Clinical Study

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Alveolar Soft Part Sarcoma in Japan: Multi-Institutional Study of 57 Patients from the Japanese Musculoskeletal Oncology Group

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Key Words
Alveolar soft part sarcoma · Bone involvement · Chemotherapy · Local control · Prognosis · Radiotherapy

Abstract

Objective: The clinical features and the management of alveolar soft part sarcoma (ASPS) are not well known. The efficacy of chemotherapy for soft tissue sarcoma, including high-dose ifosfamide and cisplatin, has not been established yet. Some reports suggest ASPS may occur primarily in bone. ***Methods:*** We report on a series of 57 patients with ASPS over 20 years. Their ages ranged from 7 to 75 years (mean 25). ***Results:*** There were 37 females and 20 males. Thirteen lesions (23%) showed bone involvement at the primary site, and 6 of them were diagnosed as bone tumors at presentation. Thirty-seven patients had distant metastases at presentation. Tumor size, bone involvement at the primary site and the presence of metastases at presentation were prognostic indicators ($p < 0.05$). Marginal excision with radiotherapy or wide excision without radiotherapy achieved good local control. Chemotherapy was performed in 47 patients with different regimens. Two patients treated with intra-arterial chemotherapy regimens responded partially, but intravenous chemotherapy with high-dose ifosfamide or cisplatin failed. ***Conclusions:*** ASPS can present primarily as a bone tumor. No advantage of chemotherapy with high-dose ifosfamide or cisplatin could be demonstrated.

Introduction

Alveolar soft part sarcoma (ASPS) is a rare soft tissue neoplasm that accounts for 1% of soft tissue sarcomas. Histologically, a prominent alveolar structure, marked vascular invasion and polygonal cells with granular cytoplasm and prominent nucleoli characterize the tumor [1–6]. Liberman et al. [3] reported a 5-year survival rate of 60%, and only 15% of patients demonstrated long-term disease-free survival. Successful chemotherapy for ASPS has never been reported in a large series, and although ifosfamide and cisplatin have recently been shown to be effective in soft tissue sarcomas [7–10], the efficacy of ifosfamide and cisplatin in ASPS needs further elucidation.

ASPS originates most commonly in the deep soft tissue of the lower extremities [1–6], but it may also arise from the tongue, uterus, stomach, vein and bone [11–18]. To further elaborate on clinical features, prognostic factors and treatment of ASPS, we performed a retrospective study in patients with ASPS who presented to institutes of the Japanese Musculoskeletal Oncology Group (JMOG).

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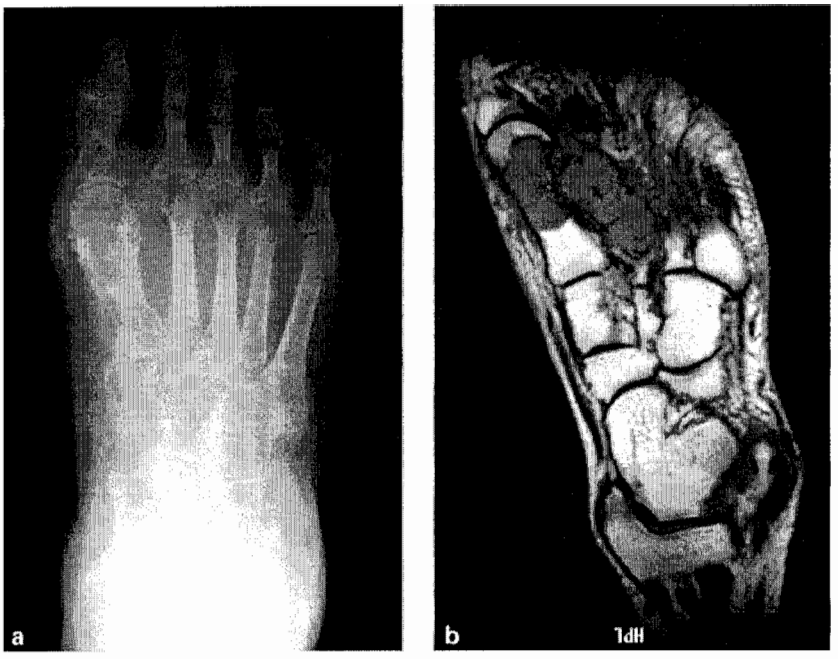


