Clinical Features, Prognostic Factors, and Treatment Strategy of Osteoblastoma-like Osteosarcoma: A Case Report and Review of the Literature

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Case report

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Abstract

Background

Osteoblastoma-like osteosarcoma is a very rare tumor and the diagnosis and the treatment can be challenging and controversial. Herein, we presented a case occurring in the occipital bone firstly and summarized all of the previously published cases to delineate the clinical characteristics, prognostic factors and treatment strategy of osteoblastoma-like osteosarcoma.

Case presentation:

A 34-year-old man presented with a headache with bitter and dull for 6 months, underwent a suboccipital posterior median approach to remove the lesion without intra-operative complications. Computerized tomography revealed osteogenic bone destruction on the occipital protuberance involving inner and outer table of the skull. Preoperative Magnetic Resonance Venogram revealed that the tumor invades sinus convergence, disappeared sinus convergence, narrowed bilateral transverse sinus, and abundant tumor blood vessels. Re-examination on the day after operation revealed that the post-operative changes of the occipital bone and the tumor were resected completely. The pathologic examination identified the Osteoblastoma-like osteosarcoma. The postoperative course was uneventful. Patient was discharged home 7 days after operation. By four months, re-examination Magnetic Resonance Imaging scan revealed that there was no evidence of recurrence and was not leaded to any problem.

Conclusion:

For patients with presumed osteoblastoma-like osteosarcoma, gross total resection should be recommended. If tolerable, radiotherapy or chemotherapy could be an alternative treatment modality. Patients with areas of conventional osteosarcoma had poor progression-free survival and tumors with metastasis had a poor overall survival. The lung was the most frequently involved regions of metastasis. Trial registration: CRD, CRD42019121880. Registered 13 February 2019, https://www.crd.york.ac.uk/PROSPERO

Background

Osteoblastoma-like osteosarcoma (OBLOS) is a benign pathology, which is a primary bone-forming tumor [1–3]. OBLOS is a rare variant of osteosarcoma accounting for approximately 1% of cases overall [4, 5]. OBLOS is a malignant lesion, and without appropriate treatment, which has a high risk of mortality. In the current WHO classification,it is classified within the group of conventional osteosarcomas [6]. Although differentiating OBLOS from osteoblastoma and conventional osteosarcoma can be challenging and controversial, there are enough knowledge about these three kinds of tumors are distinct ones [3, 5, 7–9].

The most of reports were case reports [1, 8, 10, 11] or case series and most of those, without statistical analyses, were less convincing [2, 12], because of this, little clinical features and outcome patterns were

acknowledged. Herein, we reported 1 case from our center and an additional 59 cases from published studies to describe the clinical characteristics of this tumor and to evaluate the prognostic factors and treatment strategy of OBLOS.

Case Presentation

The patient was a 34-year old man with a headache with bitter and dull for 6 months, and the pain could be alleviated by analgesics. The patient did not have any other associated symptom, and had no other comorbidities, or medication use. Previous medical history and physical examination were unremarkable except a trauma on the occipital bone in 2012. The patient did not have any symptoms except the occipital pain and the pain went away shortly. The patient did not take any interventions on the trauma. Computerized tomography (CT) revealed osteogenic bone destruction on the occipital protuberance involving inner and outer table of the skull. The whole presentation was elliptical mixed high-density shadow and plaque-like bone density (Fig. 1a, b). The size was 37*43*54 mm. Preoperative Magnetic Resonance Venogram (MRV) revealed that the tumor invades sinus convergence, disappeared sinus convergence, narrowed bilateral transverse sinus, and abundant tumor blood vessels (Fig. 1c, d).

The patient underwent a suboccipital posterior median approach to remove the lesion without intra-operative complications. Re-examination on the day after operation revealed that the post-operative changes of the occipital bone and the tumor were resected completely (Fig. 1e). The postoperative course was uneventful. Patient was discharged home 7 days after operation. The pathologic examination identified the OBLOS. Mature osteoblasts, osteoclasts and neoplastic bone accompanied by calcification, fibrous and vascular tissues could be seen between bone tissues; focal infiltration of host bone growth; aging and fusion of some new bone; epithelial differentiation of focal osteoblasts; broad eosinophilic cytoplasm; irregular components of immature hypertrophic bone sorghum and hypertrophic osteoblastoma-like tumor cells with abnormalities, clear nucleoli and mitotic figures (Fig. 1f). Gamma knife was given for treatment as the adjuvant radiotherapy. By four months, re-examination Magnetic Resonance Imaging (MRI) scan revealed that there was no evidence of recurrence and was not leaded to any problem. (Fig. 1g, h).

