Introduction

Extraskeletal Ewing’s sarcoma (EES) is a member of the Ewing’s sarcoma family of tumours (ESFTs). The ESFTs include various types of lesions, such as EES, skeletal Ewing’s sarcoma (SES), peripheral primitive neuroectodermal tumour (pPNET), and Askin tumour (1,2). Members of the ESFTs have the same morphologic features, ultrastructure, immunophenotypes, cytogenetics,
and tumour gene expression (3,4). Given that Ewing’s sarcoma (EWS) and pPNET have the same neoplastic processes, the World Health Organization has recognized EWS/pPNET as the preferred terminology for soft tissue and bone tumours; terms such as “Askin tumour” are no longer used (5,6).

The skeleton is the most common site for EWS, and EES only accounts for about 20% of all ESFTs (7,8). As such, there exists a significant quantity of studies focused on the diagnosis and prognosis of SES, such that currently, the treatment strategies for SES are relatively mature (9). However, due to the rarity of EES, there is still a lack of high-quality research of EES and its clinical characteristics, therapeutic strategies, and prognostic factors vary among studies (1,10-12). EES can originate from any site in the body (12); thus, in the current study, patients presented to a wide range of departments, not just the department of bone oncology. Therefore, standard application of appropriate diagnostic and therapeutic strategies is crucial for specialists from various departments (13), such as preoperative biopsy and neoadjuvant chemotherapy. Preoperative diagnosis and thorough imaging of the characteristics of EES are important, especially for endoceliac tumours which are hard to identify by physical examination and are always misdiagnosed preoperatively.

Here, we report and compare the clinical characteristics and outcomes of a series of EES patients at our institution. The aim was to identify significant risk factors for EES prognosis. We also aimed to describe the imaging characteristics of endocelial EES by reviewing the data from computed tomography (CT). These findings are important for accurate diagnosis and treatment of EES. We present the following article in accordance with the STROBE reporting checklist (available at https://dx.doi.org/10.21037/tcr-21-607).

**Methods**

**Study subjects**

The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The retrospective study was approved by the Institutional Ethics Committee of Peking University First Hospital (No. 2021-365) and the verbal informed consent was obtained from all the patients. In total, 25 patients with primary EES, who were admitted to our centre from January 2000 to January 2020, were identified in the institution’s clinicopathological database. The inclusion criteria were as follows: (I) primary EES and the oncological diagnosis confirmed by two experienced pathologists specialising in sarcoma; (II) patient has detailed clinicopathological data; (III) information on overall survival (OS) of patients is available in the follow-up data. The extraskeletal origin of each patient was confirmed by imaging and pathological examinations, with findings indicating no bone marrow involvement; however, patients with oppression and reactive changes in the cortex of the bone adjacent to the EES were not excluded from our study. For the pathological examination, after excluding other small round cell tumours, such as lymphoma, rhabdomyosarcoma, and neuroblastoma, the still inconclusive cases underwent fluorescent in situ hybridization (FISH) to confirm the presence of chromosomal translocation involving the EWSR1 gene [t(11;22)(q24;q12)].

**Treatment**

All patients who underwent surgery or had unresectable disease received chemotherapy as an adjuvant or maintenance therapy. The chemotherapy protocols varied among the patients due to the time span of our study. Eight patients were treated with vincristine 2 mg/m², doxorubicin 25–75 mg/m², cyclophosphamide 900–1,200 mg/m² (VAC) alternating with etoposide 100 mg/m² and ifosfamide 1,800 mg/m² (IE) (1). The remaining patients received multiagent chemotherapy protocols based on vincristine, doxorubicin, cyclophosphamide, etoposide, ifosfamide, and cisplatinum. Only three patients received core needle biopsy and neoadjuvant chemotherapy before surgery. Seventeen patients underwent resection and postoperative chemotherapy. In addition, five patients obtained the histological diagnosis by core needle biopsy and received non-surgical treatment because of unresectable lesions, including metastatic diseases or lesions involving important body parts, such as the facial region. Only two patients underwent postoperative radiotherapy because of suboptimal margins.

