BASILAR IMPRESSION (PLATYBASIA)
A CASE SECONDARY TO ADVANCED PAGET'S DISEASE
WITH SEVERE NEUROLOGICAL MANIFESTATIONS.
SUCCESSFUL SURGICAL RESULT

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Basilar impression had been known to the German pathologists for
some time. Rokitansky\(^\text{11}\) apparently was the first to mention the de-
formity in 1844. Boogaard,\(^\text{1}\) in 1865, described the condition and re-
lated its causes and consequences. Virchow\(^\text{14}\) submitted more detailed de-
scriptions of the condition in 1876. Grawitz,\(^\text{2}\) in 1880, made careful ana-
tomical studies of six skulls with basilar impression. Homé,\(^\text{5}\) in 1901, was
the first to correlate the neurological abnormalities with the postmortem
findings. He demonstrated the local pressure effects of the odontoid process
on the medulla oblongata.

It was not until 1911 that the condition was recognized as a clinical
entity. At this time Schüller\(^\text{12}\) made the first premortem diagnosis, describing
the effects of the bony deformity on the central nervous system. Since that
time, clinico-pathologic reports have appeared by Sinz,\(^\text{13}\) Kecht,\(^\text{7}\) Krause,\(^\text{8}\)
Juhlin-Dannfelt,\(^\text{6}\) Merio and Risak,\(^\text{10}\) and Ebenius.\(^\text{3}\)

In 1939, Chamberlain\(^\text{2}\) described four cases of basilar impression with
bizarre neurological abnormalities. In two cases, suboccipital decompression,
carried out by Dr. Temple Fay, resulted in improvement. Chamberlain’s
report is important in that it stimulated a growing interest in this condition
in this country. The excellent report by List\(^\text{9}\) on the neurological changes
associated with developmental anomalies of the occipital bone, atlas and
axis, further elucidated the problem.

From the standpoint of etiology basilar impression may be “primary”
(result of a congenital, developmental anomaly) or “secondary” (result of
bony softening and moulding). Trauma may be an aggravating feature in
any case but is believed never to be the underlying cause of the disease.
Rickets, osteomalacia, hyperparathyroidism and osteitis deformans (Paget’s
disease) have been listed as causes, in many instances without convincing
proof. Walsh, Camp and Craig,\(^\text{15}\) in reporting a successful surgical result,
list Paget’s disease as a cause of secondary basilar impression but do not cite
specific examples.

A case is here reported, for the first time, of basilar impression secondary
to Paget’s disease producing severe neurological manifestations. The patient
was treated successfully by high cervical laminectomy combined with suboc-
cipital decompression.
CASE REPORT

Mr. R. K., aged 50, was admitted to the Neurosurgical Service of Temple University Hospital, January 11, 1944. He complained of severe bilateral occipital neuralgia, bilateral loss of hearing, weakness of the right arm and leg with dragging of the latter extremity, numbness of the right forearm and foreleg, difficulty in speech with regurgitation of fluids through the nose, and wobbling gait.

He stated that he was entirely well until five years before admission when his brother-in-law suddenly pulled his head in a tussling match. He experienced sudden pain at the back of his head which radiated to the vertex. At first this pain would come in paroxysms but for the last year it had been almost continuous. Loss of hearing had been bilateral and progressive, existing for a number of years, the right ear being involved more than the left. Approximately one year before admission there was onset of progressive weakness of the right arm and leg so that, on admission, he had to drag the right leg in attempting to walk. Simultaneously there was noted a feeling of numbness, starting in the region of the right elbow and progressing to the fingers, and also starting in the region of the right knee and progressing to the toes. He likewise complained of numbness of the four fingers of the left hand. The right foot always felt cold. During this year his gait had become quite unsteady. For the last few weeks his speech had been thickened and indistinct and he noted, on occasions, that fluids would regurgitate through the nose on attempted swallowing. Vision was impaired in the right eye, which he attributed to an ocular injury sustained many years ago. During the last four years the head had steadily increased in size. He had been accustomed to wearing a hat sized 6½; on admission it was 8½. There had been a 33-lb. loss in weight in the last few months.

At the age of 11 he fell from a tree and fractured his coccyx. Herniorrhaphy was performed in 1929.