His family was informed of these conditions and signed a consent form prior to the operation. After the patient was intubated and anesthetized, the patient was lying on the left side and skin preparation and draping routinely. After preparing the head frame, the surgeons cut the skin and subcutaneous tissue according to the suboccipital posterior median approach incision and separated along the midline to reach the occipital bone. The retractor retracted neck muscles on both sides. The lesion was found to be tough inconsistency with the unclear surgical plane, adhering to muscles and dura, and extremely vascularized. The occipital bone around the tumor was drilled and carefully separated along the approximate border of the tumor. The tumor and the tumor tissue that was attached to the dura mater and muscle were completely excised. The peripheral nerves and blood vessels were well protected and hemostasis was completed. The dura related tumor tissue was removed completely, then artificial dura

was used for dura repair. Cranioplasty with a titanium plate was completed. The muscles, fascia, subcutaneous and scalp were sutured. The intraoperative total bleeding volume was 800 ml.

To best of our knowledge, fifteen- nine cases were reported to have OBLOS from 3 series or case reports (Supplemental table 1). We took 57 cases into analysis because of the other cases without the detailed clinical information. GTR was defined as the extent of resection is amputation, en-bloc resection and resection with wide margins. Progression-free survival (PFS) was defined as the duration from the diagnostic date to the date of death or recurrence, whichever came first. Overall survival (OS) was defined as the duration from the diagnostic date to the date of death or the date when this study ended, whichever came first. Necrosis was defined as the tumor had the areas of necrosis. The extent of surgical resection was evaluated as Gross total resection (GTR) or non-GTR (NGTR). Adverse factors for PFS and OS that were statistically significant in univariate analysis were further evaluated by multivariate Cox proportion analysis. The log-rank (Mantel-Cox) test and a Cox regression model were used to perform univariate and multivariate analysis, respectively. According to the treatments administered in each case, multimodal regimens were classified as follows: Regimen 1, surgery alone; Regimen 2, surgery plus radiotherapy (RT); Regimen 3, surgery plus chemotherapy (CMT); Regimen 4, surgery plus RT and CMT. These treatment regimens were further sub-grouped by the extent of resection to identify the optimal treatment model compared to the PFS and the OS of the whole cohort (n = 57) that was designed as a baseline. We performed pairwise comparisons among each combined modality by univariate Cox analysis (log-rank test). Statistical analysis was performed using R version 3.5.1 (R Foundation for Statistical,2018). The R package is survival (Terry M. Therneau, Patricia M. Grambsch), survminer (Alboukadel Kassambara and Marcin Kosinski), and ggplot2 (Hadley Wickham). A P value < 0.05 was considered to be statistically significant.

Among the whole cohort (n = 57, 56.1% males), patients' age ranged from 6 years to 58 years and the median and mean age were 25 years and 26.1 years (Table 1). This lesion often developed in the 1st the 2nd decade of life (Fig. 2a). The tumor size ranged from 1 cm to 10 cm (5 cm, median;4.87 cm, mean). Nineteen (86.4%) patients suffered from pain, one condition aggravated when move and one combined with weakness and numbness,which represented the most common presentation. The second (n = 10, 47.6% patients) and third (n = 7, 33.3% patients) most presentation were the swelling and tenderness. One each patient suffered from restricted movement, bilateral leg numb, 9.1 kg weight loss, ear drainage and blocking, obstruction of the nose with pus and tiredness of legs. The median duration from the onset of symptoms to diagnosis was 2 (range 1–36) months and the mean duration was 6.43 (9.07) months. The tibia area was the most frequently involved regions, found in 15 (26.4%) patients. Overall, 16 (27.6%) tumors were located in the tibia bone. Ten tumors involved the vertebra, six the femur, four the calcaneal and humerus, three the ilium and fibula, two the cuboid, metatarsus, maxilla and radius, and one each the ethmoid, scapula, temporal, hip, hand and occipital. Necrosis was found in 11 (55.0%) patients and areas of conventional osteosarcoma (AOS) was found in 11 (44.0%) patients. Nine (15.8%) patients accepted radiotherapy and fourteen (24.6%) patients accepted chemotherapy. Seventeen (29.8%) exhibited recurrence and fourteen (24.6%) had evidence of metastasis. The lung was the most frequently involved regions, found in 13 (86.7%) patients. Three involved the bone and one involved the brain. Notably, 11 of

the 57 patients had areas of conventional osteosarcoma. Accordingly, GTR alone, NGTR alone, GTR plus RT, GTR plus CMT, NGTR plus RT, NGTR plus CMT and NGTR plus RT and CMT were used in 15 (26.3%), 18 (31.6%), 2 (3.5%), 6 (10.5%), 4 (7.0%), 6 (10.5%), and 3 (5.3%) patients, respectively. The median follow-up of the entire group was 39 (mean 84.6) months. For the entire group, 6 (10.5%) patients suffered from disease progression with a 3-year PFS of 39.5% and a 5-year PFS of 25.6%, respectively; tumor recurrence was observed in 17 (29.8%) patients with a 3-year OS of 62.7% and a 5-year OS of 49.4%, respectively; 14 (24.6%) patients developed metastasis with a 3-year OS of 46.4% and a 5-year OS of 37.1%, respectively.

