Skull Metastasis from Uterine Leiomyosarcoma: A Case Report

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Abstract- Uterine leiomyosarcoma is a rare malignancy whose presenting symptoms usually are not specific for the disease. The clinical presentations include vaginal bleeding, pelvic pain or pressure, and awareness of an abdominal-pelvic mass. The diagnosis should be considered if rapid uterine enlargement occurs, especially in a post-menopausal woman. Local spreading of the tumor could involve the myometrium, pelvic blood vessels and lymphatics, contiguous pelvic structures, and then the abdominal organs, whereas distant metastasis most often involve the lungs. A 63-year-old female presented a rare uterine leiomyosarcoma metastasis to the skull with the manifestions of a bulging mass over her left occipito-parietal region.On admission, neurological examination revealed right hemiplegia. She underwent total resection of the tumor with the reconstruction of the dura, the skull plate and the scalp. Her post-operative course was smooth and the muscle power of her right limbs was improved from grade 1 to grade 3 after the procedure. The histological diagnosis is leiomyosarcoma. Aggressive management of the metastatic skull tumor is recommended in selected patients at least for a betterquality of life.

Key Words: Uterine leiomyosarcoma, Metastatic skull tumor

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INTRODUCTION

Leiomyosarcoma is an uncommon malignant neoplasm of smooth muscle origin, usually occurring in the retroperitoneum, subcutaneous tissues, blood vessels, gastrointestinal tract, genitourinary tract, and the uterus. Uterine leiomyosarcoma is a rare soft tissue neoplasm comprising about 1.3% of all uterine malignancies and approximately 25% of uterine sarcomas, with a female incidence of approximately 0.67/100000[(1)]. The tumor tends to invade and spread locally, but could also have an aggressive growth pattern with hematologic dissemination. The most common metastatic sites are lung, liver, and peritoneal cavity[(2-6)], whereas bone and brain metastases are rare. Several intracranial metastases from uterine leiomyosarcoma have been reported[(4,5,7-9)], with only one case of uterine leiomyosarcoma metastasis to the skull reported in English literature to our knowledge[(2)]. Here, we report another case of metastatic uterine leiomyosarcoma of the skull.

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CASE REPORT

A 63-year-old woman presented to us in July 2003 with the chief complaint of a progressive bulging mass over her left parietal region for about 6 months and progressive weakness of her right side extremities for more than one month. The past medical history shows that she was a case of hypertension and diabetes mellitus for years and was regularly followed up at a clinic. In April 2002, under the impression of uterine myoma, she underwent subtotal abdominal hysterectomy and bilateral salpingo-oophorectomy at a local general hospital, but the pathological diagnosis was uterine leiomyosarcoma. She was then referred to our Department of Gynecology and Obstetrics for chemotherapy. She received two courses of chemotherapy in May 2002 and June 2002 respectively. Due to the intolerable side effects, she finally gave up additional chemotherapy.

On admission, physical examination revealed a large fixed hard mass, which was about 8 cm in diameter and located at her left parietal region. Neurological examination demonstrated clear consciousness but right hemiplegia. The muscle power of her right side extremities was grade 1. Mild to moderate right limb contracture was also detected.

Cranial magnetic resonance imaging (MRI) disclosed a big tumor located at left high parietal region with involvement of the skull, as well as extension to the adjacent scalp. The tumor seemed to compress to the underlying brain parenchyma markedly with suspicious involvement of the brain tissue. The tumor was relative iso- to hypo- intensity on T1-weighted images, and gave homogenous signal on T2-weighted images. Gadolinium enhancement images showed strong enhancement of the tumor (Fig. 1).

Because her general condition was fair and she had a significant neurological deficit caused by the left parietal tumor, surgical intervention was advised and a surgical team including a neurosurgeon and a plastic surgeon were in charge of the procedures. Ten days after admission, she underwent a left occipito-parietal craniectomy with total removal of the tumor, and the involved scalp, skull plate, and dura, after which duraplasty and cranioplasty was done. During the operation, a demarcated lobulated hypervascular firm bulging mass, which was about 7cm ×6cm ×5cm in size and invades the galeal layer of overlying scalp, was found at her left occipito-parietal areas. The mass also penetrated the left parietal skull and invaded the underlying dura. On the inner surface of the involved dura, multiple nodules of the tumor were found. However, the arachnoid membrane between the nodular mass and the brain parenchyma was preserved. Grossly, the brain parenchyma was compressed, but not invaded by the tumor. The dural defect was repaired with Dura-guard, and cranioplasty was carried out with bone cement. Thereafter the scalp defect,

