# A case of B-cell lymphoblastic lymphoma involving the uterus

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## Summary

A 59-year-old postmenopausal woman presented with vaginal bleeding, lower abdominal pain, severe anaemia, leucocytosis, and an ultrasonographic finding of a large mass arising within the pelvis, most likely ovarian in origin. The patient was taken to the operating theatre with the possible diagnosis of acute haemorrhage into an ovarian cyst. At laparotomy there was a large mass at the posterior uterine wall extending retroperitoneally into the left pelvic side-wall. There was also significant paraaortic lymphadenopathy. The tumor was not resectable and biopsies were taken for pathological examination which showed a precursor B cell lymphoblastic lymphoma. Although the existence of lymphomas involving the uterus is well documented, the presentation of the lymphoma in this case was very unusual and this is the first reported case of a confirmed precursor B-cell lymphoblastic lymphoma involving the uterus.

*Key words:* Uterus; B cell lymphoblastic lymphoma; Genital neoplasms; Ovarian cysts.

## Introduction

Lymphomas involving the uterus are not unusual as has been shown in autopsy series of patients with lymphoma [1, 2] but only rarely is uterine involvement the initial presentation. They can either be primary or secondary in cases of disseminated disease. Histologically they almost always are non-Hodgkin's lymphomas and specifically B-cell with the diffuse large B-cell lymphoma being the main type [3].

We report a case of a patient with B-cell lymphoblastic lymphoma involving the uterus presenting as an ovarian cyst rupture.

## Case report

A 59-year-old, gravida 0, postmenopausal female was admitted as an emergency patient to our Hospital because she had collapsed. For the last few years the patient had been being nursed at a psychiatric institution because of schizophrenia and metal retardation. The patient was unable to give a history, but the nursing staff of the psychiatric institution mentioned that she had had vaginal bleeding for a week. Past medical history included excision of skin squamous cell carcinoma of the leg nine years before. On an incidental full blood count done eight days before admission the patient had a haemoglobin concentration of 93 g/l and a white cell count of 9.2.

On initial examination she was alert, disoriented, pyrexial (38.9°C) and haemodynamically stable (blood pressure 98/50, pulse 100/min). The abdomen was tender and distended, and a large mass arising from the pelvis and reaching the level of the umbilicus was palpable. Laboratory examinations revealed severe anaemia (haemoglobin: 49 g/l) and leucocytosis (white cell count: 31.5) with neutrophilia (neutrophils: 29.3, lymphocytes 0.95). Microscopic examination of a peripheral blood sample revealed severe anaemia with red cell anisocytosis including targets and stomatocytes, rouleaux, marked polychromasia, neutrophilia with a shift to the left and a leucoerythroblastic reaction. The morphology was suggestive of acute blood loss and possible underlying neoplasia. Biochemistry tests showed elevated urea (22.4 mmol/l), alkaline phosphatase (123 IU/l), lactate dehydrogenase (821 U/l), blood glucose (9.7 mmol/l), and C-reactive protein (64 mg/l). Serum sodium and potassium, creatine, aspartate aminotransferase, amylase and arterial blood gases were within normal limits. Total protein and albumin were decreased (44 g/l and 26 g/l, respectively). CEA was normal (2 µg/l) and CA 125 was mildly raised (67 U/l).

There were no EGG abnormalities. In the abdominal X-ray there was a large opacity in the lower abdomen and pelvis, fecal loading of the colon and distention of bowel loops. An abdominal ultrasound scan revealed a large mass arising within the pelvis, most likely ovarian in origin, bilateral hydronephrosis and hydroureter and left-sided pleural effusion but no evidence of free fluid in the abdomen.

The patient received a transfusion of six units of packed red blood cells within the next 36 hours. Following the transfusion the patient remained haemodynamically stable and haemoglobin concentration rose to 91 g/l. The patient was taken to the operating theatre with the possible diagnosis of acute haemorrhage into an ovarian cyst. On examination under anaesthetic a large abdominal mass arising from the pelvis was felt. At laparotomy the ovaries and the uterine fundus were normal. There was a large mass at the posterior uterine wall extending retroperitoneally into the left pelvic side-wall. There was also significant paraaortic lymphadenopathy. The liver was normal. A needle aspiration of the mass was done and no fluid was retrieved. The tumor was not resectable, and biopsies were taken for pathological examination. Following closure of the wound a hysteroscope was inserted into the vagina, which revealed tumor at the vaginal vault similar to that seen in the abdomen but the cervix could not be seen.

