bo-sacral magnetic resonance imaging (MRI) showed a heterogeneously enhanced 6.7 cm sized lesion involving the sacrum, sacroiliac joint, and both ilium with extraosseous mass formation containing a necrotic portion (Fig. 1A).

We performed positron emission tomography/computed tomography (PET/CT) and bone scan to assess for metastatic lesion. PET/CT and bone scan revealed hot uptake in the sacropelvic area, T8, and occipital skull area. Thoracic MRI showed an epidural enhancing 1.7 cm sized lesion in the posterior element of T8 (Fig. 1B). In addition, findings on brain imaging studies showed an enhancing mass measuring 3.8 cm with occipital bone destruction and mild brain compression (Fig. 1C).

Percutaneous biopsy of S1 was performed for diagnosis. Pathology of the biopsy indicated a type undetermined malignant neoplasm, suspected as a malignant spindle cell tumor.

On angiography, large feeding vessels were observed in sacral lesion, and tumor embolization using Gelfoam was performed prior to perform an open surgery. Partial removal of the tumor in sacroiliac area was done; then, gross total removal of the tumor at T8 was subsequently performed in a single stage. The tumor was extremely hypervascular and not well demarcated.

INTRODUCTION

Solitary fibrous tumor (SFT) is a rare tumor which was first described as originating from the pleura and occurring most commonly in the thoracic cavity. However, it is now recognized that these rare tumors can occur throughout the body. There have been only 8 case reports of extradural spinal SFT, also only 8 case reports of head and neck SFT involving bony structures. Although the majority of SFT are benign, sometimes these tumors show aggressive clinical course including recurrence or multiple invasions, report on a malignant SFT of tandem lesions in the various bony structures, including skull, thoracic spine, and sacral spine, with a rapid recurrence and metastasis. Although malignant SFT is extremely rare, it should be considered in the differential diagnosis and careful follow-up is needed.

Key Words: Solitary fibrous tumors · Metastasis · Skull · Spine.

CASE REPORT

A 54-year-old female patient was referred to our spine center with a two-month history of lower back pain and radiating pain in both legs, which was persistent even at night. She had constipation with a symmetric leg muscle power grade of IV/V. Lumbosacral magnetic resonance imaging (MRI) showed a heterogeneously enhanced 6.7 cm sized lesion involving the sacrum, sacroiliac joint, and both ilium with extraosseous mass formation containing a necrotic portion (Fig. 1A).

We performed positron emission tomography/computed tomography (PET/CT) and bone scan to assess for metastatic lesion. PET/CT and bone scan revealed hot uptake in the sacropelvic area, T8, and occipital skull area. Thoracic MRI showed an epidural enhancing 1.7 cm sized lesion in the posterior element of T8 (Fig. 1B). In addition, findings on brain imaging studies showed an enhancing mass measuring 3.8 cm with occipital bone destruction and mild brain compression (Fig. 1C). Percutaneous biopsy of S1 was performed for diagnosis. Pathology of the biopsy indicated a type undetermined malignant neoplasm, suspected as a malignant spindle cell tumor.

On angiography, large feeding vessels were observed in sacral lesion, and tumor embolization using Gelfoam was performed prior to perform an open surgery. Partial removal of the tumor in sacroiliac area was done; then, gross total removal of the tumor at T8 was subsequently performed in a single stage. The tumor was extremely hypervascular and not well demarcated.
Malignant Solitary Fibrous Tumor of Tandem Lesions

S Son, et al.

The tumors showed multifocal necrosis, high cellularity, and marked cellular atypia. Mitotic figures were frequently observed [up to 10/10 high-power fields (HPFs)]. By immunohistochemistry, the tumor cells were positive for CD34, CD99, Bcl-2 and EMA (focal-like +). As a result, the tumors were diagnosed as malignant solitary fibrous tumor (Fig. 2).

Although there was a lack of clinical evidence about adjuvant chemotherapy or radiotherapy, adjuvant chemotherapy with adriamycin and cisplatin was administered. However, just 2 days after starting adjuvant chemotherapy, one month after the initial surgery, she complained of aggravated upper back pain and paraparesis. Findings on follow-up thoracic MRI showed a recurred expansive posterior epidural mass with cord compression at T8 (Fig. 3). For the recurred tumor at T8, stereotactic radiosurgery was administered at a dose of 4 fractionated 32 Gy. After radiosurgery, her upper back pain and paraparesis showed improvement.

Gradually, she complained of progressive whole body pain. Despite chemotherapy, follow-up whole spine MRI and chest radiography revealed metastasis in multiple vertebrae and both lungs. Although palliative cervical and thoracic spine radiotherapy (dose of 5 fractionated 15 Gy) was performed for relieving intractable pain, her symptoms did not show improvement and the patient finally died due to poor general condition six months after first open surgery.

