EDORIUM Journals

J Case Rep Images Oncology 2024;10(2):1–5.
www.ijcrioncology.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Primary alveolar soft part sarcoma of the ilium: A case report and literature review

Ikuo Kudawara

ABSTRACT

Introduction: The primary alveolar soft part sarcoma (ASPS) of bone is extremely rare, and the number of published reporting cases is limited.

Case Report: We present a 30-year-old woman with primary ASPS of ilium. Computed tomography and magnetic resonance imaging revealed a destructive and moth-eaten appearance in the ilium with a soft tissue mass. ^{18}F-fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) revealed high uptakes of FDG in the left ilium. *ASPSCR1-TFE3* gene fusion was detected by reverse transcriptase polymerase chain reaction (RT-PCR). Wide resection was performed. One year after the surgery, a local recurrence developed. However, there has been no evidence of disease in the last eight years after the second surgery.

Conclusion: ^{18}F-fluorodeoxyglucose positron emission tomography-computed tomography and RT-PCR besides routine radiological and histopathological studies helped achieve the accurate diagnosis and provide appropriate treatment in the present case.

Keywords: Alveolar soft part sarcoma, Bone, Gene fusion, Ilium, Magnetic resonance imaging

How to cite this article

Kudawara I. Primary alveolar soft part sarcoma of the ilium: A case report and literature review. J Case Rep Images Oncology 2024;10(2):1–5.

Ikuo Kudawara[1], MD, PhD

Affiliation: [1]Department of Orthopaedic Surgery, National Hospital Organization Osaka National Hospital, 2-1-14 Hoenzaka, Chuo-ku, Osaka, Japan.

Corresponding Author: Ikuo Kudawara, Department of Orthopaedic Surgery, National Hospital Organization Osaka National Hospital, 2-1-14 Hoenzaka, Chuo-ku, Osaka 540-0006, Japan; Email: sugar36-dumpling99@w9.dion.ne.jp

Received: 31 March 2024
Accepted: 30 May 2024
Published: 12 July 2024

Article ID: 100133Z10IK2024

****

doi: 10.5348/100133Z10IK2024CR

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is a rare soft tissue sarcoma that mainly occurs in young adults and is located in the deep soft tissue of the extremities [1, 2]. This tumor is a slowly growing, predominantly hyper-vascular, and is occasionally present a pulsatile soft tissue mass with bruit [3]. Its local recurrence rate is 11–50% and following its distant metastases, in the long term, develops in the lung, liver, bone, and brain [1]. Local wide resection is generally thought to be the sole treatment given that both chemotherapy and radiotherapy are ineffective in this regard. Histologically, the tumor is composed of polygonal cells with eosinophilic cytoplasm it often forms the cell nests [1, 2]. Molecular analysis has also shown that *ASPSCR1-TFE3* gene fusion is characteristic of ASPS [2]. Additionally, the occurrence of primary ASPS of the bone is extremely rare, and only a limited number of case reports have been published in this regard. To the best of our knowledge, only two cases of primary ASPS of the ilium have been reported [4, 5]. Here, we report another case of primary ASPS of the ilium and present the corresponding clinical, radiological, and histological findings after surgical treatment and a 9-year follow-up period.

CASE REPORT

The case reported here is about a 30-year-old woman, who presented with a 6-month history of left buttock pain. She had no major past history. On physical examination, swelling and skin redness were observed on her left buttock. Further, an antero-posterior radiograph of the pelvis revealed the presence of osteolytic lesion in the left iliac crest (Figure 1A) and computed tomography (CT) revealed a destructive and moth-eaten appearance in the ilium as well as the presence of soft tissue mass

on both sides of the bone (Figure 1B). Furthermore, magnetic resonance angiography (MRA) revealed diffuse hyper-vascularity in the lesion (Figure 2A), and axial non-enhanced T1-weighted magnetic resonance imaging (MRI) of the lesion revealed a homogeneous higher signal intensity than the adjacent muscle. On both T2-weighted image and short tau inversion recovery (STIR) images, the high signal intensity predominantly denoted the lesion, while the low signal intensity corresponded to the central portion and flow voids (Figure 3). Additionally, based on MRI, the tumor measured 48×63×73 mm. These radiological findings strongly suggested the presence of a malignant bone tumor, possibly Ewing sarcoma or lymphoma.

