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Primary Ewing Sarcoma of the Mesentery - A Rare Case Report with Brief Review of Literature

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ABSTRACT

Primary Ewing sarcoma (ES) of the mesentery is an exceedingly rare malignancy, with only a few cases reported in the literature. Here we present a case of 31-year-old woman diagnosed with primary mesenteric Ewing sarcoma after a prolonged diagnostic course. The patient initially presented with recurrent abdominal pain while she was on treatment for infertility, and subsequent evaluation revealed mesenteric lesions along with venous thrombosis. Histopathologic and molecular analysis confirmed the diagnosis.
A review of the literature is included to provide insights into the clinical features, diagnostic approach, and management strategies for this rare entity.

Keywords: Extra-osseous Ewing sarcoma, soft tissue tumor, mesentery

CASE REPORT

A 31-year-old woman has been undergoing treatment for infertility when she developed recurrent abdominal pain. Initial evaluation at an external center revealed mild clumping of bowel loops in the right iliac fossa on computed tomography (CT). She was treated symptomatically without a definitive diagnosis. The symptoms recurred multiple times over a period of 2 years which prompted further evaluation, and repeat CT revealed terminal ileal thickening. The patient then presented to our institute and was re-evaluated. Tuberculosis work-up was negative. Colonoscopy showed edematous terminal ileal mucosa, and the mucosal biopsy showed eosinophilic infiltration; for which the patient was prescribed a short course of steroids. Despite treatment, abdominal pain persisted. A repeat Contrast enhanced CT enterogram revealed a heterogeneously enhancing, ill-defined infiltrating lesions in the small bowel mesentery, favoring a mesenteric infiltrative desmoid tumour.
She underwent diagnostic laparoscopy which showed multiple irregular, necrotic lesions extending along the mesentery of small bowel loops, predominantly involving the mid and distal ileum, causing bowel loop clumping. The lesion was sampled for biopsy and histopathological examination showed diffuse arrangement of medium-sized round cells having hyperchromatic nuclei, occasional distinct nucleoli, and scant cytoplasm (Figure 1a). Tumor showed areas of necrosis. Immunohistochemistry showed the neoplastic cells positive for NKX2.2 and CD99 (strong membranous staining) (Figure 1b,c). Negative staining

for synaptophysin, INSM1 and Chromogranin ruled out a neuroendocrine neoplasm. Desmin, CK were negative which ruled out Desmoplastic small round cell tumor. Similarly CD3, CD20, TdT were negative which ruled out a Lymphoma. FISH analysis confirmed EWSR1 gene rearrangement, establishing the diagnosis of primary mesenteric Ewing sarcoma (Figure 1d). The patient is now on neoadjuvant chemotherapy with VAC (vincristine, actinomycin-D, and cyclophosphamide) and IE (ifosfamide and etoposide phosphate) regimen.