DISCUSSION

Classically, SFT, also known as localized fibrous tumor, has been known as a rare spindle-cell neoplasm originating from the
pleura. Since the first case report in 1931, studies of SFT from the pleura were prolific in the 1980s. Although SFT occurs mainly in the pleura, it can be seen in various organs, and has recently been considered to be a mesenchymal neoplasm originating from ubiquitous dendritic interstitial cells. According to previous studies, approximately 30% of SFT arise in extrathoracic locations. Reported extrathoracic locations include the mediastinum, pericardium, peritoneum, retroperitoneal space, pelvis, adrenal gland, kidney, liver, peristomeum, salivary gland, thyroid gland, lacrimal gland, breast, nasopharynx, orbit, urogenital system, skin, meninges, and spinal cord.

Due to overlapping histologic features, differentiation of SFT from other soft tissue tumors may be difficult. Performance of immunohistochemical staining is necessary in order to rule out other differential diagnoses. Typical immunohistochemical features of SFT are a positive result for CD34, Bcl-2, and CD99, and a negative result for α-SMA, desmin, pan-cytokeratin, and S-100 protein. However, these markers are frequently overexpressed in a range of other soft tissue tumors including hemangiopericytoma. As a result, a differential diagnosis of hemangiopericytoma with SFT is difficult, even though their clinical courses are obviously different. Therefore, 2002 World Health Organization criteria for soft-tissue tumors treat those two neoplastic entities as a single section, and many lesions that were called hemangiopericytomas prior to 1990 could now be called SFT. However, in general, hemangiopericytomas are more cellular and have higher Ki-67 rated of 5-10%. In the current case, light microscopic and immunohistochemical features were typical findings for malignant SFT rather than hemangiopericytoma.

Most thoracic SFTs are asymptomatic at presentation and are diagnosed as incidental finding on radiographs and CT images of the chest. Extrathoracic SFTs, however, are usually symptomatic; depending on location, the manifestations are a painless mass or local pressure effects. In addition, an association of approximately 5% of SFTs with hypoglycemia due to secretion of insulin-like growth factors has been reported. In the current case, initially, the patient complained lower back pain with radiating pain in both legs due to compression of nerve roots of S1 and S2. In imaging studies, including MRI and CT, imaging features of SFT are relatively nonspecific. However, in general, SFTs have discrete margins, and the majority of SFTs have shown a lobulated contour. SFTs were typically well defined, tending to displace adjacent structures, and local invasion is not common. However, in our case, the tumor margin was not well defined, and local invasion was obvious. These findings strongly implied that tumor should be malignant. As in the current case, a useful distinguishing imaging feature of SFTs is the presence of large collateral feeding vessels. However, the presence of these vessels does not appear to be related to the histologic subtype of the tumor. On the other hand, the tumor occurred in multiple tandem regions, including the skull, thoracic spine, lumbar spine, and pelvic bone. The current report is first case about the malignant SFT of multiple tandem lesions.

At present, due to the rarity of the disease, standard therapies for malignant SFT have not been well established. Surgical en bloc removal has been recommended as the treatment of choice for SFT. Recurrence occurred commonly in cases involving incomplete excision, possibly caused by the level of difficulty in achievement complete resection. In these cases, adjuvant chemotherapy or radiotherapy may play a role in prevention of recurrence. However, no previous studies have demonstrated the effect of chemotherapy or radiotherapy.

The majority of SFTs are benign, and the malignant form accounts for 9-22%. Based on previous case reports, malignant SFT showed rapid local recurrence and distant metastasis. Some authors have suggested that the size of a SFT is one of the best indicators of malignancy. In addition, findings such as nuclear atypia, increased cellularity, necrosis, and greater than 4 mitoses/10 HPFs, are suggestive of the malignant potential of SFTs. Otherwise, some studies have reported that the prognosis of an SFT is most likely dependent upon complete resection rather than histologic findings. Hence, careful follow-up may be needed, even if the tumor is small and benign in appearance at the time of presentation.

The current patient showed malignant characteristics with respect to tumor size, multiple lesions, local invasion, impossibility of complete resection, and histopathologic findings. Fortunately, at initial presentation, the current patient showed a significant response to open surgery and radiosurgery. However, despite multidisciplinary treatment including adjuvant chemotherapy and radiotherapy, the tumor showed rapid local recurrence and aggressive metastasis to the whole spine and lung.

