Annals of Nuclear Medicine Vol. 19, No. 8, 729–732, 2005

Extraskeletal osteosarcoma: Extensive tumor thrombus on fused PET-CT images

Ukihide TATEISHI,* Umio YAMAGUCHI,** Takashi TERAUCHI,*** Tetsuo MAEDA,* Noriyuki MORIYAMA,*** Yasuaki ARAI* and Tadashi HASEGAWA****

*Divisions of *Diagnostic Radiology and Nuclear Medicine, and **Orthopedic Surgery, National Cancer Center Hospital*
****Division of Radiology, Research Center for Cancer Prevention and Screening, National Cancer Center*
*****Department of Clinical Pathology, Sapporo Medical University School of Medicine*

A 30-year-old woman developed extraskeletal osteosarcoma in the right buttock and thigh. Radiographs and unenhanced computed tomography (CT) showed a large, multilobulated mass accompanied by mineralized matrix. Contrast-enhanced CT and magnetic resonance (MR) images showed extensive tumor thrombus in the right internal- and external iliac veins. Co-registered positron emission tomography (PET) and CT images showed abnormal F-18 2-fluoro-2-deoxy-D-glucose (FDG) uptake in the tumor thrombus. PET study in our patient provided information concerning disease extent and viability of tumor thrombus.

Key words: FDG-PET, osteosarcoma, extraskeletal osteosarcoma, tumor thrombus

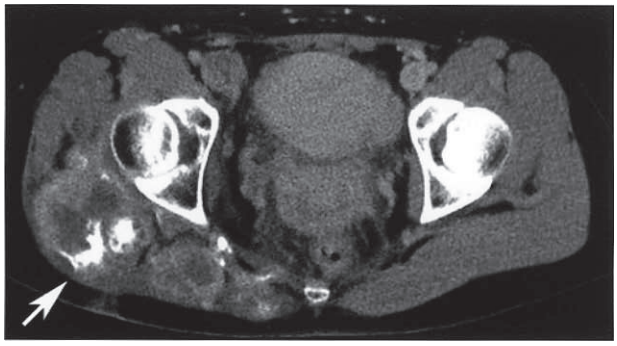
INTRODUCTION

EXTRASKELETAL OSTEOSARCOMA is a rare tumor that accounts for 5% of all osteosarcomas.[1] The tumor is typically highly malignant lesion and manifests as a uniform sarcomatous pattern with mineralized matrix and arises in the soft tissue without attachment to bone or periosteum.[2–5] The objectives of the present report are to demonstrate a patient with extraskeletal osteosarcoma presenting intravenous tumor thrombus that shows abnormal FDG uptake on fused PET-CT images. To our knowledge the PET characteristics of extraskeletal osteosarcoma have not been previously reported.

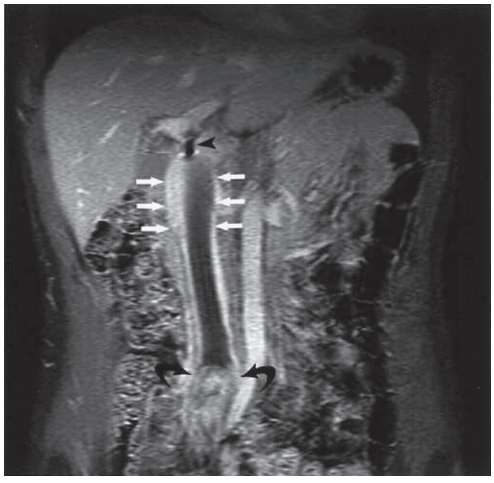
CASE REPORT

A 30-year-old woman presented with a right buttock mass and right leg discomfort. The painful mass had gradually enlarged, and the patient visited our outpatient clinic. A poorly-circumscribed hard mass was palpated in the right buttock and postero-lateral side of the thigh on physical examination. Radiographs showed a multilobulated mass in the right buttock and thigh with dense or predominantly peripheral mineralization. Unenhanced computed tomography (CT) showed a large, multilobulated mass partially accompanied by mineralized matrix (Fig. 1A). The tumor invaded the extracompartment from the buttock via the obturator foramen. The tumor was not attached to the surrounding bones. Contrast-enhanced CT showed extensive tumor thrombus in the right internal- and external iliac veins. Magnetic resonance (MR) examination from the upper abdomen to the thigh was performed after insertion of a transjugular inferior vena cava (IVC) stent. T1-weighted MR images showed a multilobulated mass in the deep soft tissue that was heterogeneously low- to isointense relative to the muscle. On T2-weighted MR images, the mass was heterogeneously low- to hyperintense. Contrast-enhanced MR images showed heterogeneous enhancement of the tumor. Homogeneous enhancement was seen in the tumor thrombus of the distal IVC beginning from the right iliac vein, while little enhancement was identified near the tumor thrombus of the proximal IVC (Fig. 1B). Tumor thrombus also extended to the contralateral external iliac vein. PET images followed by CT and MR examinations were obtained. The patient was tested for a normal glucose level (91 mg/d*l*) prior to PET scan. Emission scans from the base of the skull to the leg were obtained starting 60 minutes after