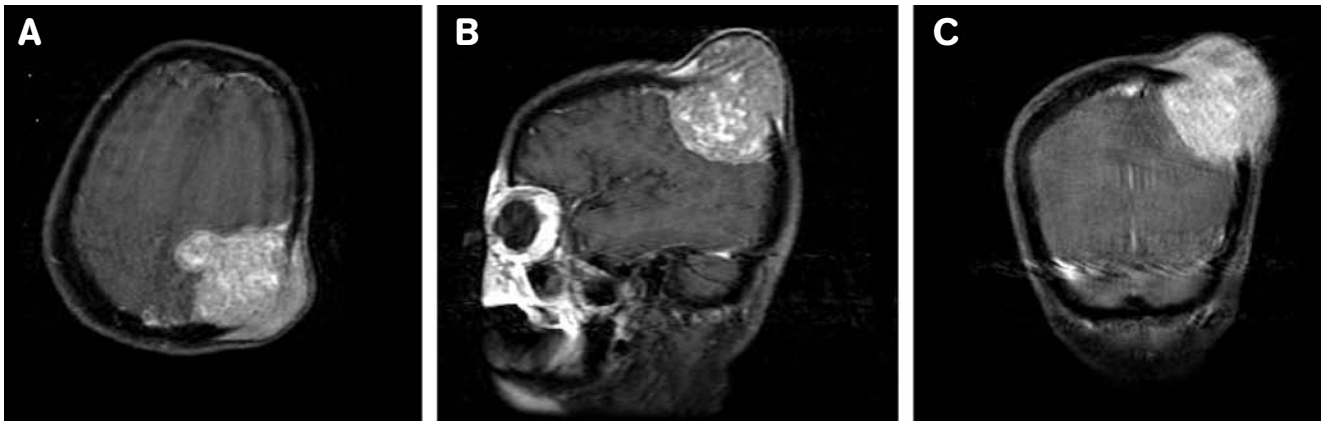

Figure 1. Gadolineum enhanced T1-weighted MRI of brain
Axial (A), Sagittal (B), and Coronal (C) views show strong enhancement of the mass as well as the adjacent dura and scalp of the left parietal region with epidural compression of the underlying brain tissues and suspicious involvement of the brain parenchyma.

roughly 9cm×9.5cm in area, was covered by a local rotation scalp flap. A sheet of split-thickness skin graft was harvested from her right thigh to cover the primary donar site.

Three pieces of tissue from the scalp, the skull bone, and the dura were sent for pathological examination. One fungating tumor was presented in each sectioned piece of the tissues, which measure 7.2cm×6cm×2.5cm, 9.2cm×8cm×2cm, 4.5cm×3.6cm×2.5cm, respectively. Microscopically, all three parts showed epitheloid tumor cells arranged in sheet and trabecular pattern. Pleomorphism and necrosis were seen. The tumor cells were diffusely positive for smooth muscle actin and focally immunoreactive for desmin, but negative for cytokeratin and EMA. These findings were consistent with metastatic leiomyosarcoma and similar in both morphology and immunophenotype to the uterine tumor resected 15 months earlier (Fig. 2). Skull metastasis from uterine leiomyosarcoma was therefore diagnosed based on the clinical features along with the histological picture.

The post-operative course was uneventful. The muscle power of her right extremities was improved to grade 3 with rehabilitation after the surgery. She was discharged with improved neurological function and without complication, and still strongly refused further chemotherapy. During the post-operative follow-up at our neurosurgical outpatient department, the muscle power of her right upper limb was improved to grade 4 and that of her right lower limb remained at grade 3, giving her and her family a better quality of life.

4 months after the surgery, she was sent to our emergency department from an outside hospital for impaired consciousness and a dehydrated state. From the refer sheet and her family, she suffered from poor oral intake for days and thus hypoglycemia with hypoglycemic encephalopathy probably ensued. The laboratory data showed elevated level of Blood Urea Nitrogen (BUN) and creatinine (Cr) with the picture of prerenal azotemia (BUN=112 mg/dL, Cr=3.7 mg/dL). The family refused any aggressive management despite of repeated explanations. She expired at home days later.

DISCUSSION

Uterine sarcomas are rare malignant neoplasms of mesodermic origin that account for about 5% of all uterine malignancies, less than a third of which are leiomyosarcoma[3,5,8,9]. Uterine sarcomas are divided into three major groups: mixed Muellerian tumor (also