Fig. 1. Radiograph (**a**) and magnetic resonance imaging (**b**) demonstrating ASPS in the foot. The tumor is located in both the intramedullary space and the soft tissue. Bone tumor was diagnosed at presentation.

Table 1. Clinical factors among all patients

Characteristics		Patients	
		n	%
Gender Total	Female	37	65
	Male	20	35
≤ 30 years	Female	29	69
	Male	13	31
>30 years	Female	8	53
	Male	7	47
Tumor size	≤5 cm	22	39
	>5 cm	35	61
Metastasis at presentation	No	20	36
	Yes	37	64
Primary tumor site	Thigh	26	46
	Leg	8	14
	Buttock	7	12
	Upper arm	3	5
	Chest wall	3	5
	Other	10	
Bone involvement at the primary site	Yes	13	23
	No	44	77

The mean age of the patients was 26 years (range: 7–75 years). Mean tumor size was 8.5 cm (range 2–30 cm).

Table 2. Local treatment and recurrence

Initial treatment	Patients	Local recurrence
Biopsy only	8	
Intralesional excision	1	1
Marginal excision	7	4
Marginal excision and radiotherapy	3	0
Wide excision	36	0
Amputation	2	0

Patients and Methods

Patients with ASPS treated at 27 institutions in Japan between 1975 and 2000 were assessed in this study. The patients were registered by members of the JMOG. In each patient, histological diagnosis was confirmed by individual pathologists of the institutions according to previously defined histological criteria [3]. Sixty-six cases were initially registered. Although we did not review all cases histologically, histological reports were all reviewed. Fifty-seven of them had proof of diastase-resistant periodic-acid-Schiff-positive granules, a characteristic feature of ASPS. Nine of them had no proof of the granules and were therefore excluded from this study.

The following definitions were used. Tumor size was defined as the maximum extent determined by pretreatment imaging or physi-

Table 3. Response to preoperative chemotherapy

Type of chemotherapy	Response
Intra-arterial chemotherapy	
cisplatin 80 mg/m^2 i.a. ×3	PR
(cisplatin 120 mg/m^2 i.a., VCR 1.5 mg/m^2, CP 1 g/m^2, DXR 50 mg/m^2/2 days) ×1	PR
(cisplatin 120 mg/m^2, caffeine 4.5 g/m^2/3 days, DXR 60 mg/m^2/2 days) i.a. ×3	NC
IFO 1 g/m^2 i.a. ×1	NC
(cisplatin 100 mg/m^2, DXR 80 mg/m^2) i.a. ×1, IFO 10 g/m^2/5 days i.v. ×2	NC
(VCR 3.5 mg/body, DXR 30 mg/body, mitomycin C 9.4 mg/body) i.a. ×3	PD
(cisplatin 100 mg/m^2 i.a., DXR 50 mg/body i.v.) ×1	PD
Cisplatin-based intravenous chemotherapy	
(cisplatin 1,000 mg/m^2/2 days, pirarubicin 40 mg/m^2/2 days, VP-16 300 mg/m^2/3 days) ×1	NC
(cisplatin 120 mg/m^2, DXR 60 mg/m^2/2 days) ×2	NC
(cisplatin 125 mg/body, DXR 70 mg/body) ×1, (IFO 3 g/body, DXR 80 mg/body, DTIC 1.5 g/body/5 days, VDS 3 mg/body) ×1	NC
(cisplatin 100 mg/m^2, DXR 30 mg/m^2) ×1	NC
(cisplatin 150 mg/body, DXR 70 mg/body) ×1	NC
(cisplatin 100 mg/m^2, DXR 60 mg/m^2) ×2	NC
IFO-based intravenous chemotherapy	
(IFO 4 g/body, DXR 70 mg/body, DTIC 1.5 g/body/5 days, VDS 3 mg/body) ×1	NC
(IFO 4 g/m^2/2 days, DXR 40 mg/m^2/2 days) ×3	NC
(IFO 7.5 g/m^2/3 days, DXR 60 mg/m^2/3 days, DTIC 900 mg/m^2/3 days) ×2	NC
(IFO 7.5 g/m^2/3 days, DXR 60 mg/m^2/3 days, DTIC 900 mg/m^2/3 days) ×2	NC
IFO 14 g/m^2/5 days ×2	NC
DXR-based intravenous chemotherapy	
(DXR 50 mg/m^2, CP 800 mg/m^2, VCR 1.4 mg/m^2) ×2	NC
(DXR 45 mg /body, VCR1.4 mg/body, CP 650 mg/body) ×1	PD
DXR 60 mg/m^2/3 days ×1	PD

