Syringomyelia as a complication of Paget’s disease

Case report

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Cranial settling and basilar invagination with medullospinal compression is believed to have resulted in the
production of a syrinx in a case of Paget’s disease of the cranium. This mechanism of compression at the
craniovertebral junction resembles the development of syringomyelia in Chiari malformations presenting in
adult life.

Key Words • Paget’s disease • basilar invagination • syringomyelia • magnetic resonance imaging

Paget’s disease of the bone is found in 7% of men
and 4% of women over the age of 55 years. Neurological syndromes associated with this disease are relatively rare. The neurological sequelae of Paget’s disease relate largely to involvement of the spine and vary according to the vertebral level, the degree of vascular compromise, and the presence or absence of malignant change. Involvement of the craniovertebral junction can result in basilar invagination with medullospinal compression, as described by Wycis. The coexistence of medullospinal compression and syringomyelia has been commonly mentioned in relation to Chiari malformations, particularly those presenting in adulthood. The occurrence of syringomyelia with Paget’s disease has rarely been mentioned, however, and no individual case report can be found in which this association has been described. We present such a case, and give details of the radiographic appearance and the operative treatment.

Case Report

This 64-year-old woman had suffered from Paget’s disease for more than 10 years. At this admission, she presented with a 3-year history of falling episodes particularly after bending forward, progressive right-sided weakness with an inability to raise the arm and dragging of the leg, ataxic gait which had worsened in the previous 6 months, and dizziness on rapid head movements. She had long-standing pain in the neck and interscapular region, and more recently she noted pain in the right arm. She denied having headaches, diplopia, dysphagia, or a sensory deficit.

Examination. The head circumference was 64 cm, with frontal bossing and asymmetrical temporoparietal enlargements. The head was held with a left tilt. The patient had a relatively immobile cervical spine and an upper thoracic scoliosis convex to the right. She exhibited jerky ocular pursuit with gaze-paretic horizontal nystagmus, and a few pulses of upbeat nystagmus were noted on upward gaze. Her hearing was decreased bilaterally and she had atrophy of the right side of the tongue. She exhibited a spastic quadriparesis (worse on the right side and involving the arm more than the leg), gait ataxia, and dysdiadochokinesia. Bilateral pyramidal signs with lower cranial neuropathies and nystagmus with pursuit difficulty were sufficient to implicate brainstem compression in the pathogenesis of this condition.

Advanced Paget’s disease of the cranial vault with flattening of the exo-occiput and basilar invagination along with a tendency toward platybasia were evident on plain films of the skull. Moderately severe degenerative disease affected the midcervical region. The C2-5 vertebral bodies were involved by osteoblastic activity secondary to Paget’s disease. Computerized tomography revealed invagination of C-1 within the posterior fossa with compression of the brain stem and cerebral structures and nonvisualization of the fourth ventricle. Magnetic resonance imaging (MRI) of the head and cervical and thoracic regions of the spine at 0.5 Tesla, using the spin-echo technique, identified a
markedly thickened cranial vault of a high signal intensity containing many vascular channels (Fig. 1). The ventricles were not enlarged relative to the cerebral volume. Cavitation of the cervical cord began at the level of the foramen magnum and extended to the lower thoracic levels.

Deformation of the skull with basilar invagination secondary to Paget’s disease was believed to have caused narrowing of the foramen magnum and compression of the medullocervical junction, with progressive interference of the free flow of cerebrospinal fluid (CSF) between the cranial and spinal compartments. Central cord cavitation immediately below the level of compression was likely to have developed as a direct outcome of this pathological sequence of mechanical events. A decompressive procedure was planned to stabilize this patient’s neurological deterioration.

