Syringomyelia Associated With Paget Disease of the Skull
—Case Report—

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Abstract

A 38-year-old man presented with a rare case of syringomyelia associated with Paget disease of the skull. Syringosubarachnoid (SS) shunting was performed. We speculate that deformation of the skull secondary to Paget disease caused narrowing of the foramen magnum with progressive impairment of the cerebrospinal fluid circulation, which led to syringomyelia and neurological symptoms. SS shunting is safe, effective, and technically simple, so may be a useful initial treatment for syringomyelia associated with Paget disease of the skull.

Key words: syringomyelia, Paget disease, syringosubarachnoid shunt, foramen magnum decompression, cerebrospinal fluid circulation

Introduction

Paget disease is a bone disease of unknown etiology affecting approximately 3% of the population over the age of 40 years.8,15) Paget disease is chronically progressive, manifesting as localized disorder of bone thickening and deformity caused by excessive turnover of bone remodeling. The skull, involved in 65% of cases, pelvis, vertebrae, and femur are common locations, in which multiple regions are often involved. Paget disease is usually asymptomatic with only 5% of patients developing symptoms.14) Paget disease involving the base of the skull results in a tendency to flatten the foramen magnum and to diminish its anteroposterior diameter.6,17) This bone deformation appears in approximately one-third of patients with Paget disease.5) Obstruction of the foramen magnum leads to various neurological complications. Basilar invagination occurs in one third of patients with long-standing Paget disease of the calvaria, and may cause hydrocephalus and brainstem symptoms.5,13,19) The coexistence of foramen magnum and syringomyelia obstruction has been commonly mentioned in relation to Chiari malformations, trauma, spinal tumor, and arachnoiditis.6) The occurrence of syringomyelia with Paget disease has rarely been mentioned.6,15) We report a case of syringomyelia associated with Paget disease of the skull which was treated with syringosubarachnoid (SS) shunting.

Case Report

A 38-year-old male had suffered from Paget disease for more than 25 years. He developed difficulty in hearing at age 16 years due to stenosis of the external acoustic meatus caused by proliferation of the skull. He underwent plasty of the external acoustic meatus at age 31 years, but the operation was discontinued due to heavy bleeding from the bone. He had severe forearm pain at age 34 years and was referred to our department.

On admission, his head circumference was 69.5 cm, with asymmetrical temporal and occipital enlargements. Physical examination revealed severe right forearm pain. No obvious motor or sensory disturbance was detected. Cranial radiography demonstrated abnormal bone formation (Fig. 1A, B). Bone computed tomography demonstrated extensive calvarial thickening, abnormal bone formation, and narrowing of the foramen magnum (Fig. 1C–E). Cranial magnetic resonance (MR) imaging demonstrated marked thickening of the skull with heterogeneous signal intensities, with narrowing of the foramen magnum and posterior fossa (Fig. 2). Cervical and thoracic MR imaging demonstrated a syrinx extending from C6 to T2 (Fig. 3A, B). Cine MR imaging demonstrated absence of cerebrospinal fluid (CSF) signal change at the craniocervical junction (Fig. 4). We thought that the forearm pain was derived from syringomyelia. The pain did not improve and gradually developed in the left forearm. As a result, surgical treatment was planned to stabilize the patient’s neurological deterioration.

SS shunting was selected. Hemilaminectomy (C7, T1) and dorsal root entry zone myelotomy (C8) were performed. One end of the shunt tube was inserted into the syrinx and the other end was placed in the ventrolateral subarachnoid space. The postoperative course was uneventful. The bilateral forearm pain improved markedly.
Cervical and thoracic MR imaging demonstrated reduction in the size of the syrinx (Fig. 3C, D).

Discussion

The term “syringomyelia” is used to describe fluid-filled cavities within the neuroparenchyma usually lined by glial cells. “Hydromyelia” is used for dilation of the central canal and the presence of ependymal cells lining the canal. “Syringohydromyelia” is a global term that has been proposed since hydromyelic cavities often include a contiguous pocket of neuroparenchyma lined by glial cells, and syrinxes within neuroparenchyma often rupture into the central canal. Histology is generally required for a definitive diagnosis. Syringomyelia and hydromyelia are difficult to differentiate based on clinical findings and di-
Syringomyelia Associated With Paget Disease

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