IFO = Ifosfamide; DXR = doxourubicin; VCR = vincristine; VDS = vindesine; DTIC = dimethyl-triazeno-imidazole carboxamide; CP = cyclophosphamide; PR = partial response; NC = no change; PD = progressive disease.

cal examination. Complete response was consistent with the complete disappearance of disease. Partial response required a 25–99% reduction in the product of perpendicular greatest dimensions of all measurable lesions. Stable disease indicated <25% reduction or <25% increase in the same parameters. Progressive disease required a >25% increase or the appearance of any new lesions. Survival was estimated by the Kaplan-Meier method. Statistically significant differences in survival curves were evaluated using the Cox-Mantel test.

Results

Clinical Features

Patient characteristics and clinical features are outlined in table 1. Patient ages at diagnosis ranged from 7 to 75 years (mean, 26 years). There were 37 females and 20 males. Of the patients aged under 30 years, there were 29 females and 13 males. Of the patients ≥30 years, there were 8 females and 7 males. Fifty-three patients presented with a soft tissue mass, and lung metastases were the first manifestation of the tumor in 5 patients. Thirty-seven patients (65%) had metastatic disease at the time of the original diagnosis.

The lower extremities were the most common tumor site: in 26 cases it was the thigh (46%) and in 8 cases the leg (14%), but there was a broad spectrum of locations: 7 buttock tumors, 3 chest wall tumors and 3 upper arm tumors. Thirteen lesions demonstrated bone involvement at the primary site, and 6 of them were diagnosed as possible bone tumors at presentation (fig. 1).

Local Treatment of the Primary Tumor and Tumor Recurrence

Initial local treatment and results are shown in table 2. Eight patients underwent only biopsy of the primary tumor without any surgery because of advanced disease.