Operation. A suboccipital craniectomy and bilateral laminectomy of the posterior arch of C-1 was carried out with a subpial resection and plication of the cerebellar tonsils, exploration of the fourth ventricle, and muscle plugging of the entrance to the central canal at the obex. A right-sided hemilaminectomy of the C-2 posterior arch was then performed and a syringo-subarachnoid shunt of the syrinx implanted. An augmentation duraplasty was performed with Lyodura prior to closure. The posterior arch of C-1 had a normal appearance, but the right lamina of C-2 seemed to be slightly involved by the disease process and had become fused to the right lamina of C-3 which was more markedly involved. Many more small vessels were encountered in the epidural space than are usually appreciated. The foramen magnum appeared to be narrow and the dura beneath it contained a tight band posteriorly which constricted the space even more. Both cerebellar tonsils were situated at the level of the foramen magnum and were squeezed together within the constraints of the narrow foramen. The upper cervical cord was obviously cystic, and the syrinx was most superficial at the root entry zone at the C-2 level. A clear cystic fluid was aspirated (protein 10 mg/dl). The C-2 nerve root exited with a slightly upward course, indicating some displacement of neural tissue relative to its vertebral encaement.

Pathological Examination. All sections of bone from the occiput and vertebral levels showed evidence of Paget’s disease with very fragmented haversian systems. Bone trabeculae were narrow and the marrow spaces were fibrotic. Very marked osteoblastic and osteoclastic activity was present throughout.

Discussion

Paget’s disease commonly involves the base of the skull, resulting in a tendency to flatten the foramen magnum and to diminish its anteroposterior diameter. Basilar invagination develops as the odontoid process begins to project through the foramen, further compressing the cervicomedullary junction and leading to a variety of neurological complications related to involvement of the lower cranial nerves, medulla, pons, and cerebellum. This bone deformation appears in approximately one-third of patients with Paget’s disease. Impairment of the CSF circulation by compression of the medullospinal junction, coupled on occasion with a leptomeningeal overgrowth at the foramina of Magendie and Luschka, has resulted in hydrocephalus with dementia in patients with Paget’s disease.
Syringomyelia in Paget's disease

The association of syringomyelia with basilar invagination secondary to Paget's disease, although correctly surmised to exist by Schmidek, has not previously been described in a case report. Neurological manifestations in cases of basilar invagination due to Paget's disease are rare. Basilar invagination can be asymptomatic or can present with clinical features identical to those seen in patients with Paget's disease without radiological evidence of basilar invagination. The local medullary and cranial nerve symptomatology in our patient suggested a direct compressive etiology, particularly in light of her spastic quadripareisis. It is interesting that segmental sensory dissociation, which would suggest the presence of syringomyelia, was neither evident from the clinical history nor found on physical examination. In a spine affected by Paget's disease it would be difficult to differentiate a cervicobrachial amyotrophy due to central cord cavitation from that due to spondylosis and degenerative disease. The same might be said of a thoracic kyphoscoliosis, another manifestation of an intramedullary cervical cord lesion of large vertical dimension. With the advent of MRI, the anatomy and pathology of the medullospinal junction can be assessed in distinct detail with the ventricular and syringoidal cavities seen in continuity in the sagittal plane in selected cases.

Gardner, et al., first drew attention to the frequent association of the Chiari malformation and syringomyelia. Similar reports have followed, indicating this coexistence in over 50% of cases of Chiari malformations, particularly during adolescence or the adult years. Although the basic mechanical pathogenesis of medullospinal compression in the Chiari malformation and that found in basilar invagination associated with Paget's disease are different in that one appears to be the reverse of the other, the tendency toward development of syringomyelia as a consequence of this compression must be similar. Dynamic factors such as straining, coughing, or postural changes are likely to be involved in the production of central cord cavitation in the presence of medullospinal compression. These maneuvers modify intracranial and intraspinal pressures and can produce large pressure gradients between poorly communicating intracranial and intraspinal compartments and thus allow a preferential diversion of flow into the central canal of the cervical cord. The low protein content of the syrinx fluid in our patient would argue in favor of a direct communication with the ventricular spaces.

Operative treatment of syringomyelia can be recommended for patients who are deteriorating. Clearly in our case the surgical goal entailed foramen magnum decompression with drainage of the fourth ventricle and syrinx cavity. A similar approach has been recommended by others. Plugging of the central canal has been controversial; however, the procedure has not led to any long-term ill effects and may be a significant factor in contributing to subsequent cord and syrinx collapse.

References


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