For further identification, we performed ^{18}F-fluorodeoxyglucose position emission (FDG PET-CT), which demonstrated a high FDG uptake in the right kidney and left ilium of this patient with SUV_{max} values of 8.1 and 4.8, respectively (Figure 2B and C). Additional enhanced dynamic CT of the kidney showed no tumor lesions in the right kidney. Meanwhile, the high level of FDG uptake in the right kidney was judged to be the result of urine pool due to the double renal pelvis. Given that needle biopsy did not lead to the confirmation of a histopathological diagnosis, incisional biopsy was performed. Thus, we observed that the tumor was composed of proliferating round-shaped cells with eosinophilic or clear cytoplasm. Via immunohistochemistry, we confirmed that the tumor cells were positive for PAS, but negative for HMB45, S100, and desmin. Moreover, occasional vein tumor embolisms were observed. These findings were suggestive of PEComa (perivascular epithelioid tumor) or ASPS. Additional reverse transcriptase polymerase chain reaction (RT-PCR) using a fresh frozen sample of the biopsy demonstrated the presence of *ASPSCR1-TFE3* gene fusion. Therefore, the tumor was diagnosed as ASPS based on the expression of this characteristic fusion gene.

En bloc excision was planned similar to standard surgical excision for primary malignant bone tumor, and during surgery, the tumor of the ilium with the surrounding iliacus, gluteus medius, and minimus muscles was resected (Figure 4). Reconstruction of the pelvis was not performed because the location of tumor resection was limited to the iliac wing. Macroscopically, the tumor of bone marrow extended to both sides, foaming a soft-tissue mass (Figure 5), and histological analysis revealed that the main tumor in the medullary canal extended to the surrounding muscles. Further, the tumor predominantly consisted of irregular sheets of round cells and vessels between trabecula, and showed necrosis at the central area in the tumor of the ilium (Figure 6). All surgical margins, including bone and soft tissue, were microscopically negative. One year after the initial surgery, a local recurrent tumor in the deep subcutis measured 2 cm in diameter was detected via MRI. The recurrent tumor was completely resected (R0) under general anesthesia, and MRI examinations of the pelvis were performed every four months for the first year after the resection of the recurrent tumor, and six months thereafter. Further, chest CT scans were obtained at every 3-month for the first year and at every 4-month intervals for the following four years, and six months thereafter. Careful physical examinations were also performed at every visit. There has been no evidence of disease within the last eight years after the surgery for the recurrence, and presently, she is ambulatory with a stick despite of mild limping at latest follow-up.

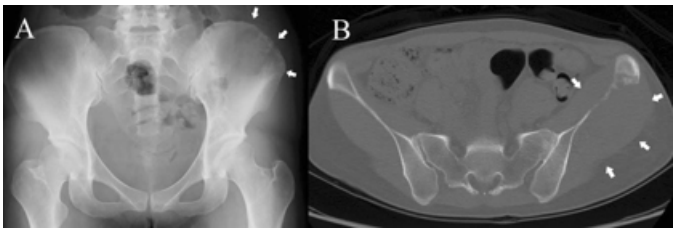

Figure 1: Plain X-ray image showing a predominantly osteolytic lesion in the left iliac crest (A). Axial CT image showing a destructive and moth-eaten appearance involving the left ilium, with the presence of a soft tissue mass on both the medial and lateral sides of the bone (arrows) (B).

