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Case Report

Intrabdominal Ewing Sarcoma of the Peritoneum: A Case Report

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ABSTRACT

Extraskeletal Ewing sarcoma (ES) is a small round-cell tumour and constitutes a sparsely reported clinical condition. We present a case of a 19-year-old man with abdominal distension and palpable masses in all quadrants of the abdomen. The patient was diagnosed with peritoneal ES and had multimodal oncologic treatment. On neoadjuvant setting, the patient received 9 cycles of chemotherapy and then underwent radical surgical excision, achieving Ro resection. Subsequent to an unremarkable postoperative course, 5 cycles of adjuvant chemotherapy were implemented to achieve optimal survival outcomes. On the current follow up, there is no evidence of local or further systematic spread of the disease. ES is a very rare malignancy and few cases have been published with only intraperitoneal spread. Chemosensitivity of the tumor is characteristic, making possible Ro resections, however local recurrence still exists.

Keywords: Extraskeletal ewing sarcoma; Extraosseous ewing sarcoma; Peritoneum

Introduction

Ewing sarcoma (ES) constitutes a malignant small round-cell neoplasm predominantly originating in skeletal and soft tissues, with a stated incidence of 1 case per million[1,2]. Extraskeletal ES is a rare clinical entity comprising 20% of ES[3]. The trunk is the most frequently impacted site, followed by the extremities, head and neck and the retroperitoneum[3]. We present a rare case of a 19-year-old man diagnosed with excessive ES of the peritoneum.

Case Report

A 19-year-old man presented to our department with massive abdominal distension, persisting for two months. His medical history was unremarkable and upon clinical examination a distended abdomen and palpable masses were found in all four quadrants, raising suspicion of sarcoma. Laboratory diagnostic tests were normal, with the exception of leukocytosis with neutrophil predominance. Following an ultrasound examination which revealed hypoechoic heterogeneous masses, a contrast-enhanced Computed Tomography (CT) scan yielded mild ascites and significant opacity of the peritoneal fat, along with heterogenous focal lesions. The largest mass was located in the lesser pelvis, measuring 20 x 14.5 x 16 cm, accompanied by concurrent enhancement in the paracolic gutters and omentum, indicating further peritoneal dissemination **(Figure 1a and 1b)**. Magnetic resonance imaging (MRI) confirmed the CT

findings, displaying a mass with restricted diffusion and central necrosis and established the differential diagnosis of a primary peritoneal lesion, sarcoma or stromal tumour **(Figure 2)**. Ultrasound-guided biopsy and histological examination established the diagnosis of extraskeletal ES and the patient commenced neoadjuvant chemotherapy. He underwent 9 cycles of chemotherapy every 15 days, consisting of 4 cycles of Ifosfamide, Etoposide, Mensa D1-D5 (IE) alternating with 5 cycles of Cyclophosphamide, Vincristine, Doxorubicin, Mensa (VDC) D1. The patient exhibited remarkable treatment response on follow-up CT scans, with a regression of ascitic fluid and a decrease in the size of the mass in the lesser pelvis to 3.5 x 2.5 cm, as well as of the concomitant lesions in the paracolic gutters and the omentum **(Figure 3)**. Subsequently, the patient underwent radical surgical excision of the peritoneal disease along with partial enterectomy 15 cm from the ileocecal valve due to local infiltration, with the objective of R0 resection. The postoperative recovery was uneventful and the patients was discharged on postoperative day 7. Histopathological analysis of the respected specimen confirmed the diagnosis of primary peritoneal ES. Following surgery, the patient received 3 cycles of Ifosfamide, Etoposide, Mensa D1-D5 (IE) alternating with 2 cycles of Cyclophosphamide, Vincristine, Doxorubicin, Mensa (VDC) D1. On the present follow-up, no evidence of recurrence has been indicated.