**Data collection**

The demographic and outcome data included age, gender, primary tumour site, tumour size, distant metastasis, site of metastasis, therapeutic strategies, status of margins, regional lymph node metastasis, and OS. The tumour site was classified as either a surface or endocelial tumour.
according to the location of the lesion: surface tumours included those on the extremities, facial region, chest wall, and inguinal region; endoceliac tumours included those located in the retroperitoneum, prostate, adrenal gland, kidney, ovary, stomach, and lungs. The tumour size, i.e., the largest diameter, was measured based on imaging examinations, including CT, magnetic resonance imaging (MRI), and ultrasound, before the patient began treatment. The metastatic status was evaluated based on bone scan, chest CT scan, bone marrow biopsy, or positron emission tomography scan. Margins and regional lymph node metastasis were confirmed by pathological examination. The OS was calculated from the date of diagnosis to the date of last follow-up or death.

CT imaging analysis

Contrast-enhanced CT images from endoceliac EES patients were reviewed by two experienced radiologists who were blind to the clinicopathological data of the patient. The two radiologists reached an agreement by negotiating when there were divergences in opinion. Eight patients with endoceliac EES had complete contrast-enhanced CT imaging data available for review in our institution; CT features included contour, calcification, necrosis or cystic degeneration, degree of enhancement, enhancement pattern, lymphadenopathy, borderline, and organ invasion were reviewed. The contour was classified as round or lobulated according to the tumour’s regular or irregular shape. The presence of high-attenuation foci in the tumour was defined as calcification. When the CT value of the object region was less than 20 Housefield units (HUs) in the portal phase, the tumour was classified as exhibiting necrosis or cystic degeneration, the degree of which was graded as follows: mild, <50% low-density area in the lesion, and severe, >50% low-density area in the lesion. The degree of enhancement was determined by comparing the CT value of the lesion to the muscle and liver in the portal phase, as described previously (14). The enhancement pattern was classified as either heterogeneous or homogeneous. Lymphadenopathy was recorded when a regional lymph node had a short diameter >1.5 cm (6). An ill-defined borderline was recorded when the lesion involved the adjacent tissue and had a blurred margin.

Statistical analysis

The associations between clinical and pathological factors and long-term OS were assessed using the Kaplan-Meier method (log-rank test). Cox proportional-hazards models were used to determine hazard ratios (HR) in univariate and multivariate analyses. Tumour or treatment characteristics for which univariate analysis reached values of P<0.05 were tested in multivariate models. To achieve an exact statistical analysis, the rule of thumb is 10 events per variable in the Cox regression. For all analyses, P<0.05 was considered statistically significant.

Statistical analyses were performed using SE STATA (Stata Statistical Software, release 15.1; Stata Corp., College Station, TX, USA).

Results

Patient characteristics

Over the 20-year study period, 25 patients with primary EES were treated at Peking University First Hospital and were eligible for inclusion in this study. The patient characteristics are shown in Table 1. The sample comprised 7 females and 18 males with a male:female ratio of 2.6:1. The median age of the sample was 29 years (range, 1–72 years). The median tumour size was 8 cm (range, 3.5–17.0 cm). Seven patients (28%) presented with surface lesions and 18 (72%) had lesions located at endoceliac sites. Overall, 80% of patients had localised disease at diagnosis and 20% had metastatic disease. The lung was the most common metastatic sites for EES in our study.

Outcome analysis

The median OS was 22 months (range, 2-139 months). Results of the univariate analyses are presented in Table 2. According to the log-rank test, age, gender, suboptimal margins, and neoadjuvant chemotherapy were not prognostic predictors of OS (all P>0.05). There was also no significant difference in OS between localised and metastatic disease (HR 2.06, 95% CI: 0.72–5.87, P=0.15). Surface tumours, rather than endocelial tumours, were associated with improved OS; though, this was not statistically significant (HR 2.51, 95% CI: 0.78–7.51, P=0.101). Significant benefits in terms of OS were observed in patients who underwent surgery (HR 5.20, 95% CI: 1.45–18.62, P=0.003) and those with smaller tumour sizes (HR 2.48, 95% CI: 0.95–6.44, P=0.045). Among those who received surgery, patients without regional lymph node metastasis had significantly increased OS (HR 3.94,
95% CI: 1.10–14.09, \( P=0.021 \). Kaplan-Meier curves for the relationships between OS and tumour size, primary tumour site, distant metastasis, surgery, and regional lymph node metastasis are presented in Figure 1. A Cox proportional hazards model was constructed for the subsample of patients who underwent surgical treatment (n=20); this model was adjusted for potential confounders. The included variables, based on the log-rank tests with \( P<0.05 \), were regional lymph nodes metastasis and tumour size (Table 3). The results revealed that regional lymph node metastasis was an independent prognostic factor associated with OS (HR 3.72, 95% CI: 1.02–13.61, \( P=0.047 \)).

