, and upon removal of the brain the cause of these compressive effects was obvious. The region of the clivus, normally concave, was seen to bulge upward into the posterior fossa in a typical fashion. The odontoid process added to the local pressure mechanism. The foramen magnum was markedly stenosed. The whole appearance was remarkably like that shown in Homén's drawing (Fig. 4).

Case III. Mrs. M. S., 43 years, housewife, first admitted on Dec. 13, 1938. *Chief Complaint*: Right hemiplegia of a few days' duration, jargon aphasia, vomiting of recent onset.

Past History: Unimportant as obtained from patient.

Neurologic Study: Entirely consistent with the final diagnosis,—syphilitic thrombosis of middle cerebral artery; C.N.S. syphilis.

Laboratory Findings: Blood serology strongly positive for syphilis. Spinal fluid negative.

Roentgen Findings: There is a perfectly definite and obvious basilar impression with stenosis of the foramen magnum, but no clinical neurologic manifestations of same. (We plan to repeat the neurologic studies after the patient's syphilis is under control.)

Case IV. H. H., male, 31 years, electrician, admitted Nov. 9, 1936. *Chief Complaint*: Loss of strength in both arms, more marked on the right; weakness in both legs; sensory disturbance, left hand.

History of Present Illness: The patient was perfectly well until 6 years ago when he noticed weakness of the left arm and loss of ability to distinguish heat and cold with the left hand. About a year later he developed weakness of both legs. About 4 years ago he began to develop the same type of weakness in the right arm. Muscular atrophy began to be noticeable about 4 years ago and has been progressive. He feels that his shoulder muscles have disappeared. For the past 6 months he has noticed that he sweats only on the left side of the face, left arm, and left side of trunk.

Neurologic Study: The positive findings included the following. Suggestion of ptosis, left; left pupil larger than right; marked atrophy of all shoulder girdle muscles, less marked in arms and forearms, slight in lower extremities; biceps and triceps reflexes absent on left, biceps absent on right; patellar and Achilles reflexes hyperactive, bilaterally, with abortive clonus; sense of position and vibratory sense unaffected and tactile sensation generally unimpaired; pain sense absent from C2 to T2 and diminished down to T9 on the right, present on the left but markedly diminished from C2 to C6 inclusive.

Physical Examination: The essential points are covered under the heading of "Neurologic Study," except that there was an apparent shortness of the neck, with a tendency for the head to be flexed and turned to the left. The patient has been aware of this deformity since it was called to his attention at the time of a physical examination 15 years ago.

Laboratory Findings: Essentially negative, including serology and spinal fluid.

Spinal Puncture: Negative, except that the Queckenstedt procedure indicated a partial block.

Ophthalmoscopic Findings: Anomalous arrangement of retinal veins.

X-ray Findings: At the time of the x-ray examinations in November, 1936, the significance of the occipital and upper cervical deformity was not appreciated. Two years later, stimulated by our increased understanding of Cases I and II, and aided by the exceptional memory of Dr. Samuel B. Hadden, who remembered that there was something unusual about the atlanto-occipital relationship, the films were reviewed and the diagnosis of basilar impression thoroughly established.

Outcome: The patient died before the diagnosis of basilar impression was reached. The patient was not under our observation at the time of his death, and no post mortem was obtained.

Comment

Anatomic studies are convincing to the effect that "basilar impression" is basically a developmental anomaly. Since indications of osteitis, rickets, osteomalacia, etc. are entirely absent, the striking tendency for the patient to escape neurologic effects during the first decade or so of his life, and then to develop a markedly progressive symptomatology, must be accounted for on some other basis. We offer the thought that young developing brain and brain stem may be able to tolerate compressive effects which later prove deleterious to older tissues. In our autopsied case (Case II) and in many other cases that have been thoroughly studied, the flattening, softening, and atrophy of the medulla at the point of impingement of the abnormally located odontoid process, have been striking. Yet we have reason to believe that the odontoid impinged in more or less the same degree during earlier years, before the onset of recognizable neurologic symptoms and signs.

It would hardly be possible to overemphasize the importance of the tendency for basilar impression to masquerade as syringomyelia, disseminated sclerosis, etc. A substantial majority of the case reports in the literature contain references to such diagnoses in patients who ultimately turned out to have this condition. Our Case I presented clinical neurologic manifestations so characteristic of syringomyelia that the proposal to do a laminectomy and reconstruction (enlargement) of the foramen magnum met with strenuous opposition from those neurologists and neurosurgeons who habitually discount roentgen evidence unless clinical evidence points in the same direction. The prompt arrest of the rapidly downhill progress of this man's condition, with sufficient actual restoration of function so that he is no longer significantly handicapped, can only mean that the skeletal

deformity was of primary etiologic importance. Furthermore, the dura was opened at the operation and thorough inspection of the cord failed to reveal any suggestion of syringomyelia or other intramedullary lesion.

In the interpretation of roentgen findings of basilar impression it must be remembered that the skeletal deformity may exist without neurologic manifestations, or may at least antedate such manifestations.

When a case of basilar impression is operated, it is essential that the exploration be wide and that the dura be opened. Only in this way can the surgeon be sure that his reconstruction is giving medulla, cerebellum, and cord sufficient leeway so that local pressure effects will be entirely obviated. (For theoretical reasons we believe that section of the tentorium may prove necessary in an occasional case in order that release from pressure-effects may be complete.) Our Case II emphasizes the importance of opening the dura, for we now know from our post-mortem findings that this patient could have been relieved by removal of the thickened dura which continued to exert local pressure after the laminectomy and low occipital craniectomy.

Summary

"Basilar impression" is an unusual but important developmental anomaly of the occipital bone and upper cervical vertebrae. Its diagnosis during life rests upon recognition of the characteristic stenosis of the foramen magnum and cephalic bulging of the clivus and neighboring bony structures into the posterior fossa of the cranium. Its chief importance lies in its effects, sometimes profound, upon the central nervous system, especially as these effects may be obviated by appropriate surgical procedures.

The neurologic manifestations of basilar impression are most often such as to simulate syringomyelia, though disseminated sclerosis and various types of spastic paralysis may also be imitated, according to the particular tracts, columns, or other parts of the central nervous system that are effectively impinged upon or otherwise disturbed by the skeletal deformity.

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