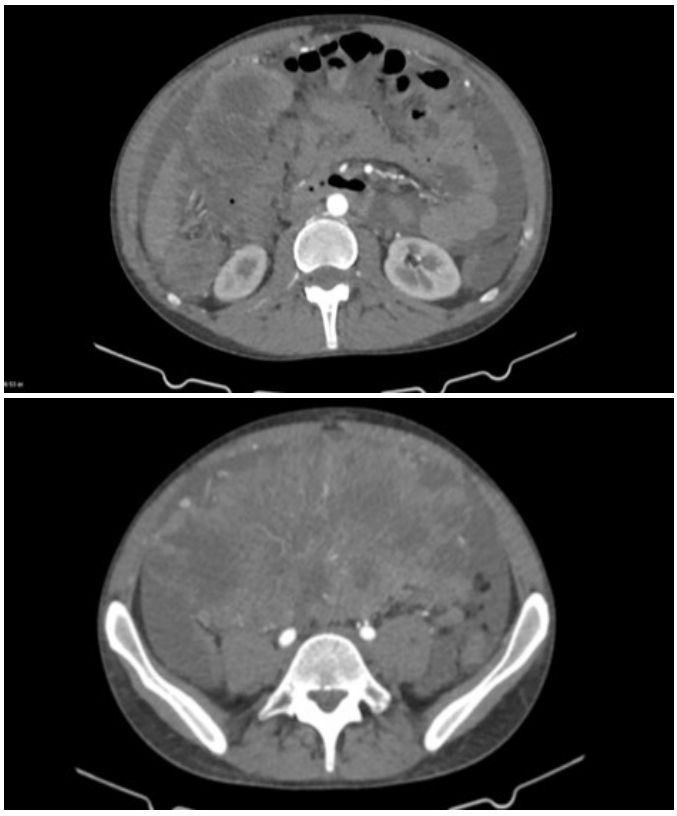

Figure 1a & 1b: Preoperative contrast enhanced CT scan of the largest lesion measuring 20 x 14.5 x 16 cm.

Discussion

Ewing sarcoma arises from a family of highly malignant small round cell tumours, defined by a shared genetic, histological and immunohistochemical basis[4]. This spectrum also encompasses extraskeletal ES, small-cell tumour of the thoracopulmonary region (Askin tumor) and soft tissue based primitive neuroectodermal tumours (PNETs)[4]. The predominant chromosomal translocation among 90% of ES family members is t (11;22) (q24; q12), resulting in the EWSR1-FLI1 fusion oncogene[1,3]. Although it continues to be a sparsely documented condition in the literature, extraskeletal ES was first described in 1969[5].

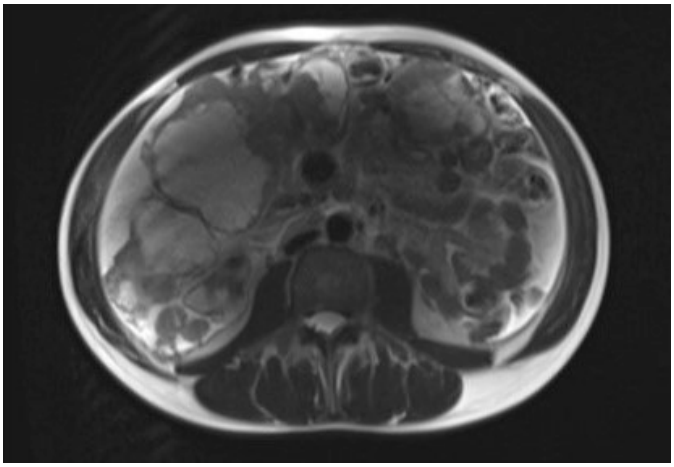

Figure 2: Abdominal MRI: the mass has high intensity on T2-weighted images.

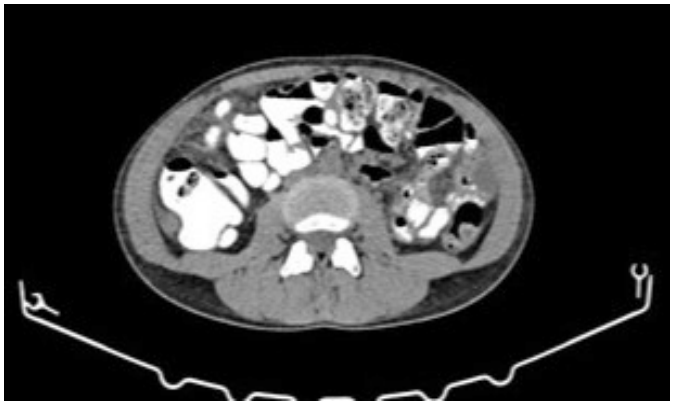

Figure 3: Post-chemotherapy contrast enhanced CT scan for tumour response assessment.