CT imaging findings

Only 8 patients with endoceliac EES have available CT data in our center and their CT features are summarised in Figure 2. Among these patients, 87.5% (7/8) presented with a lobulated contour, while calcification was detected in only 25% of the cases (2/8). Six patients (6/8; 75%) had severe necrosis or cystic degeneration and all cases presented with heterogeneous enhancement. In total, 75% of the patients (6/8) had a moderate degree of enhancement and the remaining two patients (25%) showed a low degree of enhancement. Half of the patients (50%) presented with lymphadenopathy and five patients (5/8, 62.5%) had an ill-defined borderline. Organ invasion was observed in 75% of the patients (6/8). Some typical CT images of endoceliac EES patients were shown in Figures 3-5.

Discussion

The ESFTs are a group of highly malignant diseases with short clinical courses and poor prognosis; these tumours generally present with early metastasis. Although SES is the third most common connective tissue malignancy in children (5), we still have little knowledge of the biological behaviour and prognosis of EES. Notably, to our knowledge, this study reports the largest series of Chinese EES cases to date.

Consistent with the available literature, the male preponderance of EES was also observed in our sample (10,15-17). Researches report that SES patients are younger than EES patients (1,10,13). In the current study, our EES patients were all around 30 years of age and there were no significant differences in OS between subgroups of different ages. Research also indicates that EES can originate from anywhere in the body, but the most common site for EES is the deep soft tissue of extremities (18). However, patients with surface lesions, including tumours located in extremities, only made up a small percentage of the patients with EES in our study. This may be because the general and urinary departments are the specialty departments of our institution, and thus, patients with EES originating from...
the gastrointestinal tract and urinary system were more commonly admitted to our institution. Javery et al. reported that abdominal EES commonly originates from the kidneys or retroperitoneum (7). Similarly, the most common sites of endoceliac EES in our research were the kidneys and retroperitoneum. Omori et al. reported that EES arising in cutaneous or subcutaneous tissue was associated with favourable prognosis compared to deep EES (19). Further, the prognosis of patients with EES arising from the abdomen is reported to be poorer than that of patients with EES arising from other sites (20). The abundant blood circulation and more common organ invasion may lead to the poorer prognosis of deep EES. Although the difference was not statistically significant, in our study we found that endoceliac EES tended to be associated with decreased OS as compared to surface EES. Further, in accordance

| Variables                          | No of patients | Hazard ratio (95%CI) | P value |
|-----------------------------------|----------------|----------------------|---------|
| Age                               |                | 1.90 (0.74–4.87)     | 0.156   |
| ≤29                               | 13 (52.00%)    |                      |         |
| >29                               | 12 (48.00%)    |                      |         |
| Gender                            |                | 1.28 (0.45–3.63)     | 0.624   |
| Female                            | 7 (28.00%)     |                      |         |
| Male                              | 18 (72.00%)    |                      |         |
| Primary tumour location           |                | 2.51 (0.78–7.51)     | 0.101   |
| Surface                           | 7 (28.00%)     |                      |         |
| Endoceliac                        | 18 (72.00%)    |                      |         |
| Tumor size                        |                | 2.48 (0.95–6.44)     | 0.045   |
| ≤8                                | 13 (52.00%)    |                      |         |
| >8                                | 12 (48.00%)    |                      |         |
| Surgery                           |                | 5.20 (1.45–18.62)    | 0.003   |
| Yes                               | 20 (80.00%)    |                      |         |
| No                                | 5 (20.00%)     |                      |         |
| Margins (n=20)                    |                | 1.74 (0.46–6.55)     | 0.412   |
| Wide                              | 16 (64.00%)    |                      |         |
| Suboptimal                        | 4 (16.00%)     |                      |         |
| Regional lymph node metastasis (n=20) |          | 3.94 (1.10–14.09)    | 0.021   |
| No                                | 16 (80.00%)    |                      |         |
| Yes                               | 4 (20.00%)     |                      |         |
| Neoadjuvant chemotherapy (n=20)   |                | 0.29 (0.04–2.26)     | 0.204   |
| Yes                               | 3 (15.00%)     |                      |         |
| No                                | 17 (85.00%)    |                      |         |
| Distant metastasis                |                | 2.06 (0.72–5.87)     | 0.150   |
| Yes                               | 5 (20.00%)     |                      |         |
| No                                | 20 (80.00%)    |                      |         |