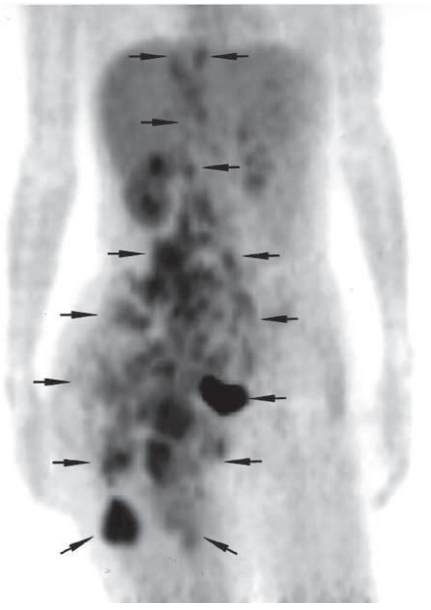
Received March 30, 2005, revision accepted July 14, 2005.
For reprint contact: Ukihide Tateishi, M.D., Division of Diagnostic Radiology and Nuclear Medicine, National Cancer Center Hospital, 5–1–1, Tsukiji, Chuo-ku, Tokyo 104–0045, JAPAN.



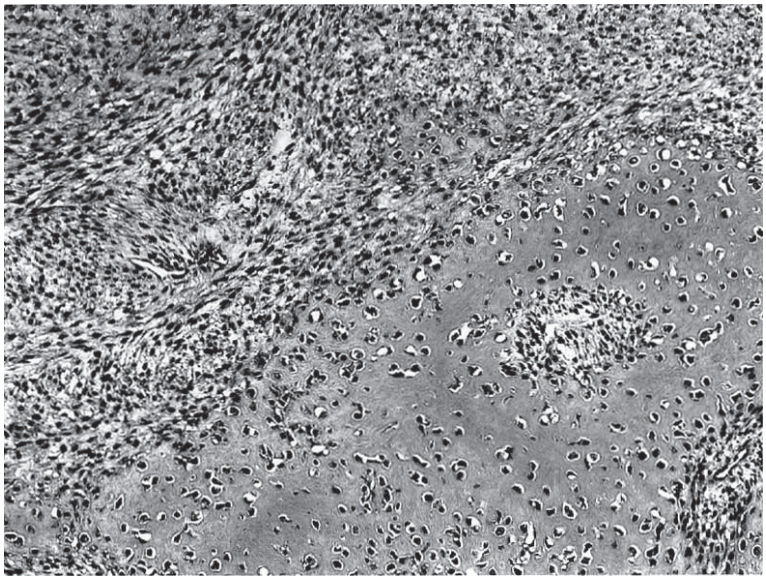
A



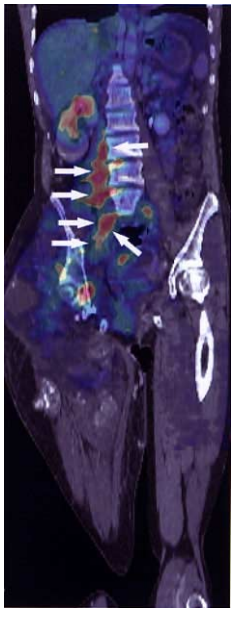
B



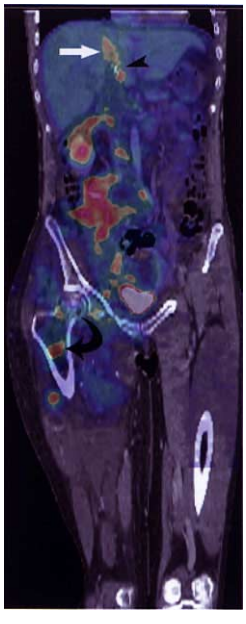
C



F



D



E

Fig. 1 A: 30-year-old woman with a right buttock mass and discomfort. Axial unenhanced computed tomography (CT) image reveals a multilobulated mass partially accompanied by mineralized matrix (*arrow*). B: Coronal contrast-enhanced fat saturated T1-weghted MR image reveals homogeneous enhancement in the tumor thrombus (*curved black arrows*) of distal inferior vena cava (IVC). The entry of the tumor thrombus is obscure. Little enhancement in the tumor thrombus of proximal IVC (*white arrows*) is noted. Metal artifact of the IVC stent is seen (*black arrowhead*). C: On FDG positron emission tomography (PET) image, area of hypermetabolism is noted in the masses of the right buttock and thigh (*arrows*). D: On fused PET-CT image, marked degree of abnormal FDG uptake is noted in the tumor thrombus of distal IVC (*white arrows*). E: On fused PET-CT image, moderate degree of FDG uptake is present in the tumor thrombus of proximal IVC (*white arrow*). The tumor thrombus of the most proximal IVC is higher than the position of the IVC stent (*black arrowhead*). FDG uptake in the right femur is caused by bone marrow stimulation (*curved black arrow*). F: Photomicrograph of pathology section reveals spindle cell proliferation with nuclear atypia and massive neoplastic bone formation. (Hematoxylin and Eosin, ×100).

