A 21-year-old male patient presented with the symptoms of weakness in both legs, urine and stool incontinence for several months. Neurological examination revealed flaccid bilateral leg. Sonography showed heterogeneously echogenic multilobulated soft-tissue mass with several cystic spaces within it. Magnetic resonance imaging showed iso-signal intensity soft-tissue mass in T1 weighted imaging with many high-signal intensity cystic areas. After intravenous administration of gadolinium contrast agent, the tumor showed moderate contrast enhancement. The most common sarcomatous lesions with cystic degeneration include chondroid tumor (such as chondrosarcoma), liposarcoma, synovial sarcoma, and malignant peripheral nerve sheath tumors. Tumor biopsy showed malignant peripheral nerve sheath tumor. The patient received arterial embolization, radiotherapy, and regular chemotherapy after the diagnostic workup.

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CASE REPORT

Imaging Presentation of Malignant Peripheral Nerve Sheath Tumor at Sacral Region

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Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) most commonly occur in the deep soft tissues, usually close to a nerve trunk [1]. The most common sites are the sciatic nerve, brachial plexus, and sacral plexus. MPNSTs and neurofibromas are not uncommon in adolescents and adults with neurofibromatosis Type 1 (NF1), with an approximate lifetime risk of 10%. These malignancies frequently arise from large plexiform neurofibromas or extensive peripheral nerve lesions [2,3].

We herein presented a case of a giant MPNST at sacral region with presentation of bilateral leg weakness, accompanied by urine and stool incontinence for several months.

Case Report

A 20-year-old male patient had a history of NF for 15 years. A mass over his left face was noted years ago. He had...
received surgical intervention for his face mass at another medical center 5 years ago and operation scar was found over his left face. A mass over lower back was detected in recent years. The patient and his family did not pay attention to it seriously. However, the mass enlarged progressively. Urine and stool incontinence occurred later. Bilateral legs weakness happened 3 months ago and paralysis developed 2 weeks ago. He was transferred to our hospital for further evaluation and management.

Plain film of pelvis revealed increased soft-tissue density in the pelvic cavity. Loss of normal cortical outline of the upper sacrum with bone destruction is considered (Fig. 1). Sonography disclosed a heteroechoic mass, composed of echogenic solid component, some irregular cystic spaces, associated with coarse calcifications in the pelvic cavity. The estimated greatest dimension was 15 cm. The sonographic pattern is compatible with neoplasm such as sarcomatous tumor or chordoma (Fig. 2).

Magnetic resonance imaging (MRI) of the lumbar spine and sacrum before and after intravenous gadolinium injection was performed. A huge tumor mass about 15 cm in size was found in the upper pelvic cavity with severe destruction of the sacrum and intraspinal extension to lumbosacral junction causing obliteration of the dural sac. The differential diagnoses included unusual neurogenic tumor (neurofibroma, neurofibrosarcoma), unusual chordoma, and giant cell tumor (Fig. 3). The thoracic spine and thoracic spinal cord are well preserved in MRI.

Fig. 1. Pelvic X-ray image shows increased soft-tissue density in the pelvic cavity. Loss of normal cortical outline of the upper sacral bone (arrows) is noted.

Fig. 2. Pelvic sonography: (A, B) transverse scans, (C, D) sagittal scans. A heterogeneously echogenic mass is noted in the pelvic region, composed of solid components, some irregular cystic spaces (curved arrows), and associated with coarse calcifications (arrows), compatible with a tumor such as sarcomatous lesion or chordoma. Color Doppler sonography with sagittal scan (D) of the tumor demonstrates color flow signals in the peripheral zone (dot arrows), compatible with a hypervascular tumor.
During hospitalization, the chest computer tomography was performed and revealed at least 20 variable size soft-tissue nodules in both lung fields, compatible with multiple lung metastases. The whole body bone scan showed destruction at sacrum and bilateral sacroiliac junctions. There were no evidences of bony abnormality in other skeleton. Then tumor biopsy was arranged and pathology report proved to be MPNST. Trans-arterial embolization therapy and radiotherapy were performed for further treatment. Postchemotherapy, the patient felt general weakness after chemotherapy. Because no major complication occurred after chemotherapy, he was discharged and received regular chemotherapy at our oncologic clinic.