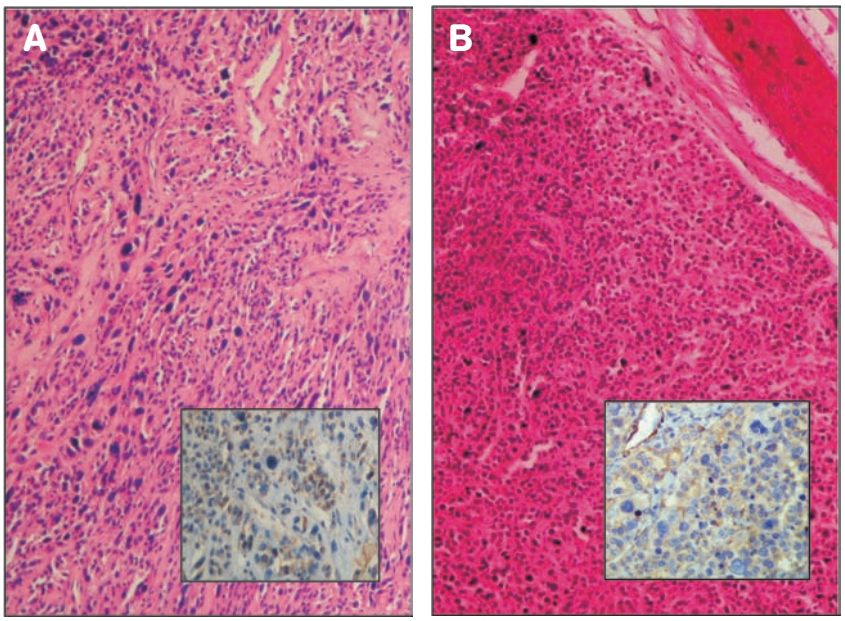

Figure 2. Histological pictures of the tumors
(A) Histopathology of the primary lesion in the uterus reveals a hypercellular tumor composed of spindle to ovoid cells with nuclear atypism and hyperchromatism in fascicular pattern. Occasional anaplastic tumor cells can also be noted. (original magnification: H&E stain ×200) Inlet: The tumor cells show cytoplasmic staining for smooth muscle action. (original magnification: smooth muscle actin stain, ×200)
(B) The metastatic tumor is composed of cells with similar morphological and immunohistochemical patterns to the primary lesion. (original magnification: H&E stain, ×200; inlet: smooth muscle actin stain, ×200)

known as carcinosarcoma), leiomyosarcoma, and endometrial stromal sarcoma[3,8,10]. Histologically, the World Health Organization classification of tumor recommends the following criteria for diagnosis of leiomyosarcoma: tumor with any coagulative tumor cell necrosis, or those without coagulative tumor necrosis but with diffuse, moderate to severe cytological atypia and a mitotic index of = or >10 mitotic figures per 10 high power fields[12]. The incidence of leiomyosarcoma arising in a leiomyoma is 0.13 to 0.81% according to the previous report[10].

Uterine leiomyosarcoma is an aggressive tumor and frequently metastasizes to lung, liver and peritoneal cavity[2-6]. The most significant prognostic factor of uterine leiomyosarcoma is the stage at diagnosis. The FIGO (Federation of International Gynecology and Obstetrics) staging for uterine sarcoma is as the following: stage I - confined to the corpus uteri; stage II - the corpus and the cervix is involved, but no extension outside the uterus; stage III - extension outside the uterus but confined to the true pelvis; stage IV - involvement of the bladder or bowel mucosa or metastasis to distant sites[10]. The primary treatment of uterine leiomyosarcoma is surgery[3,8]. Adjuvant chemotherapy and radiotherapy usually lead to unwanted side effects with an uncertain benefit of survival[3,8,10]. Because of the aggressive nature of this tumor, the recurrence rate is about 50% of stage I disease[10]. The overall prognosis of this malignant tumor is poor with the 5- year survival rate ranges from 0% to 68% reported in different series[4,5].

Skull metastases are usually seen in the advanced stage of disease. The clinical features of skull metastases include local swelling, local pain, and neurological deficits, according to on the metastatic sites. Surgery cannot alter the course of the underlying disease but can relieve the local discomfort and mitigate the neurological deficits if present. The patient may thus have a better quality of life. Surgical procedure is recommended in the following circumstances: presence of a neurologic deficit; massive destruction of bone and/or dura infiltration; painful mass; solitary metastases; confirmation of diagnosis[11].

Surgical resection of skull metastases always leaves the dural, skull and scalp defects which require reconstruction, and a watertight closure of the dura is the key to any reconstruction. When possible, primary repair is attempted, but for larger defects, both autogenous grafts (e.g. pericranial and temporalis flaps) and allogenic grafts are used for dural repair in conjunction with vascularized soft tissue coverage[13]. Bony defects can be reconstructed with either autogenous or alloplastic materials. Split-calvarial bone grafts harvested from the same operative field, split ribs and iliac bone are effective and common autogenous materials. In the cases of un-available autogenous grafts, metals such as titanium mesh, calcium ceramics, and polymers such as methylmethacrylate, hydroxyapatite cement, porous polyethylene can be used to cover intracranial contents and restore calvarial contour[14]. Scalp reconstruction after metastatic skull tumor resection is a challenging procedure, because after wide tumor excision, a complex composite wound with exposed brain is usually left. The ideal scalp flap should be durable and of the same thickness as the remaining scalp. It should also be hair-bearing and reliable enough to allow for timely postoperative adjuvant therapy[15]. Free tissue transfer including flaps of the latissimus dorsi, rectus abdominis, serratus anterior muscles[15], and anterolateral thigh[16] or local flap may provide an excellent scalp reconstruction.

Rotation flaps are local flaps made from adjacent tissues which is rotated in an arc to close a defect. Exposed skull bone or artifical skull plate requires coverage with some types of vascularized soft tissue. Rotation of the scalp flap provides an excellent coverage option. Since the pericranium of the primary donor site is preserved, a split-thickness skin graft can provide a quick and effective means of defect closure.

Our patient had a favourable surgical outcome, although she was expired 4 months after the operation because of her underlying medical disorder. Palliative surgical procedures may be indicated in selected patients of malignancy with skull metastasis.

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