Uterine Leiomyosarcoma Presenting with Neurological Symptoms: Differential Diagnosis

Nörolojik Semptomlar ile Başvuran Uterus Leiomiyosarkoma Olgusu

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ABSTRACT Brain metastases of uterine leiomyosarcoma are very rare. In this report, we present a rare case of uterine leiomyosarcoma with brain metastasis who presented with diplopia, headache and right facial paresis. Cranial magnetic resonance imaging (MRI) revealed mass lesions that occupy the right side of the pons and the left inferior part of the frontal lobe. Whole-body ^{18}F-FDG PET-CT scan showed hypermetabolic activity in the uterine mass, lungs, peritoneal cavity, right gluteus muscle and in the right posteriolateral portion of the pons. Tru-cut biopsy was performed from the uterine mass. Histopathological examination of the biopsy specimen revealed pleomorphic leiomyosarcoma. Initial presentation of uterine leiomyosarcoma with neurological symptoms is very rare.

Key Words: Uterine neoplasms; neoplasm metastasis; brain neoplasms; neurologic manifestations

ÖZET Uterus leiomiyosarkomanın beyin metastazı çok nadirdir. Bu olgu sunumunda, sağ yüz felci, baş ağrısı ve çift görme ile başvuran beyin metastazlı uterus leiomiyosarkomalı nadir bir olguyu sunduk. Kranial MRI görüntüleme, frontal lob sol alt kısmında ve pons sağ yan tarafta yerleşik kitle lezyonları gösterdi. Tüm vücut PET/BT görüntülemesi ile pons sağ posterolateralde ve sağ gluteus kası, peritoneal kavite, akciğerler, uterusdaki kitlede hipermetabolik lezyonlar tespit edildi. Uterusdaki kitleden tru-cut biyopsi yapıldı. Biyopsi örneğinin histopatolojik incelemesinde, pleomorfik leiomiyosarkoma tanısı konuldu. Uterus leiomiyosarkomanın başlangıçta nörolojik semptomlar ile ortaya çıkması oldukça nadir bir durumdur.

Anahtar Kelimeler: Uterus kanseri; kanser metastazı; beyin kanseri; nörolojik belirtiler

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Uterine sarcomas are rare malignant neoplasms of mesodermic origin that represent up to 5% of uterine cancers. Leiomyosarcomas constitute approximately 25% of uterine sarcomas and 1% of all uterine malignancies.[1] Uterine leiomyosarcoma usually metastasizes to the lung or the peritoneal cavity. In contrast, metastasis of leiomyosarcomas to the brain is very rare.[1,2] We report a female patient with uterine leiomyosarcoma who had neurological deficit findings as the initial complaint.

A 60-year-old woman was admitted to the hospital with complaints of headache, diplopia, and right facial paresis in March 2006. Neurological examination revealed right abducens nerve palsy. Other physical examinations were normal. Complete blood count, biochemical tests and tumor markers were normal on admission. Her medical history

consisted of an anal fissure operation 8 months ago.

Cranial MRI revealed a mass lesion with a large necrotic central part, occupying the pons on the right side and extending to the Meckel's cave. A second mass lesion was detected on the gray-white matter junction of the left inferior frontal lobe. Both lesions showed prominent enhancement following intravenous contrast injection (Figure 1). Cerebral lesions were evaluated as metastases. Chest CT scan showed metastatic pulmoner nodules. A following pelvic MRI depicted a centrally necrotic large mass lesion located at the uterine fundus which showed strong contrast enhancement, with adjacent heterogeneous signal intensities over the perivesical and parametrial spaces indicating a possible infiltration. A second mass lesion measuring 4 cm in diameter, with similar imaging findings was also detected on the right lower quadrant, adjacent to the anterior abdominal wall (Figure 2). PET CT imaging was performed. Whole-body ^{18}F-FDG PET CT showed hypermetabolic lesions in the uterus, lungs, peritonel cavity, right gluteus muscle and in the right posteriolateral region of the pons (Figure 3).

Tru-cut biopsy was performed from the uterine mass. Histological examination of the biopsy specimen revealed a diagnosis of pleomorphic lei-