**Discussion**

MPNSTs arise in patients with or without associated NF. Tumors that arise in the absence of NF are somewhat more common in large series of MPNSTs [1]. Men and women are equally affected by this tumor. Most tumors occur in patients between 20 years and 50 years of age. MPNSTs may arise from plexiform neurofibromas, de novo or secondary to radiation therapy [2–4]. These tumors tend to present with masses larger than 6 cm with irregular borders and a history of rapid growth. MPNST often produces neurologic deficits in the distribution of the involved nerves because of impingement or mass effect [5].
Radiologically, MPNSTs and neurofibromas may appear indistinguishable; however, certain modalities may provide insight for differentiation [5]. Recent advances in ultrasound (US) technology have enabled the echotexture of soft-tissue tumors to be presented in greater detail. The following major parameters should be determined: echogenicity, contour, margin, composition, size, related surrounding tissue in grayscale ultrasonography, the grading of color Doppler US, and resistive index in spectral Doppler. Many more specific patterns can be observed, including phleboliths, hypechoic fat lobules, central necrosis, and internal calcification [6].

Both benign peripheral nerve sheath tumors (schwannomas and neurofibromas) and MPNSTs have variable sono graphic features. The target appearance on sonography (hyperechoic center with a hypoechoic periphery) described by Lin et al [7] was presented in most benign peripheral nerve sheath tumors. The target appearance has been attributed to a central fibrocollagenous region and a peripheral myxomatous region [8]. No MPNSTs showed a target appearance, which is similar to findings described with MRI [10].

MPNST usually presents heterogeneous hypoechogenicity, infiltrated margins, scalloped or ovoid contour, solid content, large size, and mild to moderate vascularity on color Doppler US [6]. The sonographic appearance of MPNSTs depends on their histologic characteristics. They are composed of Schwann cells and fibroblasts and are accompanied by variable amounts of myxoid change [9]. Myxoid changes demonstrate significant differences in biological behavior, ranging from completely harmless to malignant transformation [2]. Sonography may show heterogeneous hypoechoic lesion with infiltrated margins. They typically have multiple internal hypoechoic cystic areas, which may be related to the high water content of myxoid tissue, and cystic changes secondary to hemorrhage and/or necrosis [10]. Peripheral hypechoic areas with central nodules may be seen because of the fat content of myelin from Schwann cells, the less cellular myxoid tissue located in the periphery or the more vascular fibrous tissue was seen centrally.

In addition, our cases displayed some internal punctate echogenic calcification, which has been described as rare but virtually pathognomonic of nerve tumors [11]. A MPNSTs that have undergone degenerative change may usually have calcification [12]. This calcification is pathognomonic and can be differentiated from myxoid type liposarcoma (the most common liposarcoma) [7]. In well-differentiated liposarcoma, US usually reveals hypechoogenicity. However, in myxoid change of liposarcoma, some mature fat cells or lipoblasts could still exist within the tumor, resulting in focal hypechoic areas. US usually appears heterogeneous and hypechoic with hypechoic areas, which retains lipoblast nets.

Most MPNSTs were hypeeremic on color and power Doppler sonography, as in other malignant tumors. But hyperemia was rarely present in benign peripheral nerve sheath tumors [13]. The characteristic dumbbell lesion, a partly intradural and partly extradural tumor, represents a neurofibroma that expands the intervertebral foramina and may be detected with ultrasonography [2,14].

Surrounding bony destruction, amorphous bony fragments, and widening of intervertebral foramina is observed in most patients [6]. The ultrasonographic appearance of MPNSTs with bony destruction differs to that of osteogenic sarcoma because they present without the sunburst periosteal reaction.

MRI findings depend largely on the histopathological characteristics of the tumor. MPNSTs show low to intermediate signal intensity on T1 weighted images and heterogeneous signal intensity on T2 weighted images. The hyperintense regions on T2 weighted images correspond to areas of cystic degeneration or myxoid matrix, whereas the hypointense regions represent collagen and fibrous tissue [15]. Hypointense areas on T2 weighted images may enhance following gadolinium administration [10]. A malignant contrast enhancement pattern is an expected finding in neurofibromas with sarcomatous changes.

Surgical resection is the first choice of therapy, ideally with total removal of the tumor. Owing to a high risk of recurrence with incomplete resection, postoperative irradiation and chemotherapy are necessary; however, they are often used as adjuvant therapies even if the tumor is completely resected. Even with aggressive therapy, local recurrence of tumor is seen in 50% of patients [2]. Heterogenous metastatic spread occurs most commonly to the lungs. The reported 5-year survival rate for patients with MPNST without NF1 is as high as 50%. It drops to as low as 10% for MPNST patients with NF1 [16].

References