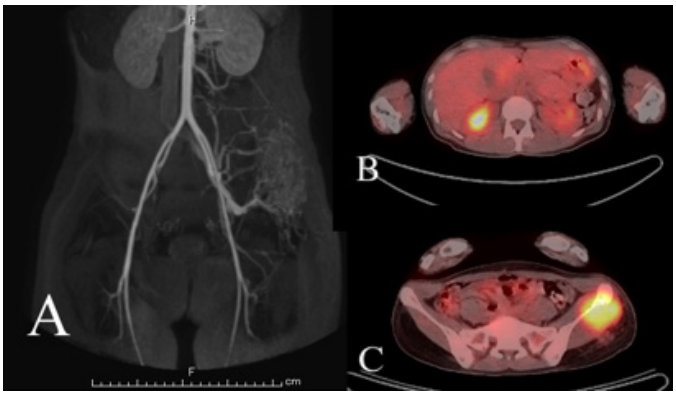

Figure 2: MRA image showing tumor hypervascularity in tumor of the left ilium (A). ^{18}F-FDG PET-CT image demonstrating increased FDG uptakes in the right kidney (B) and left ilium (C). The SUV_{max} values are 8.1 (B) and 4.8 (C), respectively.

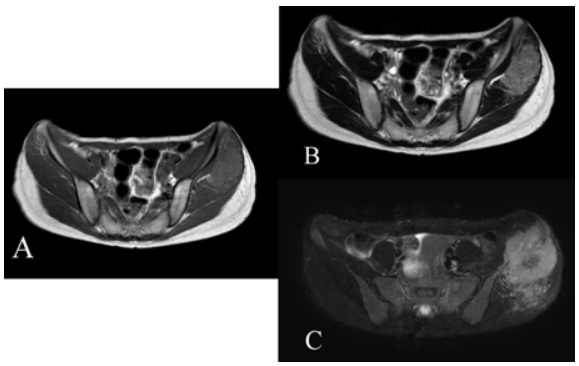

Figure 3: Axial T1-weighted MRI image revealing a higher signal intensity for the lesion than for the surrounding muscle in both intramedullary and extraskeletal tumors (A). T2-weighted and STIR images indicating that most of the tumor showed high signal intensities and occasional spotty low intensities (flow voids) (B, C). Central portion in the lateral soft tissue mass showing low intensity on all three sequences indicative of tumor necrosis (A–C).

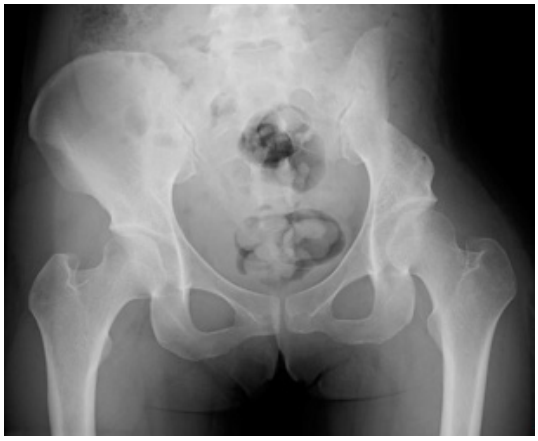

Figure 4: Postoperative plain X-ray of the pelvis.

