Journal of Bone Oncology 6 (2017) 27–31

Contents lists available at ScienceDirect

Journal of Bone Oncology

journal homepage: www.elsevier.com/locate/jbo

Research paper

Surgical management and outcomes of spinal clear cell sarcoma: A retrospective study of five cases and literature review

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ARTICLE INFO

Keywords:
Clear cell sarcoma
Spine
Surgery
Prognostic factors

ABSTRACT

Purpose: Clear cell sarcoma (CCS) is a rare high-grade malignant tumor accounting for less than 1% of all soft tissue sarcomas. CCS in the spine is much rarer and poorly understood. The objective of our study is to discuss clinical characteristics, surgical management and outcomes of spinal CCS.

Methods: Between October 2011 and December 2014, five patients with spinal CCS underwent surgical treatment in our center. Five cases from literature focusing on the spinal CCS were also reviewed. We retrospectively analyzed clinical data and outcome of all ten patients to present our understanding of spinal CCS.

Results: Three en bloc and two piecemeal resections were performed successfully. The syndromes of all the patients were obviously relieved after operation. The mean follow-up period was 24.4 months (range 10–41 months). Two patients died of lung metastasis 10 and 26 months after operation respectively. One patient was alive with tumor recurrence. Other two patients were alive with no evidence of disease at last follow-up.

Conclusions: Prognosis of spinal CCS tend to be worse than CCS in common sites. En bloc resection could be regarded as the first treatment option. Tumor size, tumor location, preoperative neurological status and resection mode might be the potential prognostic factors of spinal CCS.

1. Introduction

Clear cell sarcoma (CCS), a rare high-grade malignant tumor accounting for less than 1% of all soft tissue sarcomas [1], was first described by Enzinger in 1965 [2]. In 1983, CCS was also named malignant melanoma of the soft parts due to its histological similarities to malignant melanoma, such as the presence of melanin, immunohistochemical staining for melanoma-associated S-100 and HMB-45 [3]. With the discovery of chromosome translocation t(12;22) (q13;q12) which leads to the generation of EWSR1-ATF1 fusion gene, clear cell sarcoma was redefined as a distinct type of tumor [4,5].

CCS is often located in the tendons or aponeuroses of the extremities, especially in the foot and ankle [3,6–10]. However, CCS in the spine is extremely rare and poorly understood. Only a few cases specifically focusing on spinal CCS have been reported [11–15]. As a high-grade malignant tumor, CCS is apt to recur and metastasize, with a local recurrence rate of 84% and late metastasis rate of 63% [16]. Radical excision with negative margins is the best option for treating CCS, but it is difficult to achieve in the spine. Here, we retrospectively analyzed clinical data from our patients along with a review of the literature.

2. Materials and methods

A total of five patients with spinal CCS were diagnosed and treated in our institution between October 2011 and December 2014. All the final pathological diagnoses were confirmed by two independent pathologists according to the following histopathological criteria: 1. Tumors were microscopically characterized by a nested to fascicular growth pattern of fusiform tumor cells and/or a diffuse sheetlike fashion of much plumper polygonal or epithelioid cells. 2. In immunohistochemical study, tumors were positivity for S-100, HMB45, MITF, bcl-2, CD57, Melan A etc. 3. The chromosomal translocation t(12;22) (q13;q12) or the resultant fusion gene EWSR1-ATF1 were detected by RT-PCR or FISH. [17].

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http://dx.doi.org/10.1016/j.jbo.2016.09.002
Received 18 May 2016; Received in revised form 2 August 2016; Accepted 1 September 2016
Available online 17 September 2016

We retrospectively reviewed the hospitalization records, progress notes, surgery information, radiographic images and pathological reports of all patients. This study was approved by the hospital ethics committee and informed consent was obtained from all patients.

Of the five patients analyzed, one (case 4) had been subjected to an incomplete tumor resection in other institution. She was admitted to our center because of tumor recurrence. Other patients were regarded as "intact" cases for they had not received any treatment before admission.