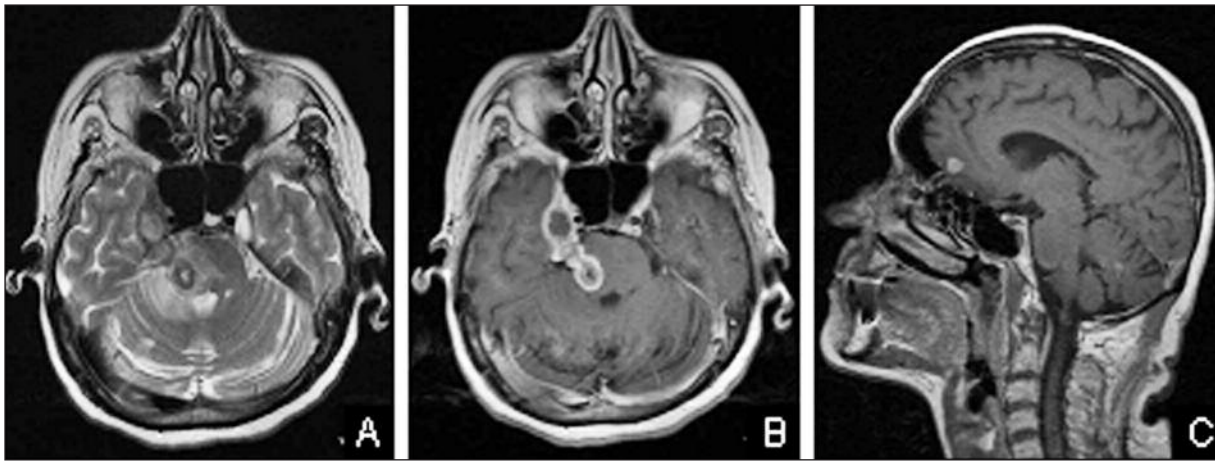

FIGURE 1: T2 weighted (A) and postcontrast T1 weighted images in the axial planes (B) show a peripheral enhancing mass lesion located in the right branchium pontis and extending to the right Meckel's cave. Postcontrast T1 weighted sagittal image (C) shows a second enhancing metastatic mass lesion on the left inferior frontal lobe.

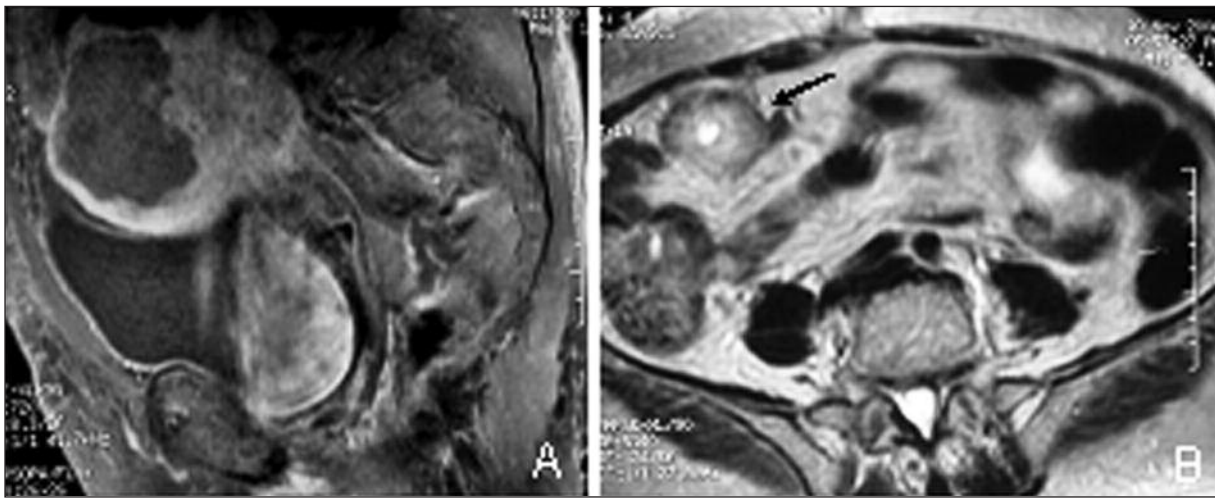

FIGURE 2: Fat suppressed sagittal postcontrast T1 weighted pelvic image (A) shows a large irregular-defined uterine enhancing mass lesion with necrotic degeneration areas and vaginal infiltration. Postcontrast T2 weighted image in the axial plane depicted a second mass lesion in the right lower quadrant (arrow) (B).