Table 1
Demographics and clinical characteristics

Variables	Value (%)
Age, years	
Mean (SD)	26.1 (11.7)
Median (range)	25.0 (6.00–58)
Gender	
F	24 (42.1%)
M	32 (56.1%)
Tumor location	
Appendage	42 (73.7%)
Torso	10 (17.5%)
Skull	5 (8.8%)
Necrosis	
Yes	11 (19.3%)
No	9 (15.8%)
Areas of conventional osteosarcoma	
Yes	11 (19.3%)
No	14 (24.6%)
Chemotherapy	
Yes	14 (24.6%)
No	40 (70.2%)
Radiotherapy	
Yes	9 (15.8%)
No	45 (78.9%)
Extent of resection	
GTR	23 (40.4%)
NGTR	31 (54.4%)
Unknown	3 (5.3%)
Recurrence	

Variables	Value (%)
Yes	17 (29.8%)
No	36 (63.2%)
Metastasis	
Yes	14 (24.6%)
No	39 (68.4%)
Treatment regime	
Regimen1	33 (57.9%)
Regimen2	6 (10.5%)
Regimen 3	12 (21.1%)
Regimen4	3 (5.3%)
Unknown	3 (5.3%)
PFS, month	
Mean (SD)	61.4 (112)
Median (range)	26.0 (2.00, 552)
1-year PFS	74.4%
3-year PFS	39.5%
5-year PFS	25.6%
OS, month	
Mean (SD)	84.6 (116)
Median (range)	39.0 (8.00, 552)
1-year OS	96.2%
3-year OS	76.8%
5-year OS	70.5%

The summary of potential factors that affected outcomes was demonstrated in Fig. 3 and Fig. 4. Kaplan–Meier analysis showed that tumor location (P = 0.001), no AOS (P = 0.035) and GTR (P = 0.046) were significantly favorable factors. Multivariate analysis confirmed that AOS (HR = 3.61, 95%CI = 1.19–10.90; p = 0.023) could independently affect PFS (Fig. 2b and Fig. 3). Though tumor location (p = 0.006), GTR (p = 0.007), recurrence (P = 0.011) and metastasis (P = 0.001) had a significant impact on OS in the

univariate analysis, only the metastasis was an independent factor for OS (HR = 4.06, 95%CI = 1.101–15.000; p = 0.035) (Fig. 2c and Fig. 4).

Four modalities were assessed (Table 2 and Table 3). Without regard to surgical resection (n = 57), the differences in PFS between regimen 1 and regime 3 were statistically significant, Regime 3 presented worse PFS, which had a significantly low 5-year PFS of 10.0% (Table 2). The estimated median survival time is 19.5 months and the confidence interval has not yet been reached in this group (Table 2). And, the differences in OS between regimen 1 and regime 2 were statistically significant, Regime 2 presented worse OS, which had a significantly low 5-year OS of 27.8%, and the estimated median survival time has not yet been reached in this group (Table 3). When the extent of surgery was considered (n = 57), some cases without information on the extent of resection were excluded, which might decrease the statistical power of this analysis. These four regimens were further sub-grouped (Table 4 and Table 5). The median PFS of this entire subgroup was 27 (17–42) months. The median OS of this entire subgroup was not achieved. The whole cohort was designed as a baseline. Compared to the baseline, patients receiving NGTR + RT or NGTR + CMT had significantly poor PFS (Table 4); Patients receiving RT follow NGTR had a significantly poor OS (Table 5).

Table 2
Clinical distribution of the four regimens for progression-free survival

	Death/total	EMST (95%CI)	1-year	3-year	5-year	HR (95% CI)	P value
Regime1	3/29	38.5(19–96)	80.8%	53.8%	34.6%	Ref.	
Regime2	2/6	14	50.0%	25.0%	25.0%	1.43(0.49–4.18)	0.5134
Regime3	1/11	19.5	60.0%	10.0%	10.0%	2.54(1.15–5.57)	0.020§
Regime4	0/3	45	100%	33.3%	0	1.42(0.42–4.82)	0.579
§P < 0.05 indicates statistical significance							

Table 3
Clinical distribution of the four regimens for overall survival

	Death/total	EMST (95%CI)	1-year	3-year	5-year	HR (95%CI)	P value
Regime1	8/33	-	97.0%	82.1%	77.8%	Ref.	
Regime2	2/11	-	83.3%	27.8%	27.8%	3.93(1.03–15.02)	0.045§
Regime3	3/6	34	100%	76.2%	76.2%	1.04(0.22-5.0)	0.960
Regime4	1/3	50	100%	100%	50.0%	1.44(0.18–11.73)	0.733
§P < 0.05 indicates statistical significance							