Table 4. Response to chemotherapy for metastatic tumor

Type of chemotherapy	Response
Cisplatin-based chemotherapy	
(cisplatin 90 mg/m^2, CP 30 mg/kg/3 days, DXR 60 mg/m^2/2 days) ×3	NC
(cisplatin 80 mg/m^2/2 days, DXR 40 mg/m^2/2 days, VP-16 360 mg/m^2/3 days) ×2	NC
(cisplatin100 mg/m^2, DXR 20 mg/m^2) ×2	NC
cisplatin 100 mg/body, VP-16 450 mg/body/3 days	NC
cisplatin 80 mg/m^2 ×3	PD
(cisplatin 120 mg/m^2, DXR 60 mg/m^2/2 days) ×3	PD
cisplatin 80 mg/m^2 ×3	PD
(cisplatin 150 mg/body, VP-16 450 mg/3 days) ×4	PD
(cisplatin 120 mg/m^2, mytomycin C 12 mg/m^2) ×2	PD
IFO-based chemotherapy	
(IFO 7.5 g/m^2/3 days, DXR 60 mg/m^2/2 days) ×2	
IFO 10 g/m^2/5 days ×2	NC
(IFO 3 g/body, DXR 80 mg/body, DTIC 1,500 mg/body/5 days, VDS 3 mg/body) ×5	NC
(IFO 10 g/m^2/5 days, VP-16 250 mg/m^2/5 days) ×2	NC
(IFO 7.5 g/m^2/3 days, DXR 60 mg/m^2/3 days, DTIC 900 mg/m^2/3 days) ×2	PD
IFO 14 g/m^2/5 days ×2	PD
(IFO 7.5 g/m^2/3 days, DXR 60 mg/m^2/3 days, DTIC 900 mg/m^2/3 days) ×1	PD
DXR-based chemotherapy	
DXR 60 mg/m^2 ×3	NC
(DXR 50 mg/m^2, CP 800 mg/m^2, VCR 1.4 mg/m^2) ×2	PD
(DXR 60 mg/m^2/3 days, VCR 2 mg/body, CP 300 mg/m^2) ×1	PD
(DXR 60 mg/m^2/2 days) ×1	PD
(DXR 30 mg/m^2/2 days) ×1	PD
(DXR 50 mg/m^2, VCR 1.5 mg/m^2, CP 500 mg/m^2, DTIC 1 g/m^2/5 days) ×2	PD
(DXR 50 mg/m^2, VCR 1.5 mg/m^2, CP 500 mg/m^2, DTIC 1 g/m^2/5 days) ×2	PD
Others	
(carboplatin 300 mg/body) ×1	PD
methotrexate 300 mg/kg ×3	PD
IFNα 900 ×10,000 U ×20, 1,000 × 10,000 U ×5	PD
IL-2 35 × 10,000 U ×3	PD

IFN = Interferon; IL-2 = interleukin-2; IFO = ifosfamide; DXR = doxourubicin; VCR = vincristine; VDS = vindesine; DTIC = dimethyl-triazeno-imidazole carboxamide; CP = cyclophosphamide; PR = partial response; NC = no change; PD = progressive disease.

One patient had intralesional excision and local recurrence 10 months after surgery. Seven patients underwent marginal excision without radiotherapy, and 4 of them had local recurrence (7–20 months after surgery, mean 14 months). Three patients underwent marginal excision with preoperative or postoperative radiotherapy with 50, 55, and 60 Gy (total dose), respectively. None of them had local recurrence. Thirty-six patients had wide local excision and 2 patients had amputation with a wide surgical margin. All these patients with a wide surgical margin never had local recurrence.

Chemotherapy and Response

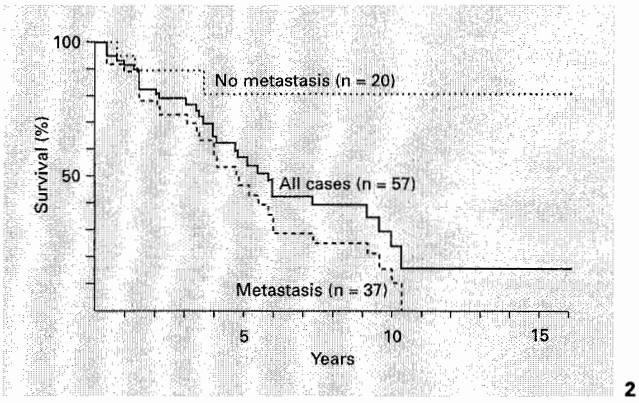
Response to the different types of chemotherapy administered is listed in tables 3 and 4. Twenty-one patients with primary tumors were treated with chemotherapy. Seven patients were given intra-arterial chemotherapy, mainly with cisplatin, resulting in a partial response in 2 patients, and 14 patients received intravenous chemotherapy based on cisplatin, ifosfamide and doxorubicin, but none of these patients showed a clinical response. Evaluable chemotherapy for metastatic tumor, including cisplatin, ifosfamide, doxorubicin, carboplatin, methotrexate, interferon and interleukin-2 regimens, was given

to 27 patients. No clinical response was observed in metastatic patients.