X-ray, computerized tomography (CT) and magnetic resonance imaging (MRI) of the spine were performed in all patients after hospitalization. Tumors were further classified according to the Enneking staging system for all patients and Weinstein-Boriani-Biagini (WBB) classification system for mobile spine based on radiographic findings. Neurologic status was evaluated by Frankel scoring system. One patient obtained her pathological diagnosis after operation in other institution. Three patients received percutaneous needle biopsy in our center. The remaining patient (case 5) refused to have a needle biopsy for fear of possible nerve damage, though we emphasized the significance.

En bloc and piecemeal resections were performed in three and two patients respectively. Patients were followed up at 3, 6, and 12 months after surgery, every 6 months for the next 2 years, and anytime when patients feel uncomfortable. X-ray and/or MRI examination were performed at follow-up. The last status of patients was obtained from office visit or telephone interview.

We also searched the articles related to the spinal CCS using MEDLINE/PubMed as searching engine, and five case reports were reviewed. Then we compared and analyzed both the data in the literature and our own.

3. Results

3.1. Epidemiology and clinical presentation

Our series composed of three men and two women. The age of patients ranged from 20 to 46 years at diagnosis, with a mean age of 27.8 years. The most common symptom was chronic pain, which often radiated to the extremities and could not be relieved by aspirin or acupuncture therapy. Tumor size ranged from 2.4 to 11.9 cm in the maximum diameter, and three of five tumors were larger than 5 cm. Tumor involved posterior elements in two cases, both vertebral body and posterior elements in two cases, and sacrum in one case. Frankel scores were as follows: one patient was Grade C; Grade D and E each were documented in two patients. The clinical data of our patients are listed in Table 1.

3.2. Radiologic studies

The plain radiographs of three patients showed bone destruction. CT scan also demonstrated lytic lesions with irregular soft tissue masses, and inhomogeneous enhancements could be seen after intravenous contrast enhancement. MRI is more sensitive for detecting soft tissue lesions. The tumor lesion was mixed hyperintense on T1WI and hypointense on T2WI, and inhomogeneous enhancement could also be seen on MRI enhancement scan (Fig. 1).

3.3. Treatment

The whole operation process comprised tumor excision, decompression of the spinal cord, reconstruction and stabilization of the spine. Intraoperative frozen section examination was performed in all five cases. Posterior approach was conducted in four patients, while the other one (case 4) whose lesion involved cervical vertebra (zone 1–9 of WBB system) used a combined anterior-posterior approach. En bloc resection was performed on three cases (case 1, 2 and 3), other two

Table 1
Clinical data of patients with spinal CCS in our institution.

No.	Age (y)/ Sex	Location	Staging*	Symptoms	F-S pre	Resection mode	Tumor size (cm)	F-S post	Adjunctive therapy	Local recurrence/Metastasis	Follow-up (month)	Last status
1	46/M	Sacrum	$G_2T_2M_0$	Radicular pain and dysuria	D	En bloc resection	8.2	D	Bisphosphonate	Local recurrence at 12 months	13	AWD
2	23/M	T10–11	$G_2T_2M_0$ 1–2/A-C	No symptom**	E	En bloc resection	2.4	E	–	–	32	NED
3	26/M	T6–8	$G_2T_2M_0$ 10–12/A-D	Palpable mass	E	En bloc resection	3.8	E	–	–	41	NED
4	20/F	C3–5	$G_2T_2M_1$ 1–9/A-D	Pain and weakness of upper extremities	C	Piecemeal resection	5.3	E	Chemotherapy, radiotherapy and bisphosphonate	Lung and neck lymph nodes metastasis before surgery in our center***	10	Dead
5	24/F	L1–3	$G_2T_2M_0$ 3–8/A-D	Pain and radicular pain	D	Piecemeal resection	11.9	E	Radiotherapy and bisphosphonate	Local recurrence at 18 months; lung metastasis at 24 months	26	Dead

M, male; F, female; F-S, Frankel score; pre, preoperation; post, postoperation; NED, no evidence of disease; AWD, alive with disease; NI, no information.

* Enneking staging was used in all cases, Weinstein-Boriani-Biagini staging was used in mobile spine.

** Tumor was found in health checks.

*** This patient experienced tumor recurrence and metastasis to lung and neck lymph nodes two years after her initial operation in other institution, and she then had an operation of cervical spine tumor in our department due to unrelieved pain and neurologic deficit.