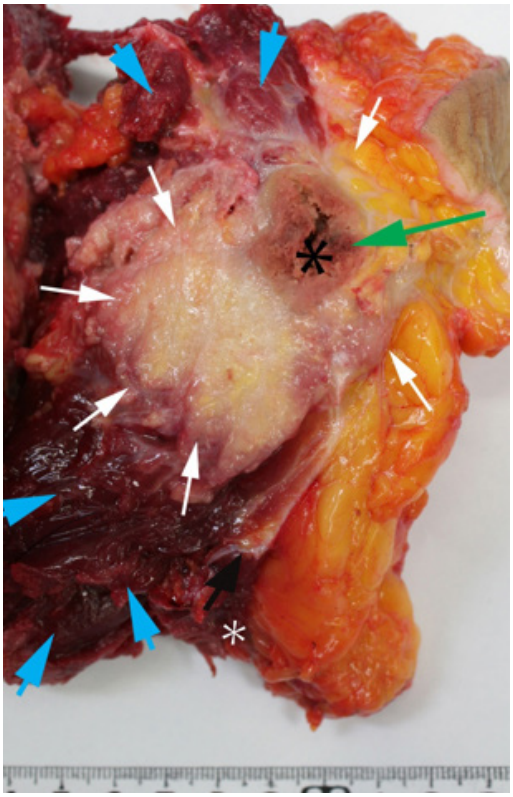

Figure 5: Axial cut surface of the resected tumor (white arrows) arising from the ilium (black arrow) and extending to the surrounding gluteus minimus, medius muscle (blue arrows), and iliacus muscle (white asterisk). The green arrow indicates the biopsy tract and the asterisk shows the tumor biopsy site.

An Institutional Review Board/Ethics Committee approval is not necessary for a case report at the author's institution.

DISCUSSION

Alveolar soft part sarcoma is a rare malignant soft tissue tumor that accounts for approximately <1% of all soft tissue tumors [1, 2]. Specifically, ASPS usually occurs

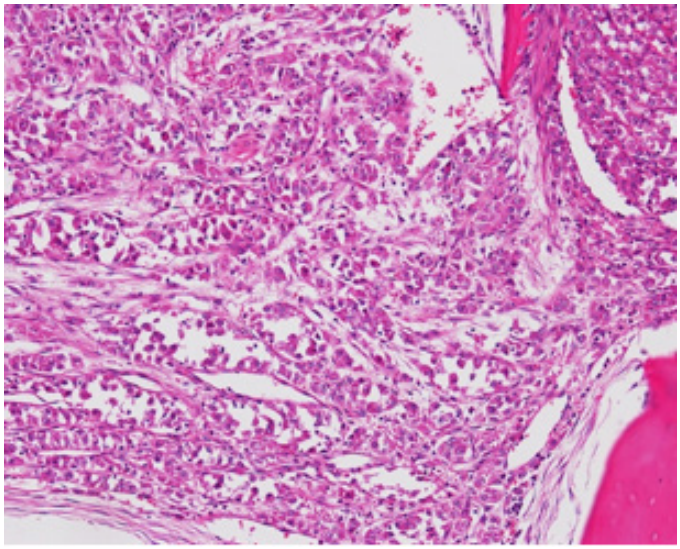

Figure 6: Photomicrograph showing the presence of irregular sheets consisting of round cells and vessels between trabecula. ×200 Hematoxylin-eosin.

in the deep soft tissue of the extremities and trunk, and its association with bone is very rare. Park et al. were the first to report six cases of ASPS arising in the bone [6]. The authors indicated that the tumor locations were the femur in three cases, tibia and fibula in two cases, and ilium in one case. Four of the six cases developed distant metastases, that is, pulmonary metastases (four cases) and brain metastases (two cases). Therefore, prior to this study, six cases of primary ASPS of the bone have been reported, with descriptions of the corresponding MRI and pathological findings (Table 1) [4, 5, 7–10]. Further, the tumor locations were: ilium (three cases), including the present case, and the fibula, thoracic spine, and sacrum for one case each. In two of the five previously reported cases, distant metastases occurred. Furthermore, the different cases showed varied survival periods and the follow-up period for the present case, including an episode of recurrence, is now nine years. However, the prognosis of ASPS of the bone is still unclear, considering the poor long-term outcomes of soft-tissue ASPS [1]. Therefore, in this reported case, careful follow-up for several more years is still necessary.