Prognostic Factors

The mean follow-up period in patients still alive was 70 months (range, 6–193 months). The overall survival rates at 5, 10, and 15 years were 56, 23 and 15%, respectively. Metastasis at presentation, tumor size and bone involvement at the primary site were factors indicating a significantly ($p \leq 0.05$) poorer survival. With respect to metastasis at initial diagnosis, the 5-, 10- and 15-year survival rates were 81% for patients without metastasis and 46, 10 and 0% for patients with metastasis (fig. 2). For tumor size, the 5-, 10- and 15-year survival rates were 72, 65, and 65% for patients with tumor diameter ≤5 cm, and 46, 9 and 0% for those >5 cm. There was no difference between patients with tumor diameters of 5–10 cm and >10 cm (fig. 3). Bone involvement at the primary site is also a prognostic indicator. The 5-, 10- and 15-year survival rates for patients without bone involvement were 66, 29, and 19%, and 21, 0 and 0% for patients with bone involvement (fig. 4).

No statistical difference in outcome was evident for age, sex, the site of the primary tumor or chemotherapy.



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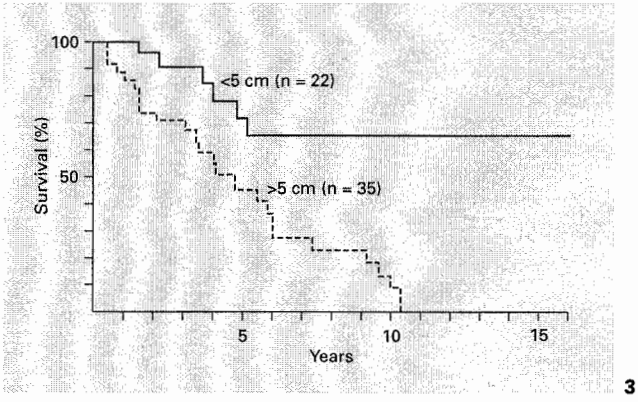

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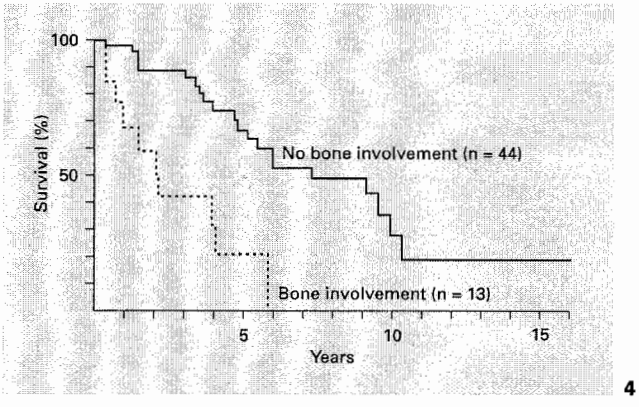

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Fig. 2–4. Overall survival plots of all patients, and patients with and without metastasis at presentation (**2**), patients with tumor diameters ≤5 and >5 cm (**3**) and patients with and without primary bone involvement (**4**).