Table 4
PFS of different stratified treatments and comparisons to entire cohort (baseline)

	Death/total	EMST (95%CI)	1-year PFS	3-year PFS	5-year PFS	HR (95%CI)	P value
Entire group	6/49	27(17–42)	74.4%	39.5%	25.6%	Ref.	
Subgroups of regimen 1							
GTR alone	1/14	39	100%	61.5%	30.8%	0.65(0.35–1.22)	0.183
NGTR alone	2/15	19	61.5%	46.2%	38.5%	1.06(0.57–1.99)	0.856
Subgroups of regimen 2							
GTR + RT	0/2	95	100%	50%	50%	0.63(0.15–2.63)	0.529
NGTR + RT	2/4	7.5	-	-	-	8.16(1.84–36.19)	0.006§
Subgroups of regimen 3							
GTR + CMT	0/5	28	100%	20.0%	20.0%	1.29(0.51–3.31)	0.591
NGTR + CMT	1/6	8	20.0%	-	-	5.36(2.00-14.35)	< 0.001§
Regimen 4	0/3	30	100%	33.3%	0	1.14(0.35–3.71)	0.830

§P < 0.05 indicates statistical significance

Table 5
OS of different stratified treatments and comparisons to entire cohort (baseline)

	Death/total	EMST (95%CI)	1-year OS	3-year OS	5-year OS	HR (95%CI)	P value
Entire group	14/53	-	96.2%	76.8%	70.5%	Ref.	
Subgroups of regimen 1							
GTR alone	1/15		93.3%	93.3%	93.3%	0.87 (0.47–1.62)	0.664
NGTR alone	7/18		100%	75.6%	69.3%	0.75 (0.38–1.47)	0.401
Subgroups of regimen 2							
GTR + RT	0/2		100%	100%	100%	0.90 (0.22–3.75)	0.883
NGTR + RT	3/4	20	75.0%	-	-	31.48 (3.18-311.56)	0.003§
Subgroups of regimen 3							
GTR + CMT	0/5		100%	100%	100%	2.48 (0.95–6.48)	0.065
NGTR + CMT	2/6		100%	60.0%	60.0%	1.39 (0.49–3.94)	0.534
Regimen 4	1/3	50	100%	100%	50.0%	1.79 (0.42–7.53)	0.430

§P < 0.05 indicates statistical significance

Discussion And Conclusion

Due to the contributions of previous studies [3, 5, 12], OBLOS were increasingly recognized. The patient from our center was the first case involving the occipital bone. Briefly, we found that AOS could affect PFS while metastasis could impact OS. A worse PFS was observed in the subgroup of patients treated with surgery + CMT, NGTR + RT and NGTR + CMT, while a worse OS was observed in the subgroup of patients treated with surgery + RT or NGTR + RT, finally the use of NGTR + RT could result in poor PFS and OS.

It was reported that OBLOS affected more male than female patients [3, 7]. A study that mainly included OBLOS reported a male to female ratio of 1.125:1 (9/8) [3]. One patient from our center and the pooled 56 patients also showed a minor male predominance (56.1% and 42.1%, respectively) for OBLOS, which

corresponded with the findings reported by Gambarotti et al [7]. This neoplasm often developed in the 2nd half of the 2nd decade of life [13]. Similar results were also observed in other studies [3, 7, 13, 14]. However, 1 patient from our center and the pooled 56 patients showed this lesion often developed in the 1st the 2nd decade of life (Fig. 2a). The median symptom duration of 2 months indicated a relatively aggressive disease course. The tumor occurred in various sites of the skeleton. The most common location was the tibia in fifteen cases, followed by the vertebra, ten cases. Small bones of the skull, face and foot were also involved, that were relatively uncommon locations of conventional osteosarcoma (Supplemental table 1).

Kathrin et al. believed that OBLOS was a low-grade lesion, similar results were reported by Luigia et al. and Bertoni et al [5, 11, 15]. However, we agreed with the report that OBLOS was a kind of high-grade neoplasm [5, 8]. The report that recurrence was common, and metastases occurred in some patients [3, 5] while our study verified that OBLOS had a high risk of recurrence (n = 17, 29.8%) and metastasis(n = 14, 24.6%) (Table 1). OBLOS was a kind of malignant lesion because of the high risk of recurrence and distant metastasis.

It was usually easy to differentiate from conventional osteosarcoma, which usually had a short clinical presentation, more aggressive radiographic findings, and high-grade histologic features [16]. However, there are unusual variants of osteosarcoma that are so histologically similar to osteoblastomas that there may be some diagnostic confusions. The symptoms at presentation may not raise suspicion about such a specific disease. However, progressive symptoms and new-onset clinical findings after an initial diagnosis of osteoblastoma must be alarming for the clinicians in favor of OBLOS.

