International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Case Report of ALL Presenting as Solid Ovarian Tumor

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Abstract: ***Lymphoma/ leukemia can rarely present as solid ovarian tumors. Secondary ovarian involvement with disseminated lymphoma is more common than primary ovarian lymphoma. Possibility of secondary ovarian lymphoma should be considered in differential diagnosis for solid ovarian tumors presenting in adolescents when there is multi-system involvement in the form of lymphadenopathy involving multiple lymph nodal groups as well as extra-nodal involvement. Absence of typical radiological signs associated with germ cell tumors on imaging and normal levels of different tumor markers on biochemical evaluation should raise suspicion of lymphomatous etiology in such patients.Early diagnosis on imaging and histopathological examination can assist in timely initiation of appropriate therapy.***

Keywords: Ovarian tumor, ovarian lymphoma, disseminated lymphoma, ALL

1. Introduction

Ovarian involvement by lymphomatous processes is rare, however in the female genital tract, it is the organ most frequently affected by hematological malignancies [1]. Since no lymphoid tissue is seen in the normal ovaries on histology, the likelihood that lymphomas arising in the ovaries has been a topic of dispute[2]. Malignant lymphoma involvement of the ovary can be primary or secondary, arising primarily form the ovary or secondary to dissemination of lymphomatous malignancy [3]. In this report, we describe a case of ovarian lymphoma that manifested as an ovarian tumor with gastrointestinal symptoms and concealed extra-ovarian illness.

2. Case Report

A 13-year-old girl hailing from a village in north India, presented with complaints of on and off fever with evening rise in temperature, significant weight loss and night sweats. Patient also complained of abdominal pain and distension with non bilious vomiting, suggestive of subacute intestinal obstruction. On physical examination patient was severely emaciated and had generalised lymphadenopathy. Per abdomen examination revealed an abdominopelvic mass which was non-tender. Per rectal examination revealed a hard mass anterior to rectum about 4cm proximal from the anal verge.

Initial routine biochemical investigations revealed counts, liver enzymes and renal function within normal limits. Electrolytes were deranged with hyperkalemia, and hyperuricemia- which suggested rapid cell death. Extended panel for pyrexia evaluation included CRP, ESR which were elevated. Gastric aspirate was sent for CBNAAT for Tuberculosis which came out to be negative. However, as tuberculosis is endemic in India and on probing patient's family gave positive contact history for tuberculosis, the child was empirically started on ATT. Meanwhile ultrasound was done which revealed a large solid pelvic mass of likely adnexal origin with non visualisation of bilateral ovaries and CT scan was planned. A large well defined heterogeneously enhancing solid mass showing areas of necrosis was seen without any foci of calcification or fat attenuation within the mass. Left ovary could not be visualised separately from the lesion *(Figure 1a,b)*. Circumferential thickening was also seen involving the proximal ileal loop in right iliac fossa with complete luminal obliteration of this segment and proximal dilatation of bowel loops *(Figure 1a,c).* Distal small bowel loops and colon were collapsed. Multiple enlarged homogeneously enhancing mesenteric and inguinal lymph nodes were seen *(Fig 1a).* Also, two lesions showing similar characteristics as the adnexal mass were noted in the pancreas *(Fig 1a)*.

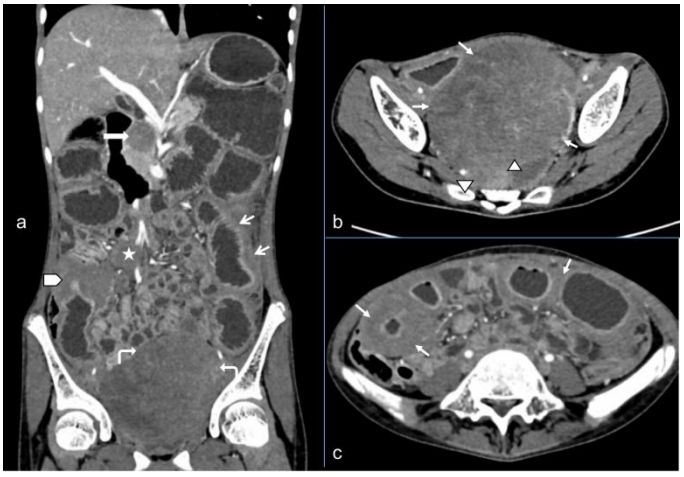

Figure 1: Coronal and axial images of CECT scan.
Fig. 1a- Coronal reformatted images of contrast enhanced CT scan of abdomen showing a large solid mass in the pelvis (curved arrows); multi-focal small bowel wall thickening (thin arrows and arrowhead); mesenteric lymph node (star) and hypo-enhancing solid lesion in pancreas (thick arrow).