Discussion

Although the exact nature and tissue type of ASPS remain to be elucidated, the tumor is often located in the skeletal muscles of the thigh [1–6]. Rare locations, where skeletal muscle is absent, are cervix, uterus, orbit, pulmonary vein, stomach and bone [7–18]. Recent molecular analyses demonstrated a specific chromosomal translocation of ASPS: der(17)t(X;17)(p11;q25) [19, 20]. The fusion gene ASPL-TFE3 is also seen in renal carcinomas of children and adolescents [21]. However, these molecular analyses do not provide additional clues to the cell of origin or differentiation [21]. Because ASPS often develops skeletal metastases, the exact site of the primary bone involvement is subject to controversy even in cases of pure intramedullary bone lesions. The present study showed that in 13 of 57 lesions (23%) bone was the primary tumor site, and 6 of them were diagnosed as bone tumors at presentation. These findings indicate that ASPS can present as a bone tumor for clinicians, although its true histogenesis is still unknown.

The mean age of the patients in the current series was 26 years, with a female preponderance in younger patients (<30 years old), as previously reported [3]. The long-term

prognosis of patients without metastasis at presentation is favorable. In this series, overall survival rates of localized ASPS were 81% at 5, 10, and 15 years. However, metastatic ASPS remains a disease with a poor prognosis, with overall survival rates of 46% at 5 years, 10% at 10 years, and 0% at 15 years in this series. This is similar to 20% at 5 years in a series of 48 patients at the MD Anderson Cancer Center [6] and 40% at 5 years and 0% at 10 years in a series of 22 patients at the Memorial Sloan-Kettering Cancer Center [3].

The curative potential of surgery alone has remained unclear. Local recurrences have been reported to occur in up to 20% of patients with ASPS [3]. In this series, none of the 38 patients operated with wide surgical margin developed local recurrence. Of the patients with marginal surgical margins, 4 of 7 patients without radiotherapy developed a local recurrence, while all 3 patients treated with radiotherapy achieved local control. Sherman et al. [22] documented excellent local control in 6 patients with ASPS who received radiotherapy and thus recommended the routine use of radiotherapy. In our opinion, local radiotherapy is not necessary in patients with complete tumor excision, but it is recommended in patients with inadequate surgical margins.

In agreement with previous reports [3, 6], the size of the primary tumor (≤ 5 vs. >5 cm) and presence of metastasis at initial diagnosis were important prognostic indicators in this series. However, there was no difference between patients with a tumor diameter of 5–10 cm and those with tumor >10 cm. Interestingly, primary bone involvement was also an important prognostic indicator in this series. In a study by Park et al. [16], patients with primary ASPS of bone had also a poor prognosis.

Active chemotherapy agents have not been identified for patients with unresected or metastatic ASPS. In the series of Lieberman et al. [3], 22 patients received chemotherapy, and no significant effect was noted. Pappo et al. [4] reported that none of 8 patients who underwent chemotherapy with mainly doxorubicin-based agents responded to the treatment. In a study by Portera et al. [6], 29 patients with ASPS received doxorubicin-based chemotherapy, and only 1 patient responded to therapy. However, the efficacy of ifosfamide and cisplatin, which are recently often used for soft tissue sarcomas [8, 9], has not been elucidated. In this series, 2 of 8 patients who received preoperative, intra-arterial chemotherapy with cisplatin responded partially. Of 44 patients with various regimens of pre- and/or postoperative intravenous chemotherapy including cisplatin, ifosfamide, and doxorubicin-based agents, no patient responded. There are some exceptional reports that advanced patients with ASPS respond to chemotherapy with doxorubicin, methotrexate, thiotepa, cisplatin, interferon, and Chinese herbs [23–27]. On the other hand, spontaneous regression of ASPS has been reported [28]. Portera et al. [6] and Pappo et al. [4] proposed that newly diagnosed patients with unresectable or metastatic ASPS be enrolled in phase I and II chemotherapy trials in an attempt to identify novel active agents. On the basis of the present study, we agreed with trails of those investigational therapies for advanced ASPS.

In conclusion, ASPS can present as bone tumor. Presence of metastasis at presentation, tumor size, and primary bone involvement were prognostic indicators. Marginal excision with radiotherapy or wide excision without radiotherapy achieved good local control. No advantage of chemotherapy including high-dose ifosfamide or cisplatin could be demonstrated.

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