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Magnetic Resonance Imaging in Paget Disease of the Skull

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Four cases of Paget disease of the skull were studied with magnetic resonance (MR) imaging. With optimal use of projection and technical factors, MR permits simultaneous demonstration of osseous deformity and its relation to the underlying soft tissues. Effects on the brain caused by thickened calvaria and brainstem compression from basilar impression can be detected readily on a single sagittal scan.

Paget disease of bone is a multifocal disorder characterized by increased and disorderly resorption and formation of bone, resulting in enlargement and deformity of osseous structures [1, 2]. As the disease progresses, extensive deformity may develop. The bones of the skull, which are often affected by Paget disease, are close to the brain. Deformity by Paget disease may therefore give rise to neurologic symptoms [3, 4]. In this communication, we report the value of magnetic resonance (MR) imaging as a possible diagnostic tool to assess the effects of osseous changes of Paget disease of the skull on the brain.

Subjects and Methods

One normal volunteer (fig. 1) and four patients, all women, with histologically confirmed Paget disease with skull involvement were examined by MR. Although there was evidence of Paget disease in all cases, all patients had received (3-amino-1-hydroxypropylidene)-1,1-bisphosphonate (APD) and were in biochemical remission [5]. The examinations were performed on a Philips 0.15 T MR system with 2D Fourier transformation. Each signal acquisition was averaged twice. In all four patients and in the volunteer, a single sagittal slice was selected using a matrix of 256×256 , with a slice thickness of 1.5 cm. All images were acquired with spin-echo (SE) technique. The repetition times (TRs) and the echo delay (TE) used were 500 and 1000 msec TRs and 50 msec TE.

Case Reports

Case 1

A 62-year-old woman with involvement of the skull and cervical spine by Paget was examined by plain films and MR imaging. A decompression procedure had been performed earlier to alleviate acute brainstem symptoms caused by severe basilar impression. The patient then received APD therapy and was in complete remission. Figure 2A shows a grossly thickened and enlarged calvaria with multiple osteolytic and osteosclerotic areas diffusely scattered throughout the skull. Basilar impression is seen.

On MR imaging (fig. 2B), the osseous changes of Paget disease appeared as irregular broadening of the diploic space and thickening of the inner and outer tables as high- and low-intensity zones, respectively. In addition, the vertical and horizontal diameter of the cranial cavity were reduced. The upward protrusion of the foramen magnum and surrounding

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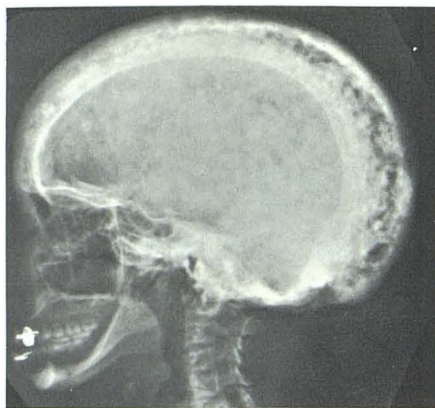
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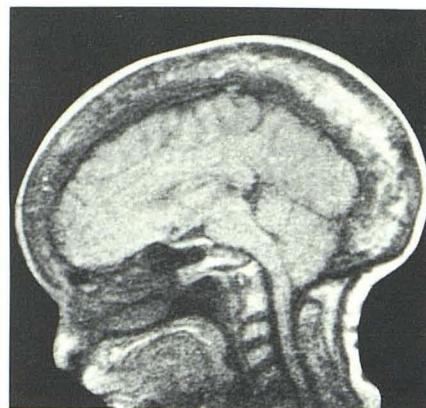
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Fig. 1.—Sagittal SE image, 1000 msec TR, 50 msec TE, of normal skull. Diploic space is thin and has high signal intensity. Tables are represented by low-intensity lines on both sides of diploë. Cerebrospinal fluid cannot be differentiated from inner table with this pulse sequence.



A



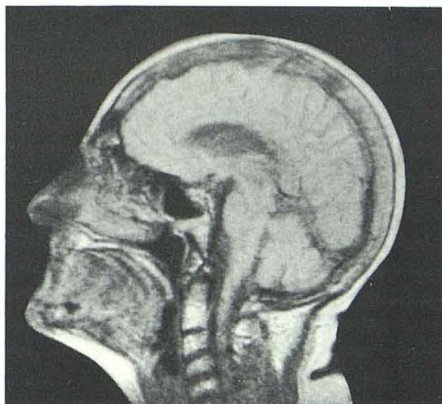
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Fig. 2.—Case 1. A, Lateral film of skull. Severe involvement by Paget disease with thickening of calvaria and marked basilar impression. B, Sagittal SE image, 500 msec TR, 50 msec TE. Widening of diploic space and irregular thickening of tables. Decrease in intracranial space with consequent flattening of brain. Compression of brainstem caused by severe basilar impression.