Osteolytic lesions, such as destructive or moth-eaten appearance in plain X-ray or CT images and soft-tissue masses in CT or MRI images, are the common radiological features in all ASPS cases (Table 1) [4, 5, 7–10]. These findings are common to primary malignant bone tumor, such as osteosarcoma, Ewing sarcoma, and lymphoma. However, the distinctive MRI features of ASPS are intermediate-high signal intensity in T1-weighted images and high signal intensity in T2-weighted images. In three of the six previously reported cases, signal voids and/or low signal intensity in the central necrosis were observed [5, 7]. These MRI findings are similar to that of soft tissue ASPS [11, 12].

Table 1: Summary of the published cases of ASPS of the bone

Author	Age/ Gender	Location	X-ray, CT	Border	Soft-tissue mass (CT or MRI)	Intensity of MRI: T1WI/ T2WI	Metastasis	*ASPSCR1-TFE3* gene fusion	Overall survival
Durkin (1999) [4]	22/F	Ilium	Lytic	Ill-defined	(+)	Low/high	Lung	n.a.	2Y
Koguchi (2005) [5]	33/F	Ilium	Lytic, destructive	Ill-defined	(+)	High/high	Lung, brain	n.a.	5Y
Aisner (2008) [7]	41/F	Fibula	Lytic, expansile, destructive	Ill-defined	(+)	High/high	Brain	(+)	1M
Zhu (2009) [8]	23/M	Thoracic spine	lytic, destructive	Ill-defined	(+)	Intermediate/ high	n.a.	(+)*	n.a.
Yavuz (2013) [9]	33/M	Scapula	lytic, moth-eaten	Ill-defined	(+)	High/high	Lung	n.a.	n.a.
Zandnik (2014) [10]	28/F	Sacrum	Lytic	n.a.	(+)	Intermediate/ high	(–)	(+)*	24M
The present case	30/F	Ilium	lytic, destructive/ moth-eaten	Ill-defined	(+)	High/high	(–)	(+)	9Y

M: male, F: female, *Immunohistochemistry, Y: years, M: months, n.a.: not assessed

Additionally, ASPS occasionally develops bone metastasis [1, 2], and thus, possibly presents a hidden primary lesion somewhere in the deep soft tissue when the bone lesion is overt, making it difficult to identify the primary tumor in the soft tissue. For this reason, the other lesions throughout the whole body should be investigated using radiological methods if the bone lesion strongly suggests ASPS histology based on the biopsy. For this reported case, we performed ^{18}F-FDG PET-CT, when the definite diagnosis indicated ASPS after the biopsy. The results of this examination revealed useful information that suspected lesions were present in the left ilium and right kidney. Thus, other soft tissue lesions, that is, doubtful soft tissue primary ASPS, were ruled out. We were able to exclude the possibility of the occurrence of renal tumors via additional enhanced dynamic CT. The SUV_{max} values corresponding to ^{18}F-FDG PET-CT images are generally higher for high-grade sarcomas, such as undifferentiated pleomorphic sarcoma, angiosarcoma, leiomyosarcoma, and rhabdomyosarcoma, than for low-grade sarcomas [13]. However, information on the clinical application of ^{18}F-FDG PET-CT in cases of ASPS is limited. Only three reports on soft-tissue ASPS have demonstrated high SUV_{max} values (5.9, 10.1, and 8.8) [14–16]. Thus, the SUV_{max} values corresponding to ASPSs vary widely and may not always reflect their tumor activity. Further, our findings following PET-CT of the ilium did not show any benign lesion as indicated by the findings of the other radiological examinations. The most important point to be taken into consideration is that the results demonstrated that the ilium is the primary site of ASPS.

Additionally, for the case reported here, it was difficult to conduct definitive diagnosis only with the routine radiological and histopathological approaches for this case. Therefore, additional ^{18}F-FDG PET-CT and dynamic CT of the kidney examinations, and analysis of the gene fusion were performed. These analyses were less invasive, and provided information that allowed us to plan and provide appropriate treatment for this case. Moreover, we would like to emphasize that good oncological and functional outcomes were achieved after curative resection of the local recurrent tumor.