Fig. 1b- Axial CECT images of pelvis showing large relatively homogeneously enhancing mass marked by arrows of left ovarian origin. Right ovary is marked by arrow heads.

Fig. 1c- Axial CECT images of abdomen revealing small bowel thickening involving ileal loops (thin arrows) with luminal compromise in right sided ileal loops.

Considering these findings two possible differentials of disseminated lymphoma and bilateral ovarian germ cell tumor with small bowel involvement were kept. Tumor markers were done including LDH, AFP, CEA, CA19-9, CA-125 and BHCG, of which only LDH was elevated. FNAC was also done but was inconclusive and biopsy was done which was reported as a germ cell tumor. Hence the patient was taken up for surgery.

On exploratory laparotomy, circumferential narrowing of ileal loop was noted at 80cm from ileocecal junction. Proximal bowel loops were dilated till the level of obstruction with collapsed distal small bowel, ascending colon, transverse and descending colon. Left ovarian mass was seen, right ovary was found separate from the mass separate and was bulky (Figure 2a). Multiple matted mesenteric lymph nodes were noted corresponding to the level of obstruction (Figure 2b).

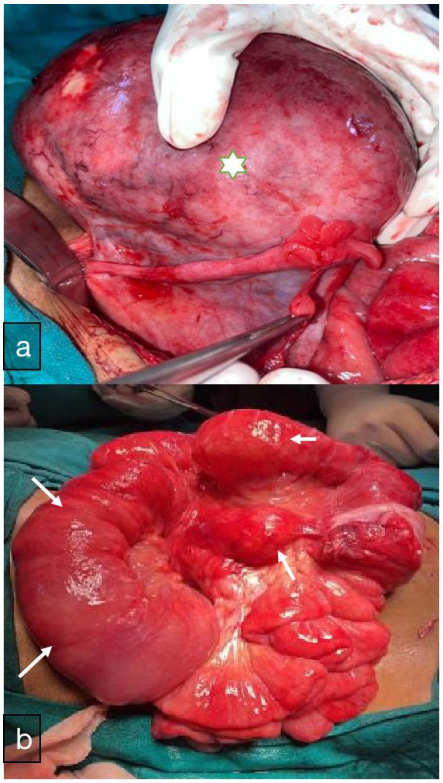

Figure 2: Intra-operative images
Fig. 2a- left ovarian mass (star marked).
Fig.2b- dilated small bowel loops with bowel wall thickening.

By the time the patient presented to our institute, a tertiary care center, she already had advanced disease with generalised lympahdenopathy and multiple solid organ and well as bowel involvement. The patient was in acute intestinal obstruction at the time of presentation, for which emergent operative management was advised. However, exploratory laparotomy had to be delayed due to financial constraints. This led to worsening of the patient's condition and she was severely emaciated with unstable vitals at the time of surgery. A multidisciplinary team including pediatric surgery, pediatric critical care, hematology, medical oncology and anesthesiology worked together to stabilise the patient and she underwent left ovarian mass resection and thickened small bowel was resected with reanastomosis of small bowel was done. The child developed persistent hypotension which was refractory to all inotropes. Despite the best efforts, she could not survive the post operative period and died on post operative day 3.

The resected ovarian mass and small bowel were sent for histopathological examination. On gross examination, the left ovarian mass measured 10x11x4.5 cm with an intact capsule and was solid and grayish white. The serosa of the left fallopian tube was intact. Microscopic examination of ovarian mass revealed ovarian tissue was replaced by diffuse sheets of small round blue cells with scant cytoplasm and high nucleus:cytoplasm ratio. There was increased mitotic activity with many atypical mitoses. Neither follicular structures nor starry-sky pattern was noted. Immunohistochemically, the cells were positive for LCA and negative for cytokeratin, which confirmed the diagnosis of lymphoma. The cells were also positive for CD79a , CD10 and TdT which suggested immature B-cell origin. Small bowel specimen showed similar histological picture with transmural involvement. A diagnosis of metastatic B-cell acute lymphoblastic/lymphoma was made.

3. Discussion

Malignant ovarian tumors are uncommon in the pediatric population and constitute for approximately 1%–2% of all childhood cancers[4]. Our patient presented with constitutional symptoms and gastrointestinal complaints for which she underwent biochemical evaluation and imaging. The initial biochemical evaluation was inconclusive with elevation of non-specific markers of systemic disease processes. On imaging, a solid mass of likely ovarian origin was found along with generalised abdominal and extra-abdominal lymphadenopathy as well as hollow viscous (small bowel) and other solid organ (pancreas) involvement.