Unlike skeletal ES, extraosseous ES exhibits a higher mean age of onset and a bimodal age distribution, being more prevalent in patients younger than 5 and older than 35 years old[1,3]. Extraosseous ES is distinguished by its axial tumor origin and rarely exceeds 8 cm in diameter[3,6]. The clinical image of extraosseous ES is non-specific, resulting thus in delay of diagnosis, with a growing mass causing localised pain as the most prevalent manifestation[7]. Other symptoms vary according on the site of origin and may include fever, weight loss, fatigue and metastasis-derived symptoms[7].

While the imaging findings are likewise non-specific, imaging serves as important for the diagnosis, staging and subsequent follow-up of patients8. Ultrasonography demonstrates a hypoechoic mass displaying heterogeneity and intratumoral flow signs on Doppler imaging[7]. Similarly, CT scan indicates a defined mass with density equivalent to or lower than that of the muscle, while MRI yields a low to intermediate and a high intensity signal on T1 and T2-weighted images, respectively. Regarding contrast enhancement, extraosseous ES exhibits an heterogenous pattern associated with intratumoral necrosis in both imaging modalities[3,7]. MRI contributes to staging by assessing the local extent of the tumor, whereas FDG-PET/CT serves as the primary staging modality for identifying distant metastases[3]. Extraskeletal ES doesn't spread through the lymphatic pathways, but rather by hematogenous routes, often

localising in lungs, followed by bone and central nervous system involvement[8]. Metastatic development to other organs of the abdominal cavity, the peritoneum or surrounding soft tissue is uncommon, with a reported rate of 10%3,8. Meanwhile, a study by Eary et al illustrated that the standardised uptake value (SUV) of the primary tumor in FDG-PET/CT provides predictive information regarding patients' overall survival[9].

The conclusive diagnosis of extraskeletal ES necessitates histological analysis in conjunction with immunohistochemistry and genetic examination1. Differential diagnosis may prove challenging and consists of several small round cell neoplasms, including other tumours from ES family, embryonal rhabdomyosarcoma, neuroblastoma and lymphoblastic lymphoma[3,7].

Despite that optimal treatment protocols remain undetermined for extraskeletal ES, National Comprehensive Cancer Network (NCCN) suggests for the localised Ewing sarcoma family of tumours localised treatment (surgery and/or radiotherapy) alongside chemotherapy[1,10]. Following neoadjuvant chemotherapy, surgical excision of the primary tumor is recommended aiming for R0 resection, while radiotherapy is implemented in case of positive margins or in inoperable cases[1,3]. The prognosis of extraosseous ES is superior as opposed to the osseous subtype with five-year overall survival rates for localised tumours being 70%, whereas for metastatic disease 33%[1,3,7]. Metastatic spread represents the fundamental prognostic indicator with pelvic involvement and older age indicating a poor prognosis[1]. On the contrary, response to the neoadjuvant chemotherapy and radical surgical excisions have been correlated with favourable outcomes[1].

Conclusion

The present case report, which presents the peritoneal infiltration of extraosseous ES, emphasises on the challenge of diagnosis arising from disease entity's rarity. A high level of clinical suspicion, alongside clinical, imaging and histopathological findings are essential for ensuring optimal patient care. Thus, a multidisciplinary team approach, implementing reductive surgeries combined with chemoradiotherapy, is required to address this uncommon clinical condition and achieve favourable survival rates.

Consent for Publication

Written informed consent was obtained from the participant for the publication of the details of their medical case.

Availability of Data And Materials

Not applicable.

Conflicts of Interest

All authors declare no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

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Author Contributions

Conceptualization- D.L, methodology- E.M, data collection and formal analysis- E.M, S.C, K.P writing-original draft- D.L, E.M, S.C, writing- review and editing- D.L, K.P, A.M, supervision- A.M. All authors have read and agreed to the published version of the manuscript.

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