OBLOS could resemble osteoblastoma histologically[1, 7]. The pathological findings included areas resembling classic osteoblastoma, sheets of cells without bone production, variable quantities of lace-like osteoid, rounded and ovoid nuclei with or without nuclear atypia, with or without prominent nucleoli and admixed spindled stromal cells, the proliferation of osteoblasts forming osteoid or bone and fibrovascular tissue between its bony trabeculae. OBLOS had a low mitotic rate. OBLOS characterized histologically by the presence of neoplastic cells cytologically identical to osteoblasts set in a trabecular osteoid matrix mimicking that of osteoblastoma [7]. All these histological features overlapped with those expected in osteoblastomas. Tumor permeation of host bone was mentioned as the primary histological feature to differentiate OBLOS from osteoblastoma [5]. Our report was consistent with this (Fig. 1f).

OBLOS shared radiological features with osteoblastoma and osteosarcoma. However, OBLOS lesions usually lack the soft-tissue component when compare to typical osteosarcoma in MRI. Radiologically the lesions were usually lytic with poorly defined borders and cortical destruction. OBLOS can be high uptake on radionuclide bone imaging [1, 2, 8, 11] and radiolucent or radio-dense [2, 5, 11]. Ossifications, a sclerotic nidus, lytic, expansile pattern and periosteal reaction can be present inside the lesion [2, 5, 8]. Mild periosteal new bone formation can be present [5]. MRI can show isointense on T1 images [1], hyperintense, edema, sclerosis or permeability on T2 images [1, 2, 4, 8] and contrast enhancement [1, 2, 8, 11]. Gambarotti et al. reported that the borders of the tumor can be poorly defined with cortical

destruction or well defined and distinct, even featuring a sclerotic rim [7, 8]. This sign was also observed in CT scans on our case ((Fig. 1a, b). The diagnosis was entirely dependent on the demonstration of peripheral permeative growth by the tumor, characterized by marrow permeation between preexisting trabeculae and trabecular appositional growth.

Bertoni et al. reported four tumor recurrence, five metastasis and four deaths over a three-year follow-up (17 cases) while Gambarotti et al. reported six tumor recurrences, five metastasis and four deaths over a mean follow-up was 128 months (range 9-552 months) (15 cases) [3, 7]. By contrast, the median follow-up in our pooled study was 39 months, and the 1-year PFS, 3-year PFS, 5-year PFS, 1-year OS, 3-year OS and 5-year OS were 74.4%, 39.5%, 25.6%, 96.2%, 76.8% and 70.5%, respectively (Table 1). The observation that OBLOS tended to develop with recurrence and metastasis frequently. The lung was the most frequent metastasis region. When Metastasis development occurred, most patients died soon. Our study proved that the metastasis could independently affect the OS (Fig. 4).

Studies on prognostic factors with exclusive emphasis on OBLOS were scattered. Gambarotti et al. reported that the presence of AOS seemed to be the critical parameter to differentiate these tumors and to predict an aggressive outcome [7]. Our study verified this argument that the presence of areas of conventional osteosarcoma independently predicted poor PFS (Fig. 3).

Surgical intervention was essential to make an accurate diagnosis and relieve symptoms by reducing the mass effect. The argument that GTR could yield good outcomes was less persuasive because our study failed to confirm the efficacy of GTR in patient outcomes. Univariate analysis revealed that the GTR could predict better PFS and OS. As the case from our institute, the patient got a GTR surgery, and the patient did not have any sign of recurrence or metastasis. Therefore, if the lesion was easily accessible, e.g. appendage bone, GTR was still advocated; otherwise, NGTR or biopsy rather than GTR should be considered to avoid the potential complications, such as malignant esophageal fistula or even death [3]. Discouragingly, the role of RT in the treatment of OBLOS was questionable. In this pooled analysis, we found that the 5-year OS of patients treated with Regime 2 was significantly shorter than that of patients treated with Regime 1 (27.8% vs 77.8%, $p = 0.020$), and a trend of poor PFS and OS were also found in the NGTR + RT subgroup (Table 3, Table 4 and Table 5). Although a trend of better PFS and OS was found in the GTR + RT group, the difference was not significant. And, our case had a good outcome after the adjuvant radiotherapy. We may conclude that the radiotherapy may be an optimal treatment modality. However, which radiotherapy and the dose of the radiotherapy were unknown.

A longer 5-year of PFS (34.6%) was reported in our studies consisting of patients with surgery alone than the patients treated with surgery plus CMT (Table 2). When compared to the entire group, a trend of poor PFS was also found in the NGTR + CMT subgroup (Table 4). Similarly, though trends of higher 5-year OS were observed in patients treated with surgery plus CMT and the GTR + CMT, the difference was not significant. We could not conclude that the CMT is useful for this kind of tumor. And, compared to the whole cohort, GTR + CMT predicted a better OS thought the difference is not significant.

