



Case Report

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Laparoscopic excision of Abdominopelvic Ewing's sarcoma in a child: a case report

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ABSTRACT

Ewing sarcoma is a rare and aggressive neoplasm that primarily affects the bones and soft tissues of pediatric and adolescent patients. When located in the abdominopelvic region, it poses challenges for treatment due to its proximity to vital organs. We present a case of a 16-year-old male diagnosed with abdominopelvic Ewing sarcoma. He presented with abdominal pain and swelling extending from the pelvis to above the umbilical region. CT imaging revealed a large mass with both solid and cystic components originating from the pelvic area, with surrounding displacement. A core needle biopsy confirmed the diagnosis. Neo-adjuvant chemotherapy was initiated to reduce the tumor size. Post-treatment imaging revealed two masses, one in the pelvis and one in the peri-splenic area. Laparoscopic surgery successfully excised abdominopelvic Ewing's Sarcoma. Histopathology showed clear resection margins and 60% necrosis. Adjuvant radiotherapy was given to minimize recurrence. This case demonstrates successful management using a multidisciplinary approach.

KEYWORDS: Sarcoma, Ewing, Case Report.

INTRODUCTION

Ewing sarcoma (ES) constitutes a notably aggressive neoplasm that predominantly targets osseous structures; however, it also possesses the capacity to present within soft tissue, commonly designated as extraosseous Ewing sarcoma (EES). This neoplastic entity predominantly affects adolescents and young adults, displaying a significant predilection for the pelvic region and the long bones. Within the classification of the Ewing sarcoma family of tumors (EFT), both Ewing sarcoma and peripheral primitive neuroectodermal tumors (PNETs) are encompassed, in addition to the aggressive small cell tumor of the thoracic wall, referred to as the Askin tumor, and atypical EES ^[1].

The occurrences of EES are relatively rare, with an incidence rate of 0.4 cases per million individuals, which is approximately an order of magnitude lower than that of bone Ewing sarcoma ^[2]. Instances of Ewing sarcoma localized within the abdominal cavity are infrequently represented in the existing scientific literature ^[3]. This pathology is often misclassified alongside other small round cell tumors, which feature embryonal rhabdomyosarcomas, neuroblastomas, and lymphomas. Implementing precise diagnostic methodologies is significant in facilitating the utilization of evidence-based, multimodal treatment regimens to achieve superior clinical outcomes ^[4].

The main objective of this investigative study was to ensure the accurate diagnosis of abdominopelvic Ewing sarcoma, enhance its management through collaborative decision-making among a multidisciplinary team (MDT), and accomplish complete resection through minimally invasive surgical techniques.

CASE PRESENTATION

A 16-year-old male patient presented with complaints of abdominal discomfort, coupled with a significant increase in abdominal swelling over the prior two months. The patient was referred to a Paediatric Oncologist through the Walk-in Clinic for further assessment. The diagnostic evaluation revealed the existence of a huge, firm neoplasm extending from the pelvic cavity to the supra-umbilical region. A computed tomography scan of the abdominal and pelvic regions demonstrated a markedly heterogeneous mass originating from the pelvic area, displacing neighbouring anatomical structures and encompassing solid and cystic components (Figure-I).

A biopsy via image-guided core needle confirmed Ewing sarcoma, characterized by the presence of CD99 and NKX 2.2 markers. An MDT decision led to the administration of six cycles of neo-adjuvant chemotherapy based on NCCN (National Comprehensive Cancer Network) guidelines ^[5], including vincristine, doxorubicin, cyclophosphamide, and etoposide, to shrink the tumor and facilitate surgical resection. Preoperative imaging identified two masses: one in the pelvis and another in the peri-splenic area (Figure- II).

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Figure-I: A huge heterogeneous abdominopelvic mass, marked with a star.