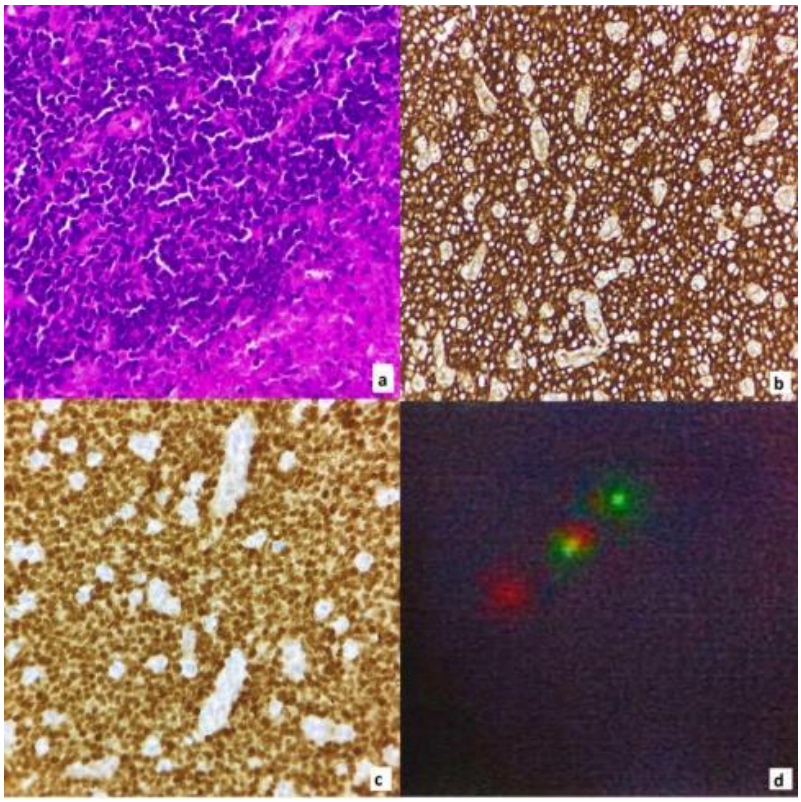

Figure 1: (a) H&E stained section showing diffuse arrangement of medium-sized round cells having hyperchromatic nuclei, and scant cytoplasm. Tumor showed areas of necrosis. Immuno-histochemistry demonstrates cells positive for (b) CD99, (c) NKX2.2. (d) EWSR1 gene rearrangement by FISH.

DISCUSSION

Ewing sarcoma (ES) is an aggressive malignant neoplasm, commonly of bony origin, and is the second most common pediatric bone tumor. Whereas extra-osseous Ewing sarcoma (EES) originating in soft tissues, accounting for 30% of all ES cases.[1] EES is more common in women, and there is a bimodal age distribution of patients being younger than five years and older than 35 years. The first reported case of EES in literature was that of a 56-year-old man, who had primary tumor in the mesocolon, with diffuse metastatic lesions in the liver at the time of diagnosis, and the patient unfortunately died despite treatments including subtotal gastrectomy and adjuvant chemotherapy. While the most common locations of EES include chest-wall, head and neck, para-vertebral region, and retroperitoneum, there are a few cases of EES involving the liver, pancreas, small bowel, uterus, and adrenal gland.[2] Mesentery of intestine is an extremely rare site. Peng et al published three rare cases of ESS that occurred in the mesentery and ileocecum, highlighting their clinical presentation, imaging findings, pathological features, and molecular genetics. He also reviewed the findings of previously documented 18 cases of EES originating in the small bowel, its mesentery, and

mesocolon.[3] Till date, to the best of our knowledge, nearly 37 cases of primary intestinal ES have been documented in the literature.(4) Clinically it is characterized by rapid growth of the soft tissue mass, often with early metastasis to lung, lymph nodes, or bone.

Patients with primary mesenteric ES may present with non-specific abdominal symptoms, such as pain, distension, or signs of intestinal obstruction. Our patient's clinical history of infertility and recurrent abdominal pain highlights the diagnostic challenges associated with atypical presentations of ES. The diagnosis of mesenteric ES requires a high index of suspicion and a multi-modal approach. The imaging features of extra-skeletal ES are nonspecific and typically demonstrate ill-defined soft-tissue mass with hemorrhagic and necrotic changes. Although some imaging features of EES have been suggested, including amorphous calcification (25%) on CT and flow voids of serpentine high-flow vessel components (90%) on magnetic resonance imaging, these findings are not specific enough to distinguish EES from other soft-tissue tumors.[5]

As the list of small round blue cell neoplasms are vast, histomorphology along with specific immunohistochemistry (IHC) and molecular studies are essential for a definitive diagnosis of ES. Ideally a panel of IHC markers should be done to differentiate ES from lymphoma (CD45), neuroendocrine neoplasm (Synaptophysin, CK), desmoplastic small round cell tumor (Desmin, WT1-C terminus), poorly differentiated synovial sarcoma (SS18-SSX) and neuroblastoma (PHOX2B). A combination of diffuse membranous staining with CD99 and nuclear staining with NKX2.2 is the most sensitive and specific IHC test for the diagnosis of ES. The NKX2.2 gene, a target of EWS-FLI1, is a useful marker with a sensitivity of 93% and a specificity of 89%.[6] The genetic hallmark is the presence of a specific translocation t(11;22)(q24;q12), which is expressed in 90-95% of patients.[7]