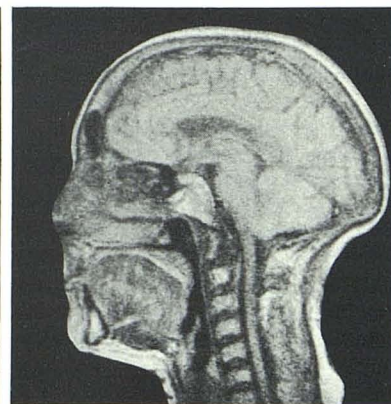
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Fig. 3.—Case 2. Sagittal SE image, 500 msec TR, 50 msec TE. Thickening of skull is predominantly affecting diploic space. Flattening of brain secondary to deformed calvaria. Severe basilar impression with compression and displacement of brainstem and moderate dilatation of ventricles.



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Fig. 4.—Case 3. Sagittal SE image, 1000 msec TR, 50 msec TE. Thickening of calvaria is predominantly localized in diploic space, with corresponding thinning of inner and outer tables. No involvement of base of skull. Compression



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on frontal lobe by wavy margins of inner table.

Fig. 5.—Case 4. Sagittal SE image, 1000 msec TR, 50 msec TE. Only moderate changes in cranial vault, with uneven enlargement of diploic spaces and minimal dilatation of lateral ventricles. Thickening in bone marrow space of clivus; however, normal anatomic relations of brain in posterior fossa persist. Diploic space is slightly enlarged, with irregular thinning of both tables.

bone, which resulted from severe basilar impression, was well demonstrated. The altered anatomy of the posterior fossa and the resulting deformity of the brainstem could be seen on a single image.

Case 2

A 60-year-old woman with Paget disease of 18 years' duration continued to have a dull headache, deafness, depression, and loss of memory despite complete biochemical remission on APD therapy. Plain radiographs showed basilar impression and other classical changes of Paget disease of the skull. A single sagittal MR image (fig. 3) clearly showed brain deformity resulting from reduced vertical diameter. Upward protrusion of the foramen magnum and slight hydrocephalus were also noticeable.

Case 3

A 74-year-old woman with a long history of Paget disease was in complete remission after treatment with APD. She was clinically asymptomatic. A conventional radiographic examination showed predominantly sclerotic bones with only a slight impression of the base of the skull.

MR (fig. 4) demonstrated that the enlargement of the cranial vault was primarily caused by an increase in the volume of the diploic bone, shown as a high-signal-intensity zone. The contour defects along the cortex secondary to the bony outline of the inner table were appreciated on the MR image. There was evidence of basilar impression. The posterior fossa did not show any abnormality. Slightly dilated ventricles may be either age-related or from focal compression with atrophy of brain.

Case 4

A 77-year-old woman known to have Paget disease for 15 years had originally been admitted with neurologic symptoms that included deafness, headache, and pain in the back and legs thought to be from the osseous changes of the disease. Treatment with APD had resulted in a complete biochemical remission of the disease activity and no symptoms related to Paget disease remained, with the exception of deafness. A recent skull radiograph showed osteolytic and osteoblastic bone lesions.

MR confirmed the moderate involvement of the skull with normal anatomic relations in the posterior fossa (fig. 5). There was some irregular thinning of the inner and outer tables, which have a low signal intensity. Broadening of the diploic spaces, seen as a high-intensity signal, accounted for enlargement of the cranial vault. No significant signs of compression were seen. The minimal ventricular dilatation may have been age-related.

Discussion

The most common radiologic finding in Paget disease of the skull is the thickening of the calvarial bones. Clinical status may vary considerably. The patient may be symptom-free throughout life despite extensive skeletal disease. Headaches and symptoms arising from compression of cranial nerves and basilar impression are well documented [6, 7]. Association of Paget disease and altered mentality, dementia, and sensory abnormalities have been suggested, without good supportive evidence [1, 2]. Basilar impression, with or without neurologic symptoms, occurs in about one-third of patients with Paget disease of skull [3, 4].

MR offers a new dimension in studying this disease, both the bony and the soft-tissue changes. It provides easily recognizable demarcation of bony structures and shows that the thickening is largely localized to the diploic space, which on MR is seen as a high-signal-intensity zone [8]. The high MR signal of the normal diploë (fig. 1) results from the short T1 and long T2 relaxation times of the fatty component of the marrow and the slow-flowing, almost stagnant blood channels of the diploë [8]. The diploë in Paget disease has a high fibrotic, connective tissue component [9] that should give a relatively low signal intensity on the MR image. In practice, however, it gives a high-intensity signal. This may be from the high hydrogen content in the connective tissue. The low MR signal with sharp margins, corresponding to the inner and outer tables of normal skull (fig. 1), is from low mobile proton density and an extremely short T2 relaxation time of the compact bones. In contrast, in Paget disease, the inner and

outer tables are shown to have an irregular contour (figs. 2B and 3-5). We studied our patients using only the sagittal view. For evaluating cranial nerves, additional views may prove to be useful.

Our study shows that MR has an advantage over conventional radiography in showing soft tissue in addition to bony changes. Its advantage over CT is in the ease of obtaining sagittal views even in a difficult patient. The spatial relation of the bones of skull to the intracranial structures, and the basilar impression and its effect on the brainstem, can be appreciated at once by looking at a single sagittal MR image. With readily available medical therapy and need for timely recognition of any correctable structural abnormality, MR imaging appears to be a useful adjunct to the management of Paget disease.

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