intravenous administration of 367 MBq F-18 FDG. PET images revealed a high degree of FDG accumulation within the masses of the right buttock and thigh (Fig. 1C). The hypermetabolism was more prominent in the thigh lesion. The maximum standardized uptake (SUV) values for the buttock and thigh lesions were 5.78 and 7.14, respectively. Co-registered PET-CT images showed abnormal FDG uptake in the deep venous tumor thrombus. Marked degree of FDG tracer accumulation was noted in the tumor thrombus of the distal IVC (maximum SUV = 5.95, Fig. 1D). In contrast, the tumor thrombus of the proximal IVC, which showed little enhancement on contrast-enhanced MR images also showed moderate degree of FDG uptake (Fig. 1E). Spinal metastases were also identified on PET/CT fused images. Incisional biopsy of the lesion was performed by an orthopedic surgeon, and pathologic assessment was obtained. On gross examination, the lesion was firm and multinodular with superficial bone formation. Most of the lobules within the tumor had osteoid or immature bone formation at the periphery. Intravenous tumor extension was prominent in the central portion of the tumor. Histologic assessment revealed spindle cell proliferation with nuclear atypia and massive neoplastic bone formation (Fig. 1F). The findings were diagnostic of extraskeletal osteosarcoma. The patient received administration of chemotherapeutic agents (Adriamycin 90 mg and Ifosfamide 16.8 g) and is alive with disease.

DISCUSSION

Extraskeletal osteosarcoma is a rare soft tissue tumor characterized by malignant osteoid formation which has no attachment to bone or periosteum. Tumor typically occurs in the thigh of patients older than 30 years old compared to its skeletal counterpart.[1,2] The histological appearance of extraskeletal osteosarcoma, which consists of several histologic subtypes, is similar to that of osteosarcoma arising from the bone.

CT findings of extraskeletal osteosarcoma are characterized by the presence of large amounts of mineralized material within soft tissue tumor.[4–7] Pathologic examination revealed spotty to massive osteoid material admixed with osteoblastic features. However, tumors often lack radiologically discernible calcific or osteoid material on radiograph or CT images. Verma DGK and colleagues reported four cases without radiologically discernible calcific or osteoid material.[8] Of these, three tumors had predominantly fibroblastic features on microscopic observation. Microscopic features in extraskeletal osteosarcomas are comparable to those described in their skeletal counterparts with fibroblastic, osteoblastic, chondroblastic, telangiectatic, and small cell variants. Histologic subtypes other than amounts of mineralized material may affect radiographic or CT findings of extraskeletal osteosarcoma.

Radiologic features of extraskeletal osteosarcoma are often similar to those of parosteal osteosarcoma.[4] Tumor often shows peripheral mineralization.[5] In our case, CT showed a large, multilobulated mass accompanied by dense or peripherally dominant mineralization in part. Peripheral pattern of the mineralization is a characteristic but nonspecific finding on CT.

MR appearance of extraskeletal osteosarcoma is nonspecific.[8] Signal characteristics are affected by mineralized, cystic, hemorrhagic, and solid components of the tumor. A multilobulated mass was visualized as heterogeneously low- to isointense on T1-weighted MR images, and low- to hyperintense on T2-weighted MR images. MR signal patterns of our case were similar to those of previous reports.[7,8]

Various histologic types of soft tissue sarcomas often show intravascular growth, the most frequent being leiomyosarcoma of deep venous origin.[9–11] Leiomyosarcomas originating in the smooth muscles of the vessels show intravascular growth spreading to the right heart chambers in about 18.6% of all cases.[9] Tumors from the middle and upper segment of IVC tend to expand intraluminally, leading to partial, and rarely to total, occlusion of the vascular lumen.

