International Surgery Journal
Aimanan K et al. Int Surg J. 2017 Dec;4(12):4079-4081
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20175415

Case Report

Extra skeletal Ewing's sarcoma: extra skeletal Ewing's sarcoma of mesentery masquerading dysgerminoma ovary

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Received: 18 September 2017
Accepted: 28 October 2017

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ABSTRACT

Extraosseous Ewing's sarcoma is a rare entity that involves the lower extremities, paravertebral regions of the spine, retroperitoneum, pelvis, and the chest wall. Involvement of mesentery as this case has been only reported five times in English literature. An eleven-year-old girl was planned for gynaecological surgery for suspected ovarian dysgerminoma. Intraoperatively she was referred to the surgical team since tumor arising from mesentery of the terminal ileum. Complete resection of the tumor with a right hemicolectomy was done. Histopathological assessment of the tumor revealed features of small blue round cell tumor which after an expert immunohistochemical staining reported as extraosseous Ewing sarcoma. She underwent a course of adjuvant chemotherapy and currently well during follow up. Due to the rarity of this tumor, a proper consensus on management has not been outlined. However, complete resection and adjuvant chemotherapy remain a standard therapy which showed an excellent post-operative outcome.

Keywords: Ewing's sarcoma of mesentery, Primitive neuroectodermal tumors, PAS-stained glycogen, Right hemicolectomy for Ewing's sarcoma of mesentery, Small blue round cell tumor

INTRODUCTION

Ewing sarcoma is a highly malignant tumor of long bones occurring in children and young adults and was first described by James Ewing in 1921.[1] However, there have been reported cases of malignant soft tissue tumors which are indistinguishable from Ewing sarcomas and have been called extra skeletal Ewing sarcoma. The most common sites are chest wall, paravertebral region, retroperitoneal space, lower extremities, and gluteal region. However, few cases have been reported in the kidney, breast, gastrointestinal tract, prostate, endometrium, the adrenal glands, brain, and lungs.[2] Its involvement in small bowel mesentery has rarely been reported (21 cases until August 2017).[3] Authors were described a case of mesenteric Ewing's sarcoma in an 11-year-old girl, which was preoperatively mistaken for ovarian dysgerminoma. The patient underwent complete resection and completed adjuvant chemotherapy. The purpose of reporting this case is to enlighten surgical trainees on this rare possible differential and management approach.

CASE REPORT

An 11 years old girl was brought into the outpatient clinic by parents with complaint of progressive abdominal distension for the past two months. She denied of any

compressive symptoms or history of trauma. There was no family history of malignancy. On examination she was well built and appropriate growth for her age. Abdominal examination revealed a firm and mobile intra-abdominal mass measures 10x15cm in size arising from right iliac fossa.

She was subsequently referred to a gynecologist. Computed tomography scan was performed for this patient and revealed a large intraperitoneal lobulated heterogeneously enhancing soft tissue mass arising from the pelvis extending superiorly up to L4 level. It measures 11.6cm x 6.0cm x 15.0cm in size. No evidence of fatty component or calcification within the mass. The mass abuts the uterine fundus and the right ovary and displaces the large and small bowels peripherally. The pre-op impression was ovarian mass or mesenteric tumor. After a multidisciplinary team discussion gynae team proceeded with tumor resection electively. Intraoperatively referred to the surgical team given mesenteric tumor adherence to the ileocecal junction. (Figure 1) There was no clear plane of dissection between the tumor and ileocecal junction, so we performed a right hemicolectomy and ileocolic anastomosis (Figure 2).

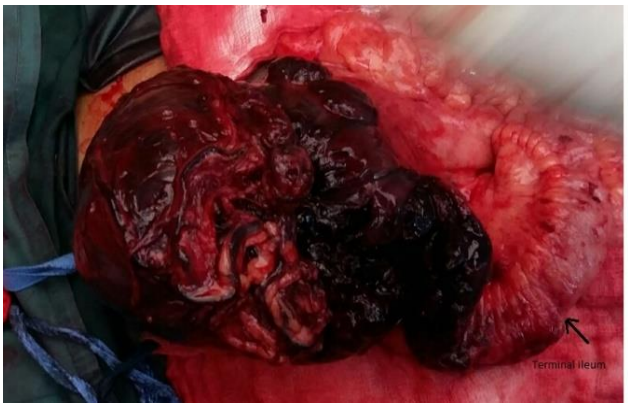

Figure 1: Mesentric tumor at terminal ileum.