We agreed with the recommendation that patients with OBLOS should be considered for wide resection [5, 11]. Unlike the treatment modalities of conventional osteosarcoma is surgical removal of all detectable tumor sites and chemotherapy [17], we did not have the enough grounds to confirm that the GTR followed by CMT could be recommended. However,if tolerable, RT or CMT could be alternative treatment modalities.

Herein, we presented a rare case and summarized all of the previously published cases which could substantially increase our experience to diagnosis and treat this rare condition. OBLOS is a rare lesion with a high preoperative misdiagnosis rate. Patients with AOS had poor PFS and metastasis predicted a poor OS. The lung was the most frequently involved regions of metastasis. The patients with OBLOS should be considered for gross total resection. If tolerable, gross total resection followed radiotherapy or chemotherapy could be an alternative treatment modality. Our findings need to be verified in the future.

Abbreviations

AM
aneuploidy mitosis;
AOOS
areas of abundant osteoid
AOS
areas of conventional osteosarcoma
Bilal
bilateral
CT
Computerized tomography
CMT
chemotherapy
EMST
estimated median survival time
GTR
Gross total resection
HN
hyperchromatic nuclei
INS
increased nuclear size
L
left
MRV
Magnetic Resonance Venogram
MRI
Magnetic Resonance Imaging

NGTR
non- Gross total resection
OBLOS
Osteoblastoma-like osteosarcoma
OS
Overall survival
PFS
Progression-free survival
R
right
Regimen1
surgery alone;
Regimen2
surgery plus radiotherapy;
Regimen 3
surgery plus chemotherapy;
Regimen4
surgery plus chemotherapy and radiotherapy.
RT
radiotherapy

Declarations

Ethics approval and consent to participate

The ethics committee of Beijing Tiantan Hospital approved this study. Informed consent was obtained from all participants.

Consent to publish

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

Availability of data and materials

All data generated or analysed during this study are included in this published article and its supplementary information files.

Competing interests

The author reports no conflicts of interest in this work.

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Authors' Contributions

Conception and design: all authors.