Differential diagnosis for solid ovarian tumor in a child includes, in decreasing order of likelihood, germ cell tumors (GCTs), surface epithelial stromal tumors, sex cord–stromal tumors (SCSTs), and miscellaneous tumors (ie, gonadoblastoma, malignant lymphoma and leukemia, small cell carcinoma, and soft-tissue tumors) [4]. SCSTs and gonadoblastomas and rarely malignant GCTs, present with precocious puberty or virilisation and menstrual irregularity related to estrogen secretion [5]. Surface epithelial tumors are rare in adolescents and its occurrence in this age group is restricted to post pubertal females. All these pertinent positive findings were absent in our case. Among the GCTs, yolk sac tumors and choriocarcinomas are likely to have elevated tumor markers (AFP and B-HCG) which was not seen in our case.[6] Lack of areas of fat attenuation and calcification also excluded teratoma. Hence the differentials considered for the ovarian mass were malignant GCT (dysgerminoma) and leukemia/lymphoma[4]. The concurrent involvement of small bowel and pancreas along with

generalised lymphadenopathy was more in favor of the latter [7], which was later confirmed on histopathological examination.

Lymphoid malignancy of the female genital tract is unusual, more so in this age group, with ovaries being the most common site to be affected. Some studies have reported up to 25% women dying with lymphoma have ovarian involvement.[8] The ovarian involvement in malignant lymphoma may be primary or secondary (more common). The initial clinical manifestation of an occult nodal lymphoma as an ovarian mass is known as having a poor outcome with a survival rate ranging from 7% to 38% at 5 years. But primary ovarian lymphoma has better prognosis, which could be because secondary ovarian involvement represents disseminated disease which would be expected to have a worse prognosis.[9]

Primary ovarian lymphoma should be considered when there is presence of an ovarian mass, confined to one or both ovaries and extensive intraoperative and postoperative staging procedures show no evidence of lymphoma elsewhere in the body.[10]Our patient did not fit this criteria as she had generalized lymphadenopathy clinically at presentation, hence secondary ovarian involvement was more likely. On subsequent CT evaluation multiple abdominal lymph nodal involvement with small bowel thickening and pancreatic deposits was seen, which supported a diagnosis of widely disseminated leukemia/lymphoma with an initial ovarian presentation.

Imaging features of ovarian lymphoma are similar to lymphomas elsewhere in the body. On Ultrasound, the lesion is hypoechoic without posterior acoustic enhancement and CT scan will show hypodense lesions with homogenous post contrast enhancement. Areas of necrosis may be seen. On MRI these appear hypointense on T1-weighted images and slightly hyperintense on T2-weighted images. Possibility of lymphoma should be considered when imaging reveals bilateral ovarian tumors that appear homogenous in the absence of ascites.[7,9]

The histological appearances of lymphoma in the ovary are generally similar to those seen in the extra-ovarian sites. The most common types of lymphomas that can have ovarian involvement are diffuse large B-cell, Burkitt and follicular lymphomas. Rarely precursor B-cell lymphoblastic lymphomas are also encountered but are relatively rare.[11] However, these need to be distinguished from other round-cell tumors such as metastatic poorly differentiated carcinoma, especially of mammary origin; primary small-cell carcinoma and dysgerminoma. Immunohistochemical studies help to distinguish between these tumors, like in our case where the cells were positive for CD79a, CD10 and TdT suggesting precuror B-cell origin. IHC for BCL2, BCL6 and MYC were negative, ruling out follicular lymphoma, diffuse large B-cell lymphoma and Burkitt's lymphoma, with the final diagnosis being B-cell ALL. Patients diagnosed with B-ALL should be started on chemotherapy as soon as possible. In our case, the patient died before the histological diagnosis could be obtained, hence she never received appropriate chemotherapy.[12]

4. Conclusion

Lymphomatous malignancies rarely can present as solid ovarian tumors in the adolescent girls, although microscopic involvement appears to be relatively more common in disseminated disease. Ovarian lymphoma should be considered in differential diagnosis for solid ovarian tumors when there are features of disseminated disease. Germ cell tumors are the most common solid tumors of the ovary in this population, hence this should also be included in the differentials. Radiological imaging, histopathological examination and immunohistochemistry can confirm the diagnosis of lymphoma. High index of suspicion of lymphomatous etiology in such patients can aid in prompt diagnosis and initiation of appropriate therapy.

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