CONCLUSION

The authors report here the first case of patient with malignant SFT of tandem lesions in the skull and spine with a rapid recurrence and metastasis. Although malignant SFT is extremely rare, it should be considered in the differential diagnosis of tumors in the spine and skull, and careful follow-up is needed.

References
1. Briselli M, Mark EJ, Dickersin GR: Solitary fibrous tumors of the pleura: eight new cases and review of 360 cases in the literature. Cancer 47: 2678-2689, 1981
2. Chan JK: Solitary fibrous tumour—everywhere, and a diagnosis in vogue. Histopathology 31: 588-576, 1997
3. Chang ED, Lee EH, Won YS, Kim JM, Suh KS, Kim BK: Malignant solitary fibrous tumor of the pleura causing recurrent hypoglycemia; immunohistochemical stain of insulin-like growth factor I receptor in three cases. J Korean Med Sci 16: 220-224, 2001
4. Cox DP, Daniels T, Jordan RC: Solitary fibrous tumor of the head and neck. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 110: 79-84, 2010
5. Demiccio EG, Park MS, Araujo DM, Fox PS, Bassett RL, Pollock RE, et al.: Solitary fibrous tumor: a clinicopathological study of 110 cases and
1. Fargen KM, Opalach KJ, Wakefield D, Jacob RP, Yachnis AT, Lister JR: The central nervous system solitary fibrous tumor: a review of clinical, imaging and pathologic findings among all reported cases from 1996 to 2010. Clin Neurol Neurosurg 113:703-710, 2011
6. Mussak EN, Tu JJ, Voigt EP: Malignant solitary fibrous tumor of the hypopharynx with dysphagia. Otolaryngol Head Neck Surg 133:805-807, 2005
7. Fukunaga M, Naganuma H, Nikaido T, Harada T, Ushigome S: Extra-pleural solitary fibrous tumor: a report of seven cases. Mod Pathol 10:443-450, 1997
8. Gengler C, Guillou L: Solitary fibrous tumour and haemangiopericytoma: evolution of a concept. Histopathology 48:63-74, 2006
9. Gold JS, Antonescu CR, Hajdu C, Ferrone CR, Hussain M, Lewis JI, et al.: Clinicopathologic correlates of solitary fibrous tumors. Cancer 94:1057-1068, 2002
10. Ha JK, Park BJ, Kim YH, Lim YJ: Orbital solitary fibrous tumor: a case report and diagnostic clues. J Korean Neurosurg Soc 46:77-80, 2009
11. Kawamura S, Nakamura T, Oya T, Ishizawa S, Sakai Y, Tanaka T, et al.: Advanced malignant solitary fibrous tumor in pelvis responding to radiation therapy. Pathol Int 57:213-218, 2007
12. Klemperer P, Coleman BR: Primary neoplasms of the pleura. A report of five cases. Am J Ind Med 22:1-31, 1992
13. Nielsen GP, O’Connell JX, Dickersin GR, Rosenberg AE: Solitary fibrous tumor of soft tissue: a report of 15 cases, including 5 malignant examples with light microscopic, immunohistochemical, and ultra-structural data. Mod Pathol 10:1028-1037, 1997
14. Okike N, Bernatz PE, Woolner LB: Localized mesothelioma of the pleura: benign and malignant variants. J Thorac Cardiovasc Surg 75:363-372, 1978
15. Shnayder Y, Greenfield BJ, Oweity T, DeLacure MD: Malignant solitary fibrous tumor of the tongue. Am J Otolaryngol 24:246-249, 2003
16. Sung SH, Chang JW, Kim J, Lee KS, Han J, Park SI: Solitary fibrous tumors of the pleura: surgical outcome and clinical course. Ann Thorac Surg 79:303-307, 2005
17. Vallat-Decouvelaere AV, Dry SM, Fletcher CD: Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. Am J Surg Pathol 22:1501-1511, 1998
18. Westra WH, Grenko RT, Epstein J: Solitary fibrous tumor of the lower urogenital tract: a report of five cases involving the seminal vesicles, urinary bladder, and prostate. Hum Pathol 31:63-68, 2000
19. Wignall OJ, Moskovic EC, Thway K, Thomas JM: Solitary fibrous tumors of the soft tissues: review of the imaging and clinical features with histopathologic correlation. AJR Am J Roentgenol 195:W55-W62, 2010
20. Fargen KM, O’Connell JX, Dickersin GR, Rosenberg AE: Solitary fibrous tumor of soft tissue: a report of 15 cases, including 5 malignant examples with light microscopic, immunohistochemical, and ultra-structural data. Mod Pathol 10:1028-1037, 1997

Malignant Solitary Fibrous Tumor of Tandem Lesions | S Son, et al.