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Figures

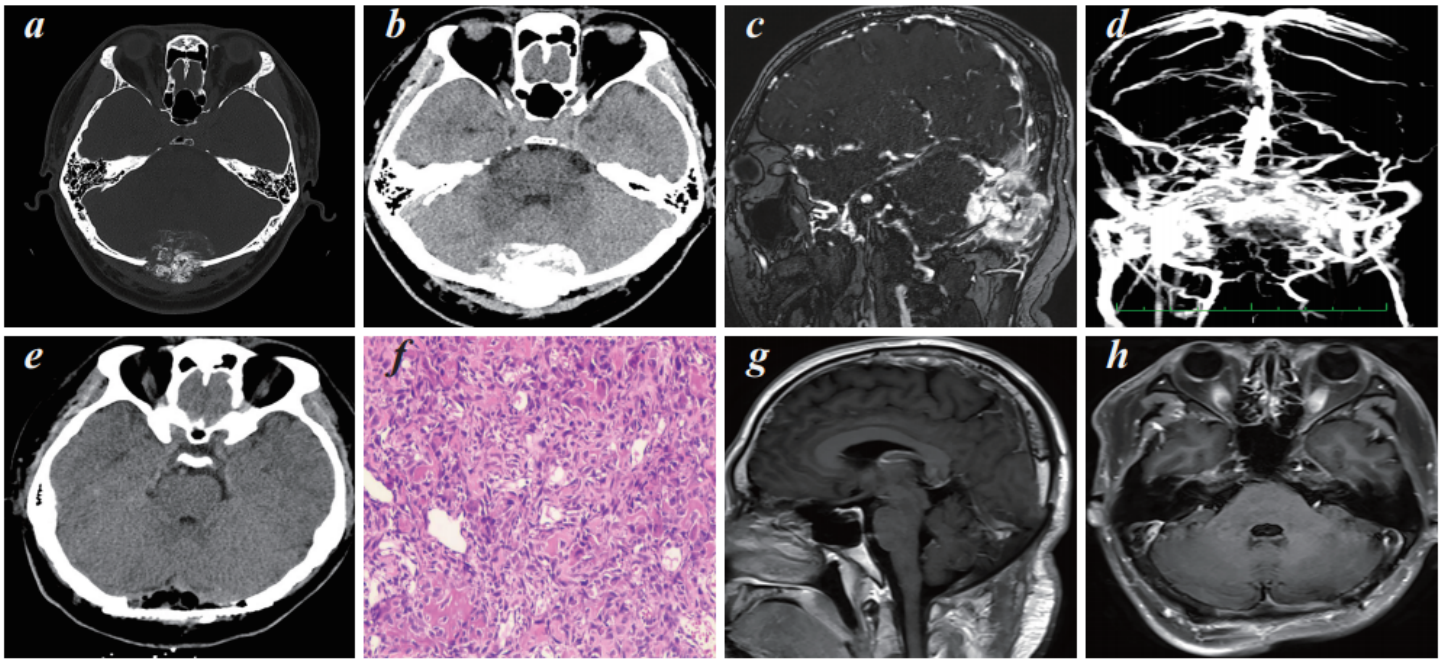

Figure 1

a–b: Preoperative CT showed a occipital lesion that had an osteogenic bone destruction (axial, bone window), the elliptical mixed high-density shadow and plaque-like bone density(axial, brain window); c-d: Preoperative MRV revealed that the tumor invades sinus convergence, disappeared the sinus convergence, narrowed bilateral transverse sinus narrow, and abundant tumor blood vessels; e: Re-examination on the day after operation revealed that the post-operative changes of the occipital bone and the tumor was resected completely; f: Staining of OBLOS with H & E revealed immature hypertrophic bone sorghum, hypertrophic osteoblastoma-like tumor cells with abnormalities, focal infiltration of host bone growth, clear nucleoli, and mitotic figures. Original magnification ×100. Figure is available in color online only; g-h: The axial and sagittal position of the MRI showed that there is no recurrence evidence.

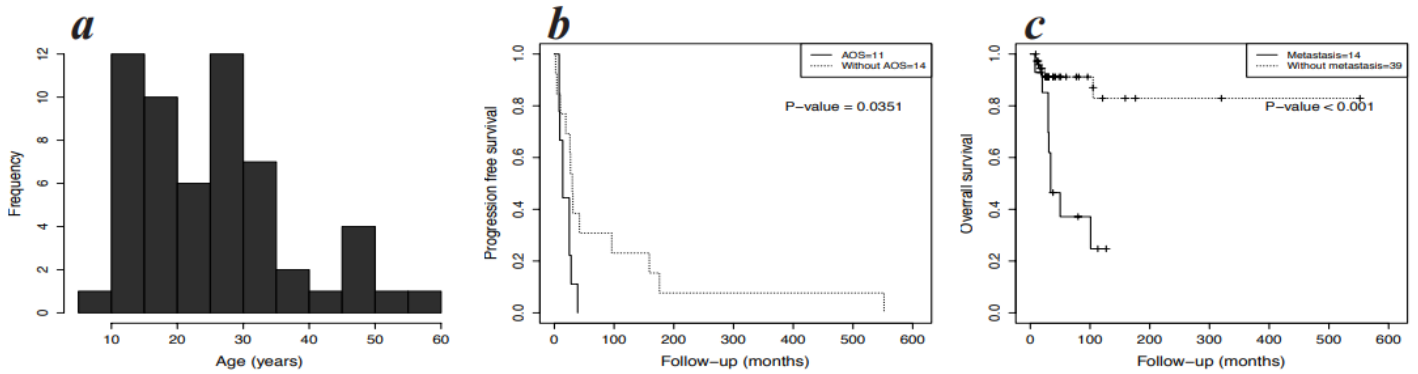

Figure 2

a: The distribution of age; Kaplan–Meier method showed that b patients with areas of conventional osteosarcoma exhibited poor PFS than its counterpart; c The patients with the metastasis had poor OS.

Risk factors	Death/total	EMST(95%CI)	1-year PFS	3-year PFS	5-year PFS	HR(95%CI)	P value
Entire	6/49	27(17-42)	74.4%	39.5%	25.6%		
Gender							
Female	4/23	29(15-60)	73.7%	36.8%	15.8%		
Male	2/25	27(17-159)	73.9%	43.5%	34.8%		
Age >= 34 years							
Yes	0/8	54.5	62.5%	50.0%	50.0%		
No	6/41	27(19-42)	77.1%	37.1%	20.0%		
Tumor location							
Torso	2/9	11	28.6%	14.3%		Ref.	
Appendage	4/35	31(26-81)	83.9%	48.4%	35.5%	1.03(0.30-3.49)	0.963
Skull	0/5	17	80.0%	20.0%	0	2.98(0.24-36.36)	0.393
AOS							
Yes	1/10	14	66.7%	11.1%		3.61(1.19-10.93)	0.023
No	0/13	30	76.9%	38.5%	30.8%	Ref.	
Necrosis							
Yes	1/11	25	70.0%	10.0%	10.0%		
No	0/8	30.5	62.5%	50.0%	37.5%		
Surgery							
GTR	1/21	34.5(27-81)	1	50.0%	30.0%	Ref.	
NGTR	5/28	17(9-60)	52.2%	30.4%	21.7%	2.95(0.98-8.90)	0.055
RT							
Yes	2/9	25	71.4%	28.6%	14.3%		
No	4/40	27.5(7-42)	75.0%	41.7%	27.8%		
CMT							
Yes	1/13	25.5	75.0%	16.7%	8.3%		
No	5/36	29(15-81)	74.2%	48.4%	32.3%		

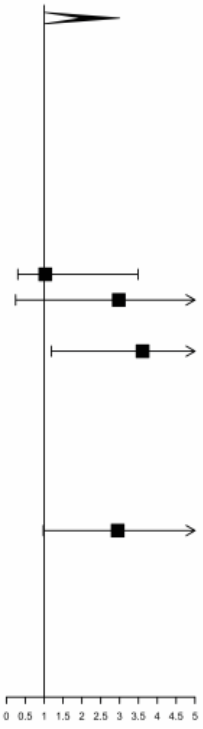

Figure 3

Multivariate Cox analyses was used to evaluate the adverse factors for PFS. Black squares indicate the hazard ratio (HR) values, error bars represent the 95% confidence interval (CI).