Univariate Cox regression used to determine hazard ratio; log-rank p-values reported; CI, confidence interval.
with previous studies (1,21), the present study found that increased tumour size, which was associated with later local staging, was a risk factor for poor EES prognosis.

Distant metastasis has been confirmed to be an independent risk factor for poor EES prognosis in several studies (10,11,13,22). Contrary to expectations, this study did not find a significant difference in OS between localised and metastatic disease. The lack of significant difference might be due to the limitation resulting from small group and nonstandard treatment strategy in our center. In a previous study, effective systemic chemotherapy was associated with a 5-year survival rate of localised EES in

Table 3 Multivariate analysis for OS in patients underwent surgery using a Cox proportional hazards model

| Variables                        | Hazard ratio (95%CI) | P value |
|----------------------------------|----------------------|---------|
| Tumour size                      |                      |         |
| ≤8                               | 2.56 (0.82–8.00)     | 0.106   |
| >8                               |                      |         |
| Regional lymph node metastasis   | 3.72 (1.02–13.61)    | 0.047   |
| No                               |                      |         |
| Yes                              |                      |         |

CI, confidence interval
excess of 65% (7). However, in our study, the OS of EES was about 20% at two-year follow-up. The therapeutic strategies for EES are based on the experience garnered from the treatment of SES. Several studies have highlighted that the neoadjuvant chemotherapy was a standard therapeutic approach for localised EES (7,8). However, only 15% (3/20) of patients in the current study underwent surgery and agreed to neoadjuvant chemotherapy. Meanwhile, radiotherapy is recommended for patients with inadequate surgical margins (23). Nonetheless, only two patients (2/4, 50%) with suboptimal margins agreed to postoperative radiotherapy in the current study. Moreover, VAC alternating with IE, which is the normative aggressive chemotherapy protocol (24), was used in less than half of our patients. Any or all of these reasons could explain why patients with localised EES in our study had poor prognosis that did not differ statistically from the prognosis of patients with metastatic EES. At present, the widely

Figure 2 The incidences of tumor imaging features were visually described using a stacked bar plot. All the variables were dichotomous.

Figure 3 EES in the pelvis. (A) Unenhanced CT image shows a large ill-defined mass in the pelvis involving the bladder and oppressing the rectum. (B) Contrast-enhanced CT image shows mild heterogeneous enhancement caused by unenhanced areas indicating the necrosis or cystic degeneration. (C) Contrast-enhanced CT image in upper slice shows multiple metastatic lesions in enterocoelia and peritoneum. The lymphadenopathy can be seen in the hepatic hilar area (black arrow) and some punctate calcifications (white arrow) exist in the metastatic lesion.
accepted view is that aggressive multimodality treatment should be performed in EES. That is, treatment should begin with neoadjuvant chemotherapy, followed by local treatment including surgery and radiotherapy, and finally, multiagent adjuvant chemotherapy (13). Research suggests that regional lymph node metastasis is associated with disseminated disease and poor prognosis (25). Applebaum et al. concluded that lymph node metastasis was common in extraskeletal ESFTs (26). Our study also found that lymph node involvement was an independent risk factor for poor prognosis. This finding has highlighted the importance of evaluating the status of regional lymph nodes when

Figure 4 EES in the stomach. (A) Unenhanced CT image shows an irregular thickening with ill-defined boundary in the lesser curvature. (B) Contrast-enhanced CT image shows mild heterogeneous enhancement in the interior of the tumour.