EES is treated in a same way as its skeletal counterpart. Treatment typically involves multi-modal therapy. Neoadjuvant/adjuvant chemotherapy is offered essentially for local control and systemic disease management, whereas surgery is the cornerstone for localized disease, and radiotherapy may be employed for residual or inoperable disease.[8] A suggested regimen of neoadjuvant chemotherapy is an alternating course of VAC (vincristine, actinomycin-D, and cyclophosphamide) and IE (ifosfamide and etoposide phosphate). After 4–8 cycles, the therapeutic effect is assessed by imaging studies and surgery is planned when possible. If treatment does not achieve a 25% regression in size on radiography, or 90 % tumor necrosis histologically, the response is judged as poor and second-line regimen such as high-dose chemotherapy with autologous stem cell rescue should be planned. Pazopanib, a multikinase angiogenesis inhibitor, has been shown to limit tumor growth modestly and has been approved by the Food and Drug Association (FDA) for treatment of advanced soft-tissue sarcomas. Pazopanib holds promise in cases of EES showing poor response to chemotherapy and also in cases of relapse following treatment, and clinical trials are underway.[9]

Prognosis of EES depends on tumor size, location, presence of metastasis, and response to therapy. Large tumor size, and old age, axial/pelvic location, presence of metastasis at diagnosis, poor response to chemotherapy, and are associated with poor prognosis. The overall survival rate is significantly improved with early diagnosis and aggressive treatment, and the 5-year survival rate after surgery and chemotherapy is approximately 70%.[10] The 5-year disease-free survival rate of patients with metastatic disease is only 35% while that of patients without metastatic disease is more than 60%. The prognosis of mesenteric ES is considered to be better as

compared to that of other sites, and is not associated with the tumor size.

CONCLUSION

Primary mesenteric Ewing sarcoma, though rare, should be considered in the differential diagnosis of mesenteric masses. Accurate diagnosis hinges on a combination of histopathological, immunohistochemical, and molecular techniques. Multi-modal therapy offers the best chance for survival in this aggressive malignancy.

Declaration by Authors

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REFERENCES

1. Turkyilmaz Z, Sonmez K, Karabulut R, Sen MC, Poyraz A, Oguz A, et al. Extraskeletal Ewing sarcoma of the mesocolon in a child. Journal of Pediatric Surgery. 2012 Sep;47(9):e1–3.
2. Li T, Zhang F, Cao Y, Ning S, Bi Y, Xue W, et al. Primary Ewing's sarcoma/primitive neuroectodermal tumor of the ileum: case report of a 16-year-old Chinese female and literature review. Diagn Pathol. 2017 Dec;12(1):37.
3. Peng L, Yang L, Wu N, Wu B. Primary primitive neuroectodermal tumor arising in the mesentery and ileocecum: A report of three cases and review of the literature. Experimental and Therapeutic Medicine. 2015 Apr;9(4):1299–303.
4. S P, P T, Br N. Primary Ewing sarcoma of the mesentery: An extremely uncommon case. J Med Sci Res. 2022 Jan 1;10(1):44–6.
5. Javery O, Krajewski K, O'Regan K, Kis B, Giardino A, Jagannathan J, et al. A to Z of Extraskeletal Ewing Sarcoma Family of Tumors in Adults: Imaging Features of Primary Disease, Metastatic Patterns, and Treatment Responses. American Journal of Roentgenology. 2011 Dec;197(6):W1015–22.
6. Yoshida A, Sekine S, Tsuta K, Fukayama M, Furuta K, Tsuda H. NKX2.2 is a Useful Immunohistochemical Marker for Ewing Sarcoma. American Journal of Surgical Pathology. 2012 Jul;36(7):993–9.
7. Saxena R, Sait S, Mhawech-Fauceglia P. Ewing sarcoma/primitive neuroectodermal tumor of the kidney: a case report. Diagnosed by immunohistochemistry and molecular analysis. Annals of Diagnostic Pathology. 2006 Dec;10(6):363–6.
8. Ahmad R, Mayol BR, Davis M, Rougraff BT. Extraskeletal Ewing's sarcoma. Cancer. 1999 Feb 1;85(3):725–31.
9. Keir ST, Morton CL, Wu J, Kurmasheva RT, Houghton PJ, Smith MA. Initial testing of the multitargeted kinase inhibitor pazopanib by the pediatric preclinical testing program. Pediatric Blood & Cancer. 2012 Sep;59(3):586–8.
10. Bernstein M, Kovar H, Paulussen M, Randall RL, Schuck A, Teot LA, et al. Ewing's Sarcoma Family of Tumors: Current Management. The Oncologist. 2006 May 1;11(5):503–19.

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