Multiple Giant Cell Tumors and Paget Disease: CT and MR Findings

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We report computed tomography (CT) and magnetic resonance (MR) findings of a patient with polyostotic Paget disease and multicentric giant cell tumor (GCT). Brain CT scan showed widening of diploic space, cortical thickening and enhancing soft tissue mass in occiput with underlying calvarial destruction. Ill-defined soft tissue masses were also detected in maxillary sinus and buttock with underlying bony destruction on CT. MR image showed multifocal nodules in wide diploic space with low signal intensity on T1-weighted image and bright signal intensity on T2-weighted image. Mass in occiput showed homogeneous hypointensity to bone marrow on T1-weighted image and homogeneous isointensity on T2-weighted image. Multiple nodules in diploic space and occipital mass showed contrast enhancement following administration of Gd-DTPA. Biopsy was performed at scalp, maxillary sinus and buttock, and histologic analysis revealed GCT.

Index Words: Bone neoplasms, CT Bone neoplasms, MR Osteitis deformans

Paget disease is a condition of unknown cause affecting approximately 3 percent of the population over the age of 40 years. It appears to be particularly common in inhabitants of Australia, Great Britain, and certain areas of continental Europe, but it is extremely rare in the Oriental (1). Neoplastic complication of Paget disease is relatively rare, with osteosarcoma being the most common histologic type and giant cell tumor (GCT) representing a considerably smaller minority of the cases (1, 2). We report computed tomography (CT) and magnetic resonance (MR) imaging findings of multicentric GCT in a patient with Paget disease of bone.

CASE REPORT

A 53-year-old man with a family history of Paget disease presented with a mass in occiput. His father was known to have Paget disease.

Brain MR imaging revealed widening of diploic space and a mass in occiput with underlying calvarial destruction. On T1-weighted (TR/TE, 500/20) spin echo image, multiple nodules with low signal intensity (SI) were dispersed in diploic space. On T2-weighted (TR/TE, 2000/80) spin echo image, nodules appeared bright. The mass in occiput showed low SI to marrow on T1-weighted image and isointensity on T2-weighted image (Fig 1a and b). Multiple nodules in diploic space and occipital mass showed heterogeneous contrast enhancement following administration of Gd-DTPA (Fig. 1c). Excisional biopsy of the mass and histologic analysis revealed GCT.

One year later, he revisited for masses on previous operation site and buttock. Brain CT scan showed a relatively well-marginated enhancing soft tissue mass on the previous operation site with bone destruction (Fig. 2a and b). Pelvis CT scan showed right iliac bone destruction with an ill-defined enhancing mass in buttock and lower back (Fig. 3). Open biopsy of the masses revealed GCT. Three months later, the patient developed nasal obstruction and epistaxis. CT scans of the paranasal sinus revealed bone destruction of medial and lateral walls of the right maxillary sinus with a relatively well-marginated homogeneous soft tissue mass in the maxillary sinus extending
Fig. 1. Paget disease with GCT in calvarium
a. T1-weighted SE sagittal image shows widening of diploic space with multifocal low SI nodules (arrows) and a homogeneously low SI mass in occiput with underlying calvarial destruction.
b. T2-weighted SE axial image shows bright SI nodules (arrows) in diploic space and the homogeneously low SI mass in occiput.
c. Contrast-enhanced sagittal image shows enhancement of the nodules in diploic space and the occipital mass.

Fig. 2. Pelvis CT shows an ill-defined soft tissue mass involving gluteus muscles and lower back with iliac bone destruction.

Fig. 3. CT of the paranasal sinus shows a homogeneous soft tissue mass in the right maxillary sinus with sinus wall destruction to the nasal cavity and infratemporal fossa (Fig. 4). Biopsy yielded GCT.

Fig. 4. The microscopic section of the occipital mass (H & E, × 300) shows the even distribution of multinucleated giant cells within stromal cells.

dcessive rate of bone turnover, and combination of osseous resorption and apposition. Therefore multiple findings may coexist in the marrow. In the active phase of the disease, the marrow is replaced by fibrous connective tissue with large, numerous vascular channels. This fibrovascular marrow resembles granulation tissue, therefore MR images show decreased SI on T1-weighted image and high SI on T2-weighted image (3-5). High signal areas on T1-weighted image is considered to be residual islands of fatty marrow, subacute hemorrhage, or dilated vascular channels with slow flow velocity (3, 4). In the inactive osteo-sclerotic phase of the disease, the marrow reverts to a more normal state, with loss of excessive vascularity and of fibrous connective tissue. The mosaic pattern of bone remains. Our case shows multifocal small hypo-intense nodules interspersed in hyperintense diploic space on T1-weighted image. It may represent fibrous connective tissues in residual fatty marrow. Contrast enhancement occurs due to hypervascularity of fibrous connective tissue as suggested in a case of Ginsberg et al (4).

