Primary Burkitt's lymphoma of the ovary

Sir,

Primary ovarian lymphomas are rare in the paediatric population. We recently had a case of Burkitt's lymphoma of the ovary that clinicoradiologically mimicked advanced epithelial ovarian cancer.

A 13-year-old female presented with rapidly progressing abdominal distension and fever. Clinically, she had ascites. The peripheral lymph nodes were not palpable. Her haemogram and renal/liver function tests were within normal limits. A computed tomography (CT) scan of the abdomen revealed bilateral adnexal masses and peritoneal/omental deposits [Figure 1]. No nodes were demonstrable. Tumour markers, including Human Chorionic Gonadotropin (HCG) and Alpha feto protein (AFP), were normal. There was mild elevation of CA–125 (112 U/ml). Serum Lactic dehydrogenase (LDH) was 2,280 U/L.

Biopsy of the peritoneal deposit demonstrated proliferation of atypical lymphoid cells with scanty cytoplasm and round nuclei with dispersed chromatin and many small nucleoli exhibiting a high mitotic rate [Figure 1]. Immunohistochemistry was performed. CD45, CD20, CD79a and CD10 were positive. BCL-2 and TdT were negative. Ki-67 was positive in >90% of the cells. Bone marrow studies and CT of the chest were normal. Hence, the final diagnosis was primary Burkitt's lymphoma of the ovary. The patient was initiated on LMB 89 protocol. She attained complete remission and remained clinically stable at 6 months of follow-up in our outpatient clinic.

Primary ovarian lymphomas account for 0.5% of all non-Hodgkin's lymphomas and 1% of all ovarian neoplasms.[1] The origin of these rare tumours is controversial. The malignant transformation of benign lymphoid infiltrates seen in up to 50% of normal ovaries, as suggested by Monterroso *et al.* may partly explain their pathogenesis.[2] The symptoms are nonspecific, but presence of constitutional symptoms and the rapid progression in a young patient should raise the suspicion of a lymphoma. The probability of tumour lysis syndrome is very high in this setting. Fox *et al.* suggested diagnostic criteria for primary ovarian lymphoma, which needs (a) the disease to be confined to the ovary, (b) absence of disease in the blood and bone marrow and (c) the extraovarian deposits, if any, should appear at least after few months.[3] High-grade non-Hodgkin's lymphomas like Burkitt's lymphoma and diffuse large B cell lymphoma predominate the histologies. Crawshaw *et al.* described the extremely rapid growth of the tumour and solid lesions with preservation of follicles in the periphery as the imaging findings of primary ovarian non-Hodgkin's lymphomas.[4] However, the bilateral ovarian masses with diffuse peritoneal deposits, as in our case, make it difficult to differentiate it from epithelial ovarian cancer. The prognosis of ovarian lymphomas is excellent.[5] B cell lymphomas fare better than T cell histologies. The treatment is mainly combination chemotherapy and the protocol depends on the primary histology. The role of surgery is debatable with present day chemotherapy regimes.

To summarise, we presented a case of primary ovarian Burkitt's Lymphoma in a 13-year-old girl, clinicoradiologically mimicking advanced ovarian

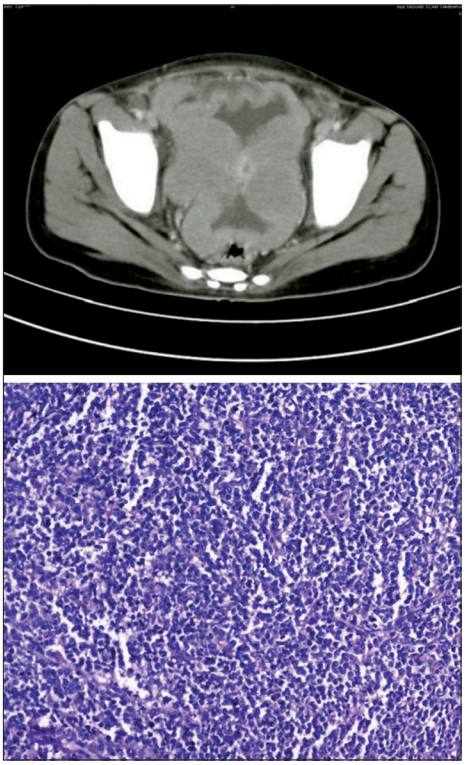


Figure 1: Top: bilateral adnexal masses and peritoneal/omental deposits Bottom: Proliferation of atypical lymphoid cells with scanty cytoplasm and round nuclei with dispersed chromatin and many small nucleoli exhibiting high mitotic rate

epithelial malignancy and successfully treated with combination chemotherapy. Ovarian lymphomas should be differential diagnosis in malignant ovarian tumours, especially in young patients, and an appropriate preoperative diagnosis can avoid extensive and unnecessary surgery in these patients.

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