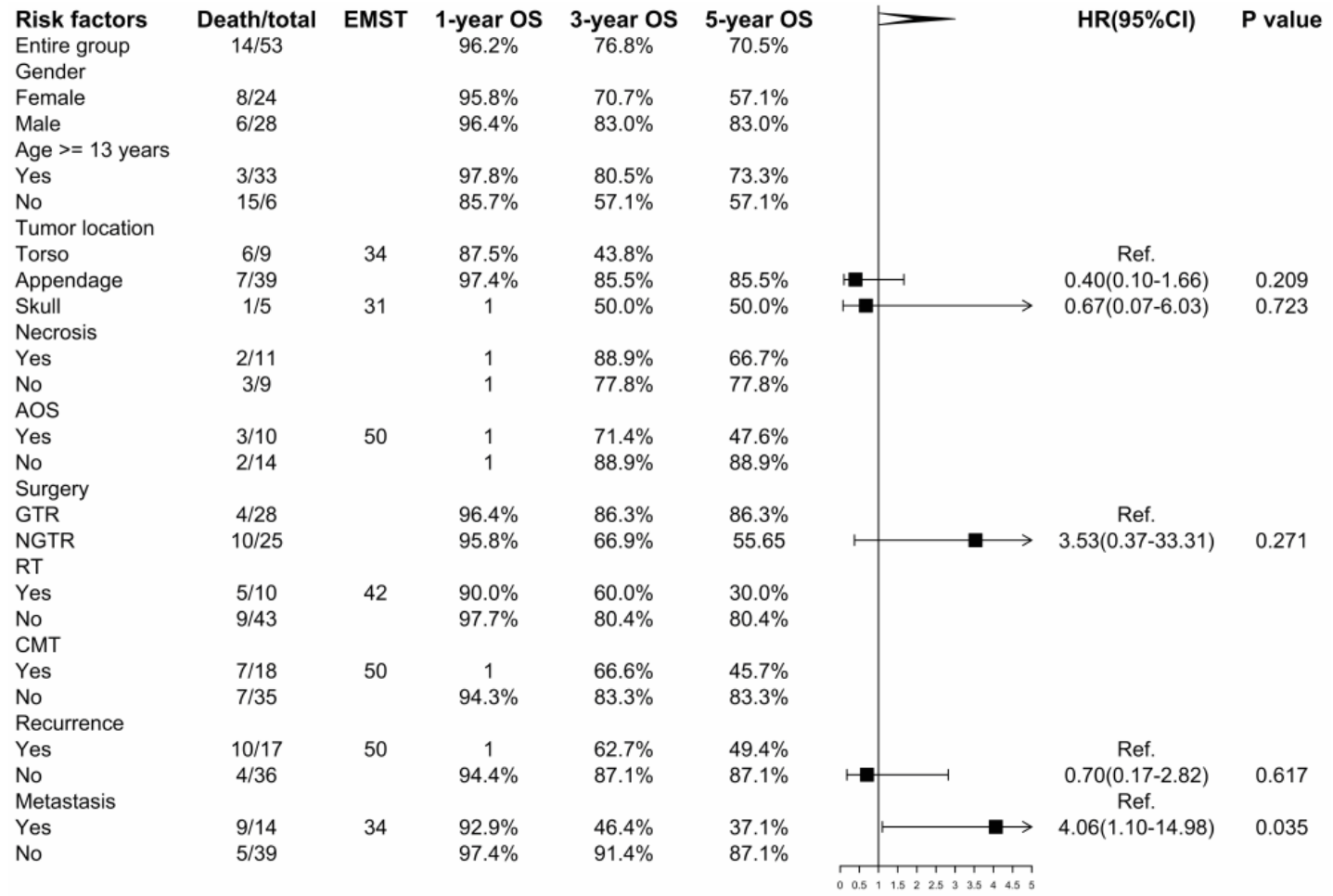

Figure 4

Multivariate Cox analyses was used to evaluate the adverse factors for OS. Black squares indicate the hazard ratio (HR) values, error bars represent the 95% confidence interval (CI).

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- CAREchecklistEnglish2013.pdf
- Supplementaltable1.docx