DISCUSSION

Paget disease is dynamic and variable as there is excessive rate of bone turnover, and combination of osseous resorption and apposition. Therefore multiple findings may coexist in the marrow. In the active phase of the disease, the marrow is replaced by fibrous connective tissue with large, numerous vascular channels. This fibrovascular marrow resembles granulation tissue, therefore MR images show decreased SI on T1-weighted image and high SI on T2-weighted image (3-5). High signal areas on T1-weighted image is considered to be residual islands of fatty marrow, subacute hemorrhage, or dilated vascular channels with slow flow velocity (3, 4). In the inactive osteo-sclerotic phase of the disease, the marrow reverts to a more normal state, with loss of excessive vascularity and of fibrous connective tissue. The mosaic pattern of bone remains. Our case shows multifocal small hypo-intense nodules interspersed in hyperintense diploic space on T1-weighted image. It may represent fibrous connective tissues in residual fatty marrow. Contrast enhancement occurs due to hypervascularity of fibrous connective tissue as suggested in a case of Ginsberg et al (4).
Patients with Paget disease of bone are 20 times more likely to develop primary malignant bone tumors than the rest of the population of comparable age. The most common tumor is osteogenic sarcoma, followed by fibrosarcoma and chondrosarcoma. GCT of bone is an unusual complication of Paget disease (6). The tumor typically develops in patients with polyostotic Paget disease. Both the anatomic location and age distribution of these patients are considerably different from those of patients with conventional GCT (6, 7). Patients with Paget disease and GCT are much older (average age, 60 years) than those with conventional GCT. The skeletal distribution favors the skull, facial bones, and axial skeleton rather than the extremities. The radiographic appearance of pagetic GCT has some features of classic GCT. A predominantly lytic lesion that lacks both a sclerotic rim and periosteal reaction has been found in pagetic GCT. An extensive soft tissue component was noted in our case as in the cases by Potter et al (2). Most GCT demonstrate low to intermediate SI on T1-weighted image and intermediate to high SI on T2-weighted image. Tissue inhomogeneity has been noted on T2-weighted image (7), but our case showed homogeneous pattern on T2-weighted image. It may be relevant to tissue homogeneity, such as homogeneous giant cell infiltration without hemorrhage or necrosis (Fig. 5). Contrast enhancement following administration of Gd-DTPA is due to highly vascular nature of GCT (6).

Although CT and MR imagings are effective in the detection of GCT, these are not adequate to differentiate lytic areas in Paget disease including osteolytic phase of Paget disease, metastatic disease, giant cell reparative granuloma and other sarcomatous degeneration (1, 2). Therefore biopsy should be considered for the correct diagnosis.

REFERENCES


Paget병에서 발생한 다발성 거대세포종: CT와 MR 소견

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다골성 Paget병에서 발생한 다발성 거대세포종의 CT와 MR소견을 보고한다. CT상 두개골의 판간층 (diploic space)이 넓아져 있고 피질골의 비후와 함께 후두부에 조영증강이 되는 연부조직 종괴가 관찰되었으며 주위 골파괴가 동반되어 있었다. 또한 상악동과 턱부에도 경계가 불분명한 연부조직 종괴와 함께 주위 골파괴가 관찰되었다. MR에서는 넓어진 판간층 내에 많은 결절들이 있었으며, 이들은 T1 강조영상에서 저신호강도로, T2강조영상에서는 고신호강도로 나타났다. 후두부의 종괴는 군질하게 T1 강조영상에서 저신호강도로, T2 강조영상에서는 높은신호강도로 나타났다. 판간층의 결절과 후두부의 종괴는 Gd-DTPA 주입후 군질한 조영증강을 보였다. 각각의 종괴를 조영증강한 결과 모두 거대세포종으로 확진되었다.
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venue: Indiana Convention Center
contact: 영동세브란스병원 방사선과 오기근 교수 (tel: 02–3450–3511)

1995/10/18—21 22nd Annual Refresher Course of the International Skeletal Society
venue: The Westin Canal Place Hotel, New Orleans, Louisiana, USA
contact: Ryals and Associates, P.O. Box 1925, Roswell, GA 30077–1925, USA

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venue: Wuerzburg, Germany.
contact: Deutsche Roentagenges e.V,
Postfach 1204, D-63232 Neu-Isenburg, Germany.
(tel: 49–6102–4032; fax: 49–6102–6668)

1995/11/26—01 81st Meeting Radiological Society of North America (RSNA)
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(tel: 1–708–5712670; fax: 1–708–5717837)

1996/03/02—07 21st Annual Meeting Soc. of Cardiovascular and interventional Radiology
venue: Seattle Conv. Center Seattle, WA, USA.
contact: Soc. Cardio. Interv. Radiol., Technical Exh. Services,
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venue: The Gleneagles Hotel Perthshire, Scotland, United Kingdom.
contact: Mrs. T. Seear, The London Clinic,
20 Devonshire Place, London WIN 2DH, Uniter Kingdom.

1996/03/17—20 Annual Meeting American Institute of Ultrasound in Medicine
venue: New York, USA.
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11200 Rockville Pike, MA 20852-3139 Rockville, USA.
(tel: 1–301–8812486; fax: 1–301–8817303)

1996/05/05—10 96th Meeting American Roentgen Ray Society
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contact: American Roentgen Ray Soc,
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1996/05/15—18 77th Deutscher Roentgenkongress
venue: Wiesbaden, Germany.
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