Examination. The general appearance in the advanced stage of Paget's disease was striking. The short, squat figure with bent shoulders, sunken chest, curved back, and great head hanging forward, wobbling along, was a true living justification of Sir James Paget's description of the disease in 1876. The great flattened head seemed to "mushroom" on top of the cervical spine. The veins and arteries of the temporal areas were unduly prominent. The movements of the head were limited in all directions but particularly in the lateral deviations to the shoulders. Palpation showed marked atrophy of the posterior cervical group of muscles. The spino processes appeared thickened and prominent. The pupils were equal, but light reaction was sluggish in the right eye due to the presence of a corneal scar. The light reflex in the left eye was normal. Vision was impaired in the right eye only. The visual fields were grossly normal. The fundi appeared normal. The external ocular movements were normal in all fields of movement but there was a horizontal nystagmus of second degree on either lateral gaze. The corneal reflexes were normal and there did not appear to be any sensory abnormalities in any of the trigeminal fields. The facial expressional movements were equal. There was bilateral eighth nerve deafness, more pronounced on the right side. The gag reflex was present on both sides and the uvula and soft palate moved in the midline. The left shoulder was slightly lower than the right. The tongue protruded in the midline without tremor or atrophy. The speech, nevertheless, was quite thickened and almost indistinct. There was difficulty in swallowing fluids and, on occasion, the fluid would regurgitate through the nose.

The gait was markedly ataxic with falling to either side in the Romberg position. In attempting to walk, he would drag his right lower limb. The deep tendon reflexes were bilaterally exaggerated, but more so on the right side. There was a Hoffmann sign on the right side with bilateral Babinski responses, more easily elicited on the right side. There was marked weakness in motor power of the right upper and lower limbs. The motor weakness was mild on the left side. There was a bilateral dysmetria and dyskinesia of the upper limbs, more pronounced on the right side. Ataxia was pronounced in both lower limbs, but also was more exaggerated on the right side.

Sensory disturbances to pain and temperature were not distinct. However, he was able to appreciate an ice tube more acutely on the entire left side with a definite increase in sensa-
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tion bilaterally at the level of the second cervical dermatomere. Vibration, position and stereognostic sense were unimpaired.

Laboratory Studies. The urinalysis and complete blood counts were normal. The spinal fluid and blood serology were normal. The blood calcium was 10.2 mgm. per 100 cc.; the blood phosphorus was 4.5 mgm. per 100 cc. while the alkaline phosphatase was elevated to 36 Bodansky units per 100 cc.

Roentgenograms. I am indebted to Dr. W. Edward Chamberlain for the following comments and descriptions of the roentgenologic findings in this case.

“Advanced Paget’s disease of the skull and upper cervical spine is here visualized (Fig.

![Image of skull showing advanced Paget's disease with secondary platybasia and basilar impression](image)

Fig. 1. Right lateral projection of skull showing advanced Paget’s disease with secondary platybasia (marked flattening of the “basal angle” between floor of anterior fossa and clivus) and basilar impression (invagination of occipital condyles, margins of foramen magnum and other basal structures into the brain space). In Paget’s disease the bones become softened, even when their mineral content is high. The deformities here visualized are a result of that softening.

1). This is characterized by thickening of the bone with loss of demarcation of the outer table of the skull. Many small distinct and conglomerate areas of increased bone density are seen. These are characteristic of the disease.

“Paget’s disease produces marked softening of the bone, even in the presence of increased bone density (increased calcium content). As a result of this softening a marked degree of secondary basilar impression has developed. The cephalo-caudal dimension of the cranium is at least 2 cm. less than the normal. The angle between the floor of the anterior fossa and the clivus, normally of the order of 135 degrees, has flattened to approximately 160 degrees. The basilar invagination is most marked at the ventral margin of the foramen magnum, less marked at its dorsal margin. As a result of this, the plane of the foramen magnum is inclined dorsad and obviously there must be a kinking of the brain stem where it passes around the invaginated odontoid process and the ventral margin of the foramen magnum.
Normally the plane of the foramen magnum is so disposed that this “high angle vertex projection” fails to visualize it. In basilar impression the invagination is more marked at the ventral margin of foramen than at the dorsal margin. Its plane thus becomes angulated so that it may be visualized by this angle of projection. When viewed in the stereoscope the outlines of the foramen can be readily distinguished. In this non-stereoscopic illustration its outlines may be confused with the more apparent somewhat concentric outlines of the upper vertebral canal.