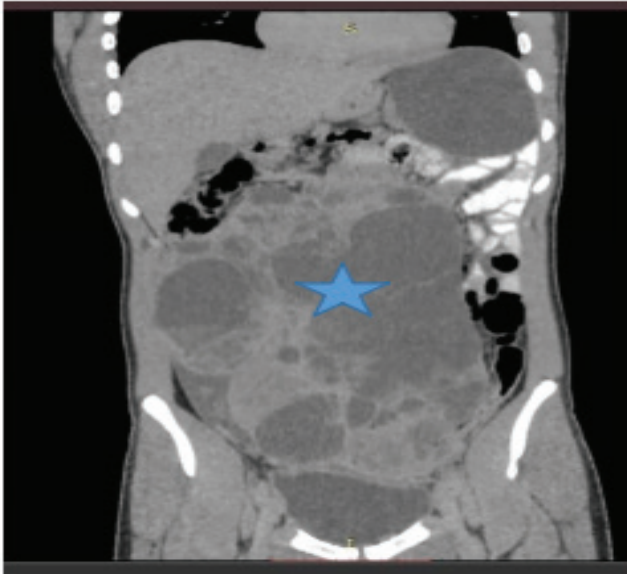


Figure-II: Reduction in size of the abdominopelvic mass and peri-splenic mass separate from tumor.

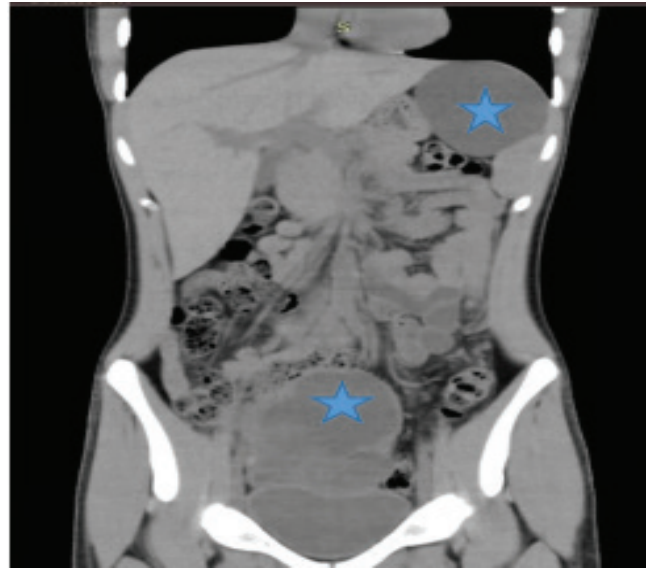
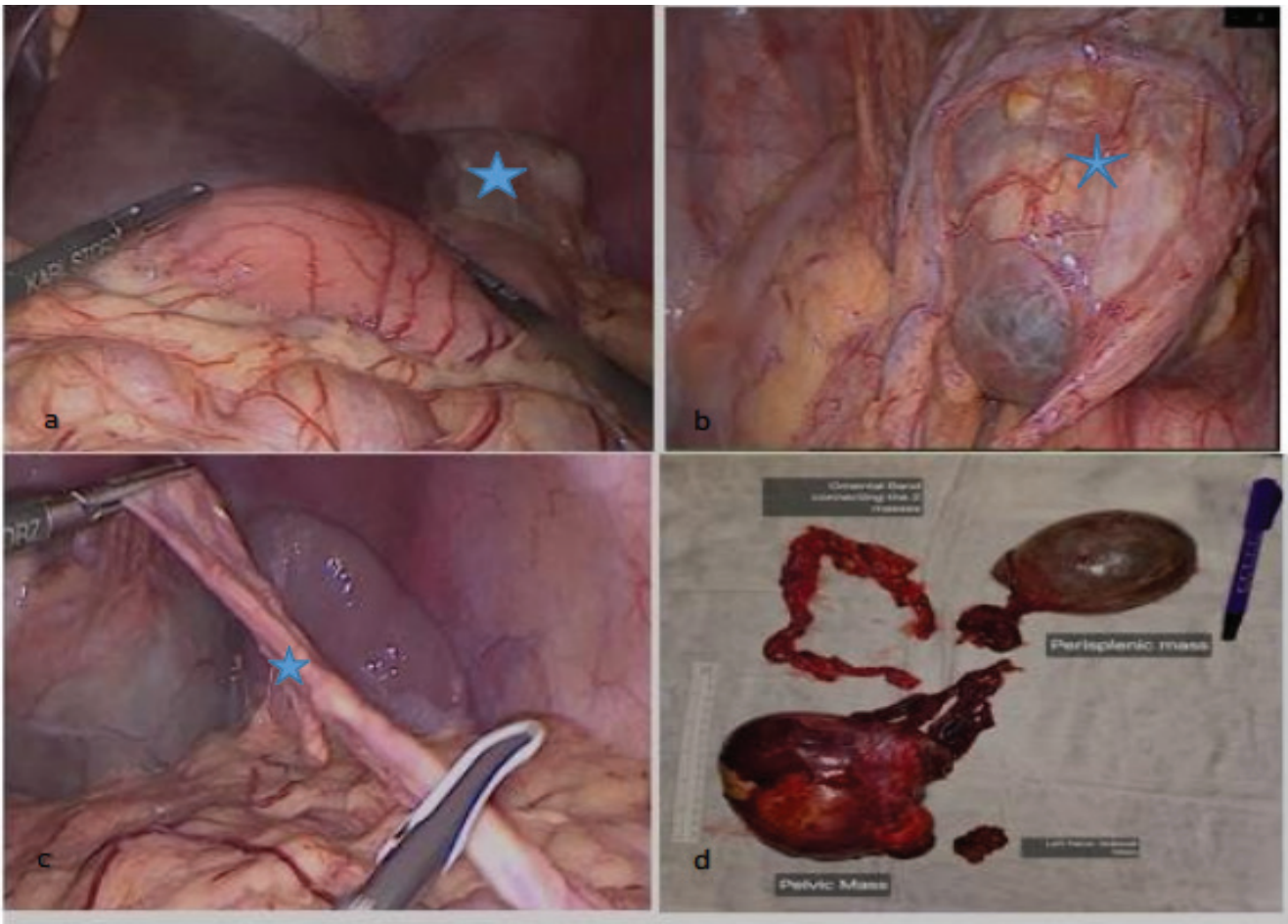