Tumors with intravascular growth present similar clinical findings according to their sites. Abdominal pain is the main complaint found in 68% of the patients.[12] Other findings are nausea, emesis, fever, anorexia, dyspnea, and weight loss. They can be asymptomatic and found incidentally in unrelated operations or necropsy. Ultrasonography, CT, and MR can be used to confirm the diagnosis. MR has been found to be superior to CT in determining the intravascular nature of the tumor and its extension. MR images in our patient revealed homogeneous enhancement in the tumor thrombus of distal IVC beginning from the right iliac vein, and little enhancement was identified near the tumor thrombus of the proximal IVC. Although MR features were similar to those of secondary proximal thrombosis due to distal tumor thrombus, the PET images in our patient showed focal intense accumulation of FDG consistent with the intraluminal tumor extension and suggested viable tumor thrombus. The fact that the PET images also revealed abnormal uptake in the tumor thrombus of the proximal IVC made no difference in either tumor stage or treatment. However, the PET study in our patient provided information concerning the viability of tumor thrombus and might be useful to monitor the tumor response to treatment.

Osteosarcoma often metastasizes to secondary bone sites. F-18-FDG PET can provide detailed information on tumor staging and detecting recurrences in osteosarcoma. However, the benefit of F-18-FDG PET for detecting osseous metastases in osteosarcoma has been sparsely reported.[13] PET/CT fused images in our case showed spinal metastases. Similar to reports on increased accuracy of F-18-FDG PET for detecting bone metastases

in various cancers compared with conventional bone scanning, F-18-FDG PET/CT fused images may have advantages for the identification of bone metastases.

In conclusion, extraskeletal osteosarcoma is visualized as a mutilobulated mass accompanied by mineralized matrix with abnormal uptake on F-18-FDG PET/CT fused images. Co-registered PET/CT images also demonstrate abnormal FDG uptake in the venous tumor thrombus. Careful follow up is required in patients with tumor thrombus of IVC because of a high risk of poor prognosis.

ACKNOWLEDGMENT

This work was supported in part by grants for Scientific Research Expenses for Health and Welfare Programs, No. 17-12, The promotion and standardization of diagnostic accuracy in PET-CT imaging.

REFERENCES

1. Bane BL, Evans HL, Ro JY, Carrasco CH, Grignon DJ, Benjamin RS, Ayala AG. Extraskeletal osteosarcoma. A clinicopathologic review of 26 cases. *Cancer* 1990; 65: 2762–2770.
2. Lee JS, Fetsch JF, Wasdhal DA, Lee BP, Pritchard DJ, Nascimento AG. A review of 40 patients with extraskeletal osteosarcoma. *Cancer* 1995; 76: 2253–2259.
3. Lidang Jensen M, Schumacher B, Myhre Jensen O, Steen Nielsen O, Keller J. Extraskeletal osteosarcomas: a clinicopathologic study of 25 cases. *Am J Surg Pathol* 1998; 22: 588–594.
4. Yi ES, Shmookler BM, Malawer MM, Sweet DE. Well-differentiated extraskeletal osteosarcoma. A soft-tissue homologue of parosteal osteosarcoma. *Arch Pathol Lab Med* 1991; 115: 906–909.
5. Okada K, Ito H, Miyakoshi N, Sageshima M, Nishida J, Itoi E. A low-grade extraskeletal osteosarcoma. *Skeletal Radiol* 2003; 32: 165–169.
6. Wesseling FJ, Tjon A Tham RT, Breed W, Klinkhamer PJ. Retroperitoneal extraskeletal osteosarcoma. *AJR* 1990; 155: 1139–1140.
7. Doud TM, Moser RP Jr, Giudici MA, Frauenhoffer EE, Maurer RJ. Extraskeletal osteosarcoma of the thigh with several suspected skeletal metastases and extensive metastases to the chest. *Skeletal Radiol* 1991; 20: 628–632.
8. Varma DG, Ayala AG, Guo SQ, Moulopoulos LA, Kim EE, Charnsangavej C. MRI of extraskeletal osteosarcoma. *J Comput Assist Tomogr* 1993; 17: 414–417.
9. Hines OJ, Nelson S, Quinones-Baldrich WJ, Eilber FR. Leiomyosarcoma of the inferior vena cava: prognosis and comparison with leiomyosarcoma of other anatomic sites. *Cancer* 1999; 85: 1077–1083.
10. Le Minh T, Cazaban D, Michaud J, Ginestet-Auge MC, Laporte MH, Patelli A, et al. Great saphenous vein leiomyosarcoma: a rare malignant tumor of the extremity-two case reports. *Ann Vasc Surg* 2004; 18: 234–236.
11. Mourra N, Tiret E, Parc Y, de Saint-Maur P, Parc R, Flejou JF. Endometrial stromal sarcoma of the rectosigmoid colon arising in extragonadal endometriosis and revealed by portal vein thrombosis. *Arch Pathol Lab Med* 2001; 125: 1088–1090.
12. Greenwald U, Newman E, Taneja S, Rockman C. Malignant epithelioid angiosarcoma of the external iliac vein presenting as venous thrombosis. *Ann Vasc Surg* 2004; 18: 493–496.
13. Brenner W, Bohuslavizki KH, Eary JF. PET imaging of osteosarcoma. *J Nucl Med* 2003; 44: 930–942.