CONCLUSION

In conclusion, in this report, we present the radiological and histopathological findings as well as the clinical outcomes corresponding to an unusual case of ASPS of the ilium. Reverse transcriptase polymerase chain reaction for the identification of tumor-specific gene fusion and ^{18}F-FDG PET-CT for whole body examination in addition to routine radiological and histological studies enable the accurate diagnosis of this tumor, allowing us to provide appropriate treatment for the case here reported.

REFERENCES

1. Jambhekar NA, Ladanyi M. Alveolar soft part sarcoma. In: WHO classification of tumours. WHO Classification of Tumours Editorial Boards, editor. Soft Tissue and bone Tumours. 5ed. Lyon: IARC; 2020. p. 297–99.
2. Goldblum JR, Folpe AL, Weiss SW. Enzinger and Weiss's Soft Tissue Tumors. 6ed. Philadelphia, PA: Elsevier; 2014. p. 1070–79.
3. Temple HT, Scully SP, O'Keefe RJ, Rosenthal DI, Mankin HJ. Clinical presentation of alveolar soft-part sarcoma. Clin Orthop Relat Res 1994;(300):213–8.

4. Durkin RC, Johnston JO. Alveolar soft part sarcoma involving the ilium. A case report. Clin Orthop Relat Res 1999;(359):197–202.
5. Koguchi Y, Yamaguchi T, Yamato M, Osada D, Saotome K. Alveolar soft part sarcoma of bone. J Orthop Sci 2005;10(6):652–5.
6. Park YK, Unni KK, Kim YW, et al. Primary alveolar soft part sarcoma of bone. Histopathology 1999;35(5):411–7.
7. Aisner SC, Beebe K, Blacksin M, Mirani N, Hameed M. Primary alveolar soft part sarcoma of fibula demonstrating ASPL-TFE3 fusion: A case report and review of the literature. Skeletal Radiol 2008;37(11):1047–51.
8. Zhu FP, Lu GM, Zhang LJ, Wang JD, An XJ, Dong YC. Primary alveolar soft part sarcoma of vertebra: A case report and literature review. Skeletal Radiol 2009;38(8):825–9.
9. Yavuz A, Göya C, Bora A, Beyazal M. Primary alveolar soft part sarcoma of the scapula. Case Rep Oncol 2013;6(2):356–61.
10. Zadnik PL, Yurter A, DeLeon R, et al. Alveolar soft-part sarcoma in the sacrum: A case report and review of the literature. Skeletal Radiol 2014;43(1):115–20.
11. Daly BD, Cheung H, Gaines PA, Bradley MJ, Metreweli C. Imaging of alveolar soft part sarcoma. Clin Radiol 1992;46(4):253–6.
12. Crombé A, Brisse HJ, Ledoux P, et al. Alveolar soft-part sarcoma: Can MRI help discriminating from other soft-tissue tumors? A study of the French sarcoma group. Eur Radiol 2019;29(6):3170–82.
13. Macpherson RE, Pratap S, Tyrrell H, et al. Retrospective audit of 957 consecutive ^{18}F-FDG PET-CT scans compared to CT and MRI in 493 patients with different histological subtypes of bone and soft tissue sarcoma. Clin Sarcoma Res 2018;8:9.
14. Montgomery JR, Conrad GR, Sinha P, Absher K. FDG PET of alveolar soft part sarcoma. Clin Nucl Med 2010;35(10):827–9.
15. Sood S, Baheti AD, Shinagare AB, et al. Imaging features of primary and metastatic alveolar soft part sarcoma: Single institute experience in 25 patients. Br J Radiol 2014;87(1036):20130719.
16. Dong A, Wang Y, Cheng C, Zuo C. CT, MRI, and FDG PET/CT in a patient with alveolar soft part sarcoma. Clin Nucl Med 2014;39(3):265–7.

****

Author Contributions
Ikuo Kudawara – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None.

Consent Statement
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Author declares no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.