Figure-III: a: Peri- splenic mass, b: pelvic mass, c: omental band, d: excised specimen with the omental band, peri splenic mass, pelvic mass, and pelvic sidewall mass

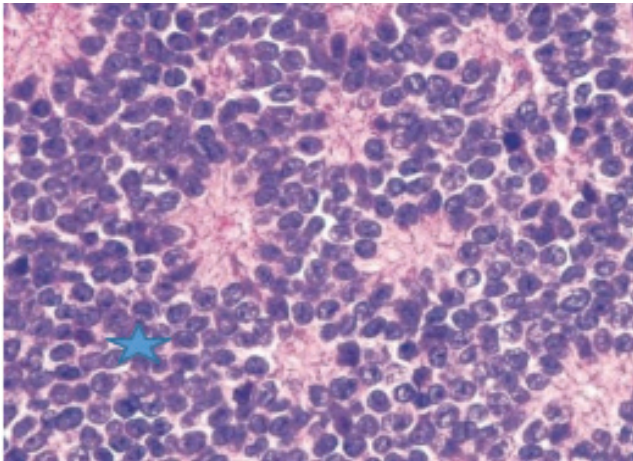


After 10 days of the last chemotherapy session, laparoscopy using four ports was performed. The primary mass was located behind the urinary bladder in the pelvis, and an omental band connected it to the peri-splenic mass. During the procedure, meticulous dissection separated the tumor

from adjacent structures, such as the bladder and rectum. A smaller pelvic sidewall mass was also removed. The peri-splenic mass was excised carefully to avoid damaging the spleen or its capsule, thus preventing tumor upstaging (Figure-III).