Air introduced by lumbar puncture for encephalomyelography visualized an almost complete block, with the caudal limit of the cisterna magna and one of the cerebellar tonsils displaced down the vertebral canal as far as the upper borders of the laminae of C-3 (Fig. 3). Some air did enter the cranial cavity, indicating that the block was incomplete or inconstant, but even after prolonged maintenance of the sitting position, most of the air remained below the level of the foramen magnum.

Encephalomyelogram. Jan. 14, 1944. At the above-mentioned encephalomyelographic procedure the initial spinal fluid pressure was normal. The fluid was clear and all laboratory findings were nega-
tive. A partial block was indicated by manometric readings during jugular compression.

Operation. Jan. 18, 1944. Suboccipital craniectomy with high cervical laminectomy (Dr. H. Wycis). Under anertin-local anesthesia, the usual midline incision was made with reflection of the cervical and occipital muscles. The angle between the occiput and the upper cervical spine was so acute that it was very difficult to free the atlas and axis. The rim of the atlas on the right side was fused with the posterior rim of the foramen magnum. Of striking interest was the fact that the second cervical spinous process was extremely huge and porous. The bone had the consistency of cheese and was extremely vascular. It appeared so soft that the laminectomy could have been completed with a stout pair of scissors. The laminae of the first five cervical vertebrae were removed. The bone over the cerebellar hemispheres was then rongeured away. The dura was thickened and deeply indented over the cord in the region of the atlas. The dura was opened over the upper cervical segments and over both cerebellar hemispheres. It was noted that the tonsils of the cerebellum were herniated through the foramen magnum for a distance of approximately one inch with greater displacement on the right side than on the left. There was a suggestive Chiari deformity due to a kinking of the posterior part of the medulla against the upper cervical cord. It appeared as if the entire cervical cord and medulla were foreshortened because of the deformity. The second cervical roots were swung in a hammock fashion about the lower poles of the tonsils. The second cervical roots were crushed on both sides. The dura was allowed to remain open and the muscle layers were closed with interrupted steel wire. Skin was closed with silk.

Postoperative Course. The postoperative course was little short of dramatic. Eight hours after operation he was alert and conscious. He stated that he had no occipital pain and that the numbness and tingling were absent from his hands and feet. Two weeks after operation he felt nearly normal. The pain had entirely subsided. He could walk without dragging the leg and the ataxia had almost completely abated. He stated that hearing had improved in the left ear. Speech was now distinct and there was a marked improvement in swallowing. Nystagmus was still present on either lateral gaze. The deep tendon reflexes were still more active on the right side with bilateral Babinski signs. Six weeks later, he presented few, if any, neurological abnormalities except for an equivocal Babinski on the right side. He was free of all pain, and had a substantial increase in appetite and gain in weight. There was an area of anesthesia over the distribution of the second cervical dermatomere due to the crushing of the second cervical posterior roots. Hearing, however, was apparently unchanged. The tortuosity of the temporal vessels had markedly decreased (Fig. 4).

Microscopic Examination. The bone removed shows the characteristic mosaic pattern of Paget's disease (Fig. 5).

COMMENT

A successful surgical result in a case of basilar impression secondary to Paget's disease is presented. The writer feels that wherever possible the
dura should be allowed to remain open so as to permit full decompression. List\textsuperscript{9} likewise recommends opening the dura for adequate decompression. In the successfully operated case by Dr. Temple Fay (Chamberlain, Case I\textsuperscript{1}) the dura was allowed to remain open.

The probability of other cases of basilar impression secondary to Paget's disease certainly exists. Patients with Paget's disease should have thorough neurological examinations, as well as thorough X-ray studies of the upper cervical region and foramen magnum area. Since the writing of this report, the author has encountered two other cases of Paget's disease with neurological dysfunction due to compression by the diseased bone. In one case there was early bilateral loss of hearing, tingling in both hands and fingers, and unsteady gait. The X-rays of the skull show typical platybasia. The second patient presented himself with progressive loss of vision in the left eye due to compression of the optic nerve in the optic canal. Both patients refused operation.

REFERENCES

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11. ROKITANSKY, C. Cited by Ebenius.