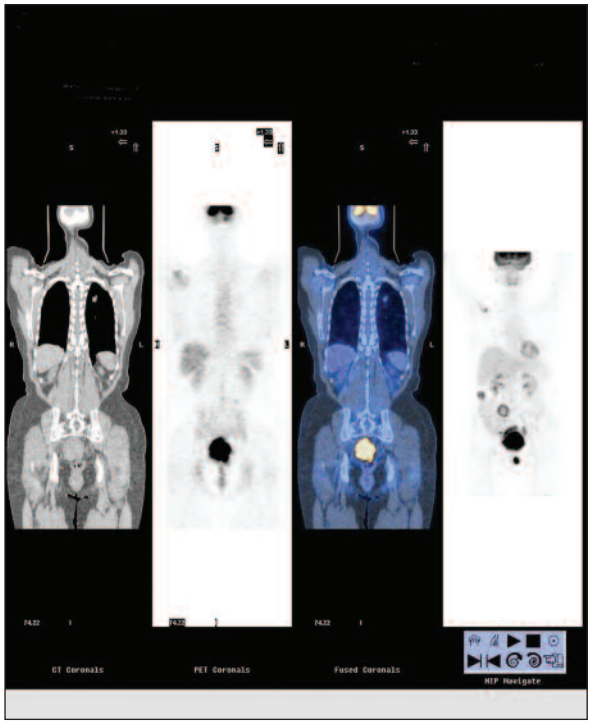

FIGURE 3: Pathological increased ^{18}F-FDG PET CT eclipse was observed in lesions in posterolateral region of the pons, in reticulonodular densities in right lung upper lob anterior, lower lob superior and middle lob lateral segments, in left lung lower lob superior and superior lingular segments, in the peritoneal cavity, in the right parotid lymph nod, in left scapula superior, in the precaval lymph node at the level of right renal hilus and in uterus.

omyosarcoma. Immunohistochemistry revealed diffuse positive staining for actin, desmin, vimentin and lack of staining for panceratin and S100 (Figure 4). 30 Gy of cranial radiotherapy was given in 3 Gy fractions, 5 days per week as initial treatment followed by a chemotherapy regimen consisting of adriamicin, ifosfamide and mesna administered in 3 cycles. The patient died during follow-up.

DISCUSSION

Leimyosarcoma tends to metastasize hematogenously, commonly to the lungs and other organs such as the liver.[3] In a study, Gadducci et al reported brain metastases in 1 of 126 patients with uterine leiomyosarcoma.[4] In the same study, common metastatic sites were detected as lungs and the abdomen; in contrast, bone and brain metastases were uncommon. In an autopsy study, Rose et al demonstrated brain metastases in 1 of 73 patients with uterine sarcoma.[2] Nevertheless, two recent studies reported that uterine leiomyosarcoma metastasized to the sphenoid sinus and the skull without pons metastasis.[5,6] Our patient was older and had multiple metastatic sites such as the lung, bone and brain as well as pons metastasis of the brain. However, she had only neurological spelling error findings at her admittance and no other spelling error.

The common prognostic factors were reported as tumor stage at diagnosis, mitotic count of tumor and age.[1,7] Large studies demonstrated 5-year overall survival rates of 20-30% and approximately 50% in patients with stage I diseases.[8]

The most common presenting symptoms of uterine leiomyosarcoma are abnormal vaginal bleeding and pelvic or abdominal pain. Less common symptoms are weight loss, weakness, lethargy, and fever.[1]

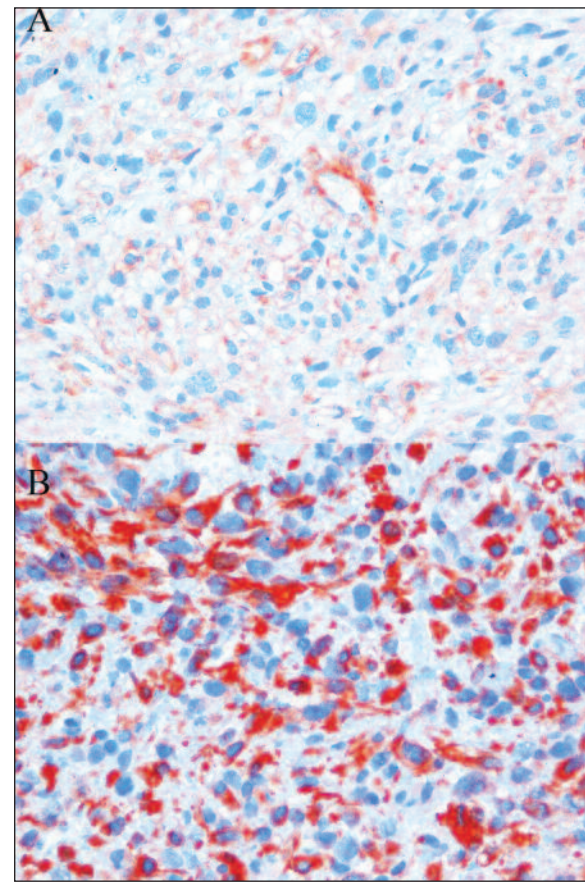

FIGURE 4: Actin (panel A) and desmin (panel B) positivity in tumoral cells in immunohystochemical (Actin & Desmin stain, x400).

The primary treatment of uterine leiomyosarcoma is surgery. Adjuvant chemotherapy and radiation therapy has limited activity on clinical outcome.[2] Chemotherapy regimens containing doxorubicine showed some benefit in adjuvant setting for uterine sarcomas.[9]

According to our knowledge, there are no reports on the initial neurological presentation in patients with brain metastasis of uterine leiomyosarcoma. This is the first case of metastatic leiomyosarcoma that presented with neurological deficit as the initial complaint.

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