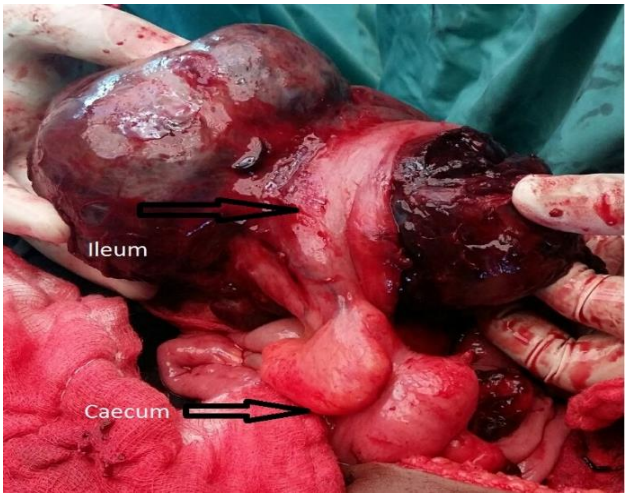

Figure 2: No clear plane between the mesentric tumor and ileocecal junction.

Postoperatively the patient was well and discharged to home after three days. The microscopic histopathology examination revealed a partially encapsulated mass that composed of small blue round cells with diffuse sheets and cords growth pattern. The tumor cells are rather primitive, displaying round to oval hyperchromatic nuclei, indistinct nucleoli and scant cytoplasm. It invaded serosal surface and the outer wall of the terminal ileum. Immunostains showed strong, characteristically diffuse membranous positivity for CD99 as well as diffuse nuclear positivity for NKX2.2, while WT-1 and inhibin were negative. The final histopathology report was Ewing's sarcoma arising from mesentery. The patient was sent to oncology and received a course of chemotherapy with vincristine, dactinomycin, ifosfamide and doxorubicin. Currently, she is well and performing her routine activities as usual.

DISCUSSION

Extraosseous Ewing's sarcoma is a rare malignant small round cell neoplasm of undifferentiated mesenchymal origin. It has been reported to be found in the soft tissues of the kidneys, retroperitoneum, intestines, and central nervous system. It is believed that this tumor is caused by genetic defects on chromosome 8, chromosome 11, chromosome 12, and chromosome 22 with a slight male predilection.[4] The factors that affect clinical signs were the site of origin and the size of the tumor. Surprisingly in our patient despite the size was more than 10cm and location at mesentery of terminal ileum she did not complain of any intestinal obstruction symptoms.

Formulating a definitive diagnosis in extraosseous Ewing sarcoma is a dilemma. Ewing's sarcoma and primitive neuroectodermal tumors (PNET) are regarded as two extremes of a morphologic spectrum of the same tumor entity based on similar clinical, immunohistochemical and cytogenetic profiles.[5] In literature, some pathologists view ESS as an entirely different disease entity from PNET, whereas other pathologists consider the two conditions as belonging to the same disease group. Although both ESS and PNET show expression of HBA-7 and the t11:22 translocation, only ESS contains PAS-stained glycogen in the cytoplasm.[6] Reciprocal chromosomal translocation t (11;22) (q24;q12) is a useful ancillary test however was not performed because of lack of this test at our institution.

Age and surgical treatment were found to be significant prognostic variables in the treatment of extraskeletal Ewing's sarcoma. No other variables, such as tumor size, tumor location, stage of disease, or radiation therapy, were found to improve survival. Surgical resection should be considered for all patients with extraskeletal Ewing's sarcoma.[7]

Chemotherapy is provided after surgery to improve overall survival rates and reduce the likelihood of tumor recurrence. First-generation regimens consisted of the

combination of vincristine, cyclophosphamide, actinomycin D and doxorubicin. Second-generation regimens incorporated ifosfamide and later etoposide with improved disease-free survival for patients with localized disease.[8] There is no proper consensus on specific treatment of extraosseous Ewing's sarcoma, which renders surgeons and oncologist to apply similar treatment as Ewing's sarcoma.

CONCLUSION

Although EES is extremely rare, it responds relatively well to a combination of surgical resection, chemotherapy and radiation therapy. Thus, an aggressive treatment protocol is needed. As there have been few reports on EES, clinicians should devise a more comprehensive and accurate pathological diagnostic method and a more systematic clinical evaluation method for the family of small round cell neoplasms of bone and soft tissue.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Aimanan K, Saprudin SA, Kanthasamy SV, Tangarathnam R, Guan CL. Extra skeletal Ewing's sarcoma: extra skeletal Ewing's sarcoma of mesentery masquerading dysgerminoma ovary. Int Surg J 2017;4:4079-81.