

Primary Burkitt's lymphoma of the ovary (Case Report)

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الخلاصة

اورام المبيض نادرة في الاطفال . لقد وثقنا حالة لمرض (بيركت) اللمفاوي الابتدائي في مبيضي طفلة عمرها ثلاث سنوات كانت تشكو من الام مزمنة في البطن مع حمى وشحوب الوجه وورم في اسفل البطن . وبعد اجراء عملية فتح البطن الاستكشافي واستئصال الاورام وارسالها الى الفحص النسيجي تاكد بان المريضة مصابة ب (ورم بيركت اللمفاوي الابتدائي في المبيضين) .

Abstract

Ovarian neoplasms are rare in children . We had reported a case of primary Burkitt's lymphoma of the ovary in a 3-year-old girl presented with chronic abdominal pain with fever, pallor, and abdominal mass which was proved after laparotomy and histopathological study of the excised tumour to be a primary Burkitt's lymphoma of the ovary.

Key words : Burkitt , ovary , lymphoma.

Introduction

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⁾

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High-grade non-Hodgkin's lymphomas like Burkitt's lymphoma (BL) and diffuse large B cell lymphoma predominate the histologies. (BL) is a highly aggressive lymphoma often presenting at extranodal sites or as an acute leukaemia. BL is a B-cell neoplasm composed of monomorphic, medium-sized cells with basophilic cytoplasm and a high proliferation fraction, characterized by translocation and deregulation of the c-myc gene on chromosome 18 (4). This disorder often presents with extranodal disease and occurs most often in children and immunocompromised hosts.

Ovarian involvement by lymphoma is usually secondary, occurring as part of systemic disease. Primary malignant lymphoma of the ovary is a rare tumour and bilateral ovarian BL involvement mimicking gynaecological malignancy in adults is unfrequent (5). 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. (6) 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. (7) 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.

Case report

Primary ovarian lymphomas are rare in the paediatric population. We recently had a case of Burkitt's lymphoma involving both ovaries, small bowel segment, and mesenteric lymph nodes.

A 3-year-old Iraqi girl (Figure.1) from Wasit governorate (Badrah subdistrict) presented on 15th of March 2009 with recurrent severe abdominal pain, prolonged fever, and progressive pallor for three months duration; there was no history of diarrhea, vomiting, or dysuria. On examination, the girl was in severe pain, pale but not jaundiced, with a firm abdominal mass (5 cm by 4 cm) localized in the suprapubic region, no other findings were observed.



Figure.1- a 3-year- old postoperative patient with abdominal mass.

Abdominal ultrasound revealed a large hypoechoic mass of irregular outline (73mm ×65mm) seen in hypogastric region (may be related to small bowel loop or ovaries) with multiple mesenteric lymph nodes (??Lymphoma) . A computed tomography (CT) scan of the abdomen and pelvis revealed bilateral adnexal masses and peritoneal/omental deposits , no nodes were demonstrable. Blood test results were normal including [white blood cells(WBCs) & differential count, erythrocyte sedimentation rate (ESR), lactate dehydrogenase (LDH) , Uric acid , Renal & liver function tests] ; except haemoglobin (Hb) =84 gm/l .Serological tests for human immunodeficiency virus (HIV) , hepatitis C virus and hepatitis B virus were negative. Bone marrow aspirate and biopsy specimens and cerebrospinal fluid (CSF) analysis were normal. Chest radiograph was clear.

Explorative laparotomy by infraumbilical transverse incision revealed bilateral large ovarian solid tumors with multiple mesenteric and para-aortic lymph nodes . Removal of both ovarian tumours with resection of a segment of small bowel and excision of two lymph nodes ; all were sent for the histopathologist .

The gross section showed a piece of ovarian mass soft , grey, homogenous with area of necrosis, 4×4.3 cm × 3 p.; mesenteric lymph node 0.8 cm grey homogenous ×2 p. ; and a piece of intestine 5 ×1.5 cm ×3 p with resection margins (Figure.2).



Figure.2 (gross section of ovaries and intestine)

Microscopical histopathological findings showed monomorphic infiltration of lymphoblast cells with scattered reactive macrophages consistent with (Burkitt's lymphoma) with starry sky appearance involving the ovarian tissue , lymph nodes showed complete effacement of architecture by Burkitt's cells , intestinal wall showed involvement by tumor with clear resection margins (Figures. 3,4,5)

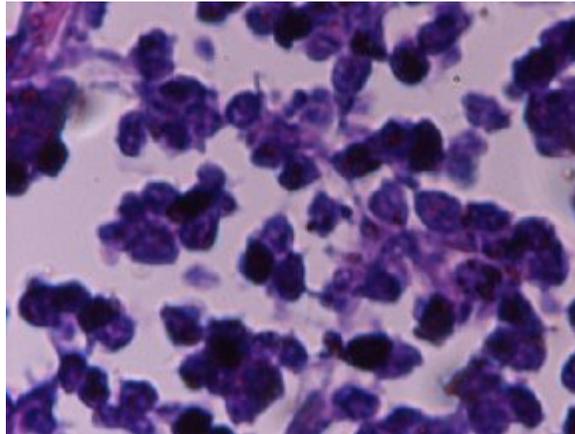


Figure.3 - starry sky appearance involving the ovarian tissue , lymph nodes showed complete effacement of architecture by Burkitt's cells .

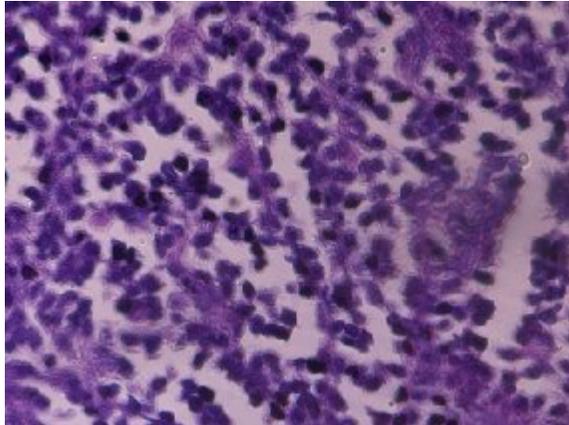


Figure.4 - monomorphic infiltration of lymphoblast cells with scattered reactive macrophages consistent with(Burkitt's lymphoma)



Figure.5- Burkitt's cells

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