Figure 5 EES in the adrenal gland. (A) Unenhanced CT image shows that a huge lesion with multiple low density areas and lamellar calcification (white arrow) compresses the right lobe of liver. (B) Contrast-enhanced CT image shows heterogeneous enhancement because of the necrosis or cystic degeneration and a filling defect in the postcava indicating the existing of tumour thrombosis (black arrow). (C) Coronal scan CT image shows that tumour displaces the normal adrenal gland and involves the postcava.
assessing EES. As such, technologies such as sentinel lymph node biopsies and routine regional lymph node dissection have potential application prospects for the treatment of EES.

The basic imaging features of EES, especially for endoceliac EES with insidious onset and poor prognosis, remain unclear. The application of CT can provide as much imaging information about ESFTs as MRI, is cheaper and more convenient than MRI in clinical practice (20). Endoceliac EES mimics other malignant lesions at corresponding sites, such as carcinoma of the kidney, prostate, colorectum, ovary, and so on. Therefore, it is crucial that we have a thorough general understanding of the CT features of endoceliac EES, as these patients tend to present to a wide range of departments, including general surgery, gynaecology, and urinary surgery departments; this would ensure that the various specialists who encounter these patients are aware of the importance of standard treatment for EES. Commonly, EES is a high-grade malignant tumour that tends to be large in size and has a quick growth rate. It is also commonly accompanied by a lobulated contour and necrosis, which can lead to a heterogeneous enhancement pattern on CT examination (1,6,20,27-29). An imbalance associated with the tumour’s fast growth rate and insufficient blood supply could be why these tumours tend to exhibit severe necrosis. Second, almost all publications on EES report ill-defined borderlines (29,30) and endoceliac lesions, like abdominal EES, more commonly exhibit organ invasion on CT, as compared to peripheral lesions (27). Finally, EES often presents with a moderate degree of enhancement (28) and absence of calcification (7,20,27). On the other hand, the current study also encountered some extraordinary findings related to the imaging features of endoceliac EES. The majority of imaging studies have indicated that lymphadenopathy is a rare CT feature in EES (7,27-29). However, half of the patients with endoceliac EES in this study presented with lymphadenopathy. Together with the findings of previous clinical analyses, the current findings suggest that serious preoperative evaluation, careful intraoperative management, and rigorous postoperative staging of regional lymph nodes are vital in the treatment of endoceliac lesions.

Our study also had several limitations. First, the main weakness of this study was the small sample size. This could be why endoceliac EES only trended towards poorer OS than surface, though this was not statistically significant. Second, our institution is famous for its urology and general surgery departments; thus, there was referral bias, and as such, this study had a large portion of patients with EES originating from the urinary and digestive systems. Third, despite the long-time span of this study, the follow-up periods for some patients were less than three years. In addition, the chemotherapy protocols varied across patients due to the time span of the study. Finally, this study was a retrospective study with inevitable selective bias. In spite of its limitations, this study is the largest published series of EES patients in China. The data acquired from reviewing clinicopathological and imaging data has potential value for improving preoperative diagnostic accuracy and formulating a standard treatment strategy for this rare disease.

In conclusion, large tumour size, non-surgical treatment, and regional lymph nodes metastasis were risk factors for poor prognosis in EES. In addition, regional lymph nodes metastasis was an independent predictor of OS in EES patients who underwent surgery. Endoceliac EES, which tends to be misdiagnosed preoperatively, exhibited the following common CT features: lobulated contour, absence of calcification, severe necrosis, moderate heterogeneous enhancement, ill-defined borderline, and organ invasion. Lymphadenopathy was also relatively common. Thus, the findings of this study contribute to the understanding of both the clinicopathological and imaging characteristics of this rare disease.

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**Footnote**

**Reporting Checklist:** The authors have completed the STROBE reporting checklist. Available at https://dx.doi.org/10.21037/tcr-21-607

**Conflicts of Interest:** The authors have completed the ICMJE uniform disclosure form (available at https://dx.doi.org/10.21037/tcr-21-607). The authors have no conflicts of interest to declare.

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**Ethical Statement:** The authors are 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. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The retrospective study was approved by the Institutional Ethics Committee of Peking University First Hospital (No. 2021-365) and the verbal informed consent was obtained from all the patients.

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