The pathology examination of the tumor biopsies showed total replacement of the myometrium by small to medium sized moderately pleomorphic lymphoid cells with occasional large phagocytes. Immunocytochemistry indicated that these cells

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were positive for B cells markers (CD20, CD79A). BCL-2 and cyclin D1 were moderately positive. T cell markers were negative in the majority of cells, while there was patchy positivity with CD21 and CD23. Overall the features were best regarded as those of a precursor B-cell lymphoblastic lymphoma according to the REAL classification.

The patient was haemodynamically stable and was having a gradual recovery during the first two postoperative days while on subcutaneous prophylactic dose of heparin. On the third day she developed acute respiratory depression and died within five minutes of the onset of the episode. A post-mortem examination was ordered.

In the post-mortem pathological examination evidence of aspiration pneumonia was found. There was also extensive lymphoma involving the para-aortic lymph nodes, the anterior abdominal wall peritoneum, the myometrium and, focally, the myocardium. No evidence of pulmonary embolism or myocardial infarction was found. The cause of death was presumably cardiac arrhythmia. With the additional information obtained from the post-mortem examination the lymphoma was staged as stage IV.

## Discussion

Lymphomas with uterine involvement, either primary or secondary, are not uncommon. In two autopsy series of patients with lymphoma uterine involvement was found in 7.6-10.1% [1, 2]. However they are not encountered frequently in gynaecological practice, possibly because the patients already have disseminated disease and thus the pelvic organ-related symptoms are trivial compared to the systemic symptoms. The presenting complaints include uterine bleeding and abdominal pain. In this case the presentation with acute blood loss and a pelvic mass probably of ovarian origin was misleading. There is no mention in the literature of a uterine lymphoma presenting as acute haemorrhage into an ovarian cyst, although the presenting symptoms have not been described in every case reported.

The lymphomas that involve the uterus are almost always non-Hodgkin's lymphomas and the main type is the diffuse large B-cell lymphoma. Other less usual types are the follicle center cell lymphoma, follicular, and small lymphocytic lymphoma [3, 4]. A MEDLINE search was done (key words: uterus, lymphoma, lymphoblastic lymphoma, genital neoplasms) for the years 1966-2001. This is the first reported case of a confirmed precursor B-cell lymphoblastic lymphoma involving the uterus. Because of the various classifications of lymphomas that have been used before the establishment of the REAL classification it is difficult to entirely exclude the possibility that the same type of lymphoma has appeared in the literature under another designation. It is certain however, that since the REAL classification has been in use there have not been any reports.

Precursor B-cell lymphoblastic lymphoma is uncommon and accounts for less than 10% of cases of lymphoblastic lymphoma and, unlike precursor T-cell lymphoblastic lymphoma, it usually involves extranodal sites, most often the skin and bones as well as the lymph nodes [5, 6]. Immunophenotyping is essential to establish the diagnosis of precursor B-cell lymphoblastic lymphoma as was done in this case.

## References

[1] Lathrop J. C.: "Malignant pelvic lymphomas". *Obstet. Gynecol.*, 1967, *30*, 137.

[2] Risdall R., Hoppe R. T., Warnke R.: "Non-Hodgkin's lymphoma. A study of the evolution of the disease based upon 92 autopsied cases". *Cancer*, 1979, *44*, 529.

[3] Vang R., Medeiros L. J., Ha C.S., Deavers M.: "Non-Hodgkin lymphomas involving the uterus: a clinicopathologic analysis of 26 cases". *Mod. Pathol.*, 2000, *13*, 19.

[4] Aozasa K., Saeki K., Ohsawa M., Horiuchi K., Mishima K., Tsujimoto M.: "Malignant lymphoma of the uterus. Report of seven cases with immunohistochemical study". *Cancer*, 1993, *72*, 1959.

[5] Lin P., Jones D., Dorfman D. M., Medeiros L. J.: "Precursor B-cell lymphoblastic lymphoma: a predominantly extranodal tumor with low propensity for leukemic involvement". *Am. J. Surg. Pathol.*, 2000, *24*, 1480.

[6] Iravani S., Singleton T. P., Ross C. W., Schnitzer B.: "Precurosr B lymphoblastic lymphoma presenting as lytic bone lesions". *Am. J. Clin. Pathol.*, 1999, *112*, 836.

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