Postoperatively, the patient recovered well and resumed normal activities. Follow-up imaging confirmed complete resection of the tumor, and histology showed clear margins with 60% necrosis, with the presence of tumor in resected specimens (Figure-IV). An MDT decision of adjuvant radiotherapy was made to address any residual disease and reduce the risk of recurrence. Whole abdomen radiotherapy was started after 14 days of surgery. Regular follow-up, including physical exams, imaging, and tumor marker assessments, was scheduled to monitor for any signs of recurrence.

Figure-IV: Small round blue cells consistent with Ewing's Sarcoma.



DISCUSSION

A comprehensive multidisciplinary paradigm, including chemotherapy, surgical resection, and radiotherapy, is imperative in the management of abdominopelvic Ewing sarcoma in pediatric patients. The MDT meetings involve specialists from various disciplines discussing diagnosis, treatment options, and surveillance protocols, ensuring patients receive the best possible evidence-based care according to their needs. By promoting collaboration and ensuring comprehensive management, tumor boards play a pivotal role in improving survival rates, decreasing complications, and improving the overall quality of life for patients affected by this challenging malignancy [5].

Ewing sarcoma, characterized as a rare and aggressive neoplasm originating from osseous and soft tissue, poses significant therapeutic challenges due to its propensity for early metastasis and its anatomical positioning within the abdominal and pelvic cavities, thereby complicating both diagnosis and surgical intervention [1]. Our scenario showed a young patient with a diagnosis of abdominopelvic Ewing's sarcoma through a Tru-Cut biopsy. He was first treated with neo-adjuvant chemotherapy, then underwent laparoscopic excision of pelvic and perisplenic mass. Postoperative recovery was uneventful.

Ewing sarcoma is treated using a set of guidelines for chemotherapy. The implementation of neo-chemotherapy featuring drugs like vincristine, doxorubicin, cyclophosphamide, and etoposide significantly reduces the tumor's size, which helps in surgical procedures. This approach aims to diminish the tumor's volume and its metastases, thereby minimizing the extent of surgical intervention and reducing the risk of residual disease [5]. This preoperative approach holds significant importance in the context of abdominopelvic Ewing's sarcoma. It may allow for the tumor's relocation from essential anatomical components, consequently helping in effective surgical excision and lowering the risk of complications after the procedure.

Surgical intervention continues to represent the cornerstone of cancer treatment. In instances where the tumor is localized (as in our case), complete excision is essential for ensuring patient survival. The decision to adopt open or laparoscopic surgery relies on the tumor's dimensions and location in the body, together with the surgeon's experience. Laparoscopic approaches offer advantages such as diminished postoperative pain, short recovery periods, and reduced scarring; however, they necessitate advanced technical skills and meticulous planning to tackle potential complications [8]. The case illustrates the successful implementation of laparoscopic techniques in resectioning a challenging abdominopelvic mass, underscoring their significance in oncologic surgical practice [9].

Postoperative management constitutes an adjuvant strategy to address residual disease and prevent recurrence. Radiotherapy is frequently administered in conjunction with chemotherapy to eradicate remaining neoplastic cells when complete surgical resection is not achievable [10]. Routine follow-up care encompasses imaging studies and surveillance protocols designed to identify early indicators of recurrence and manage the long-term sequelae of treatment.

CONCLUSION

The best treatment of abdominopelvic Ewing sarcoma in children depends on a comprehensive approach, including pre-operative chemotherapy, surgical excision/local control, and further therapy. After the treatment, complex surgery and ongoing monitoring are important to improve effectiveness and increase the chances of survival in this complicated situation.

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Bilal Qayyum : Substantial contributions to the conception ,design and the acquisition of data for the work.

Sajid Ali : Drafting the work .

Tariq Latif : Reviewing it critically for important intellectual content.