

Case Report

Ovarian lymphoma – A rare presentation

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Introduction

Non-Hodgkin's lymphoma involving the ovaries is unusual and may cause confusion for the clinician since its presentation could resemble other, more frequent tumors. Malignant lymphoid cells may occur in the ovary either as a primary neoplasm or as a secondary manifestation of a disseminated occult or known disease. The most common presenting symptoms or signs of malignant lymphomas involving the ovaries are abdominopelvic pain or mass.

Involvement of the ovary by malignant lymphoma is well known as a late manifestation of disseminated nodal disease, almost always of the non-Hodgkin's type, however, primary ovarian lymphomas are unusual [1]. Primary ovarian Non-Hodgkin's Lymphoma (NHL) accounts for 0.5% of all non-Hodgkin's lymphomas and 1.5% of all ovarian neoplasms [2].

However, contrary to their traditional presentations, we present a unique case of non-Hodgkin's lymphoma involving the ovary presenting with a Fever of Unknown Origin (FUO). Thus, the objective of this case report is to highlight the variable presentation of primary ovarian lymphoma, as is described later, to alert healthcare providers to avoid delayed or misdiagnosis and, thus, prompt treatment.

Patients and methods

A 53-year-old nulliparous female presented to the Accident and Emergency department with three weeks history of general malaise, fever and left renal angle discomfort. She presented to her family doctor eight days ago with the same symptoms, urinary tract infection was suspected and she was treated with broad-spectrum antibiotics.

Examination revealed pyrexia of 38C, tachycardia of 120/min and blood pressure of 110/85, BMI of 52. Abdominal examination revealed a soft non-tender abdomen with no evidence of any swelling, organomegaly, or lymphadenopathy. A system review revealed no other significant abnormality.

Initial investigations revealed raised White Cell Count (WCC) of 11.3 and a C-reactive protein (CRP) of 245. Septic screens were performed including blood cultures, Mid-Stream

More Information

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of Urine (MSU) and chest X-ray. She was then commenced on antibiotics against both gram-positive & gram-negative organisms.

However, on the second day of admission the patient's temperature remained persistently elevated at up to 39.5 C, therefore, for which a third antibiotic (triple antibiotics), was added to her treatment regime. Intriguingly, her septic screens returned as unremarkable.

On the third day of admission, the patient developed nausea, vomiting, and diarrhea and continued to have a spiking temperature of > 39 C. Repeat C-reactive protein (CRP) was 238 but the White Cell Count (WCC) was falling. She remained on triple antibiotics. However, as the stool culture was positive for Clostridium Difficile, antibiotics were discontinued and the patient was commenced on oral Metronidazole. At that time, the provisional diagnosis was a Fever of Unknown Origin (FUO).

Persistent pyrexia promoted further workup which included an ultrasound scan of the pelvis and abdomen, which revealed a complex-looking pelvic mass of uncertain origin; thus, a CT abdomen and pelvis was carried out on the sixth day of admission. This showed an 8 cm complex left ovarian mass, however, there was no evidence of abscess or fluid collection. The above finding promoted further workup in the shape of tumor markers namely, CEA, CA125, Ca19.9, AFP & HCG as well as an MRI of the pelvis (Figure 1). Rheumatoid factor, ANA, ANCA, ENA, C3, C4 and Immunoglobulin were all normal. The Mantoux test was negative.

Subsequently, on the 7th day, in view of the patient's age,

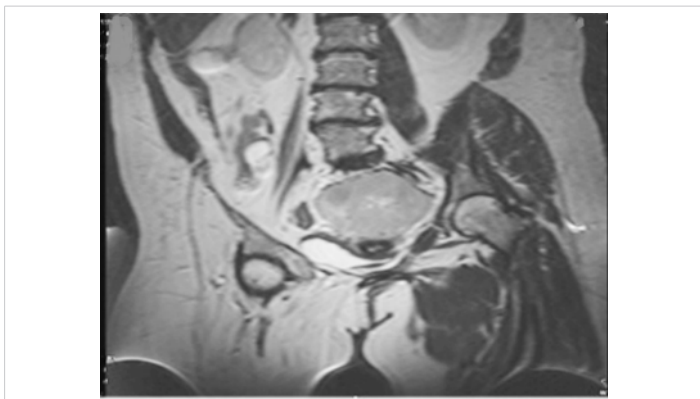


Figure 1: MRI pelvis showing large left ovarian mass.

imaging evidence of ovarian mass, and continuing symptoms, the patient underwent a diagnostic laparoscopy to assess the abdominal and pelvic cavity as well as the ovarian mass and then consider further intervention depending on operative findings.

Diagnostic laparoscopy was performed under general anesthesia which confirmed the same previous imaging findings, namely, a cystic ovarian mass, which was too friable and bled easily on touch to allow any laparoscopic manipulations, therefore, laparoscopy was abandoned in favor of laparotomy and left salpingo-oophorectomy.

The patient's temperature settled and vital signs normalized shortly postoperatively; she remained afebrile for the rest of her hospital stay and was discharged home on the fifth postoperative day.

Pathological assessment of the ovary revealed ovarian B cell lymphoma, but no other abnormalities were detected. Further workup with a gynecologist and oncologist team in the shape of bone marrow aspirate, PET scan and CT scan of the chest revealed no further abnormality.

Discussion

Most Non-Hodgkin's Lymphomas (NHL) are nodal. Extranodal NHL is frequently encountered in gastrointestinal and other mucosa-associated lymphoid tissues. Although NHL of the ovary is rare, it has been well-reported in the literature. The latter revealed two large case series: one of 42 cases, 1983 (1) and the second of 39 cases including one case of leukemia, 1993 (2). The vast majority of the cases in these series presented with an ovarian mass and most of them were referred with a suspicion of primary or secondary neoplasm [1,2].

Numerous published case reports emphasized that pelvic-related complaints, e.g., pain and/or mass were the commonest clinical presentation [3]. However, some unusual presentation was also reported, for example, some case series reported NHL cases with features suggestive of ovarian cancer, e.g., pelvic mass, pleural effusion and even marked elevation of CA-125 [4,5]. Other cases presented with rare

features, e.g., thickened omentum and ascites [6], autoimmune hemolytic anemia which occurs in 3% of NHL [7], abnormal genital bleeding [8], twisted ovarian cyst [9] and menstrual abnormalities [10] have been reported.

In general, true primary ovarian lymphoma usually carries a favorable prognosis [7,11,12], however, other favorable prognostic features should also be considered, e.g.:

1. FIGO stage IA.
2. Unilateral ovarian involvement.
3. Focal involvement of one ovary; and,
4. Follicular (nodular) lymphoma [1].

The main steps in the management are surgery and chemotherapy. The primary disease has better survival than secondary ovarian involvement.

Our case has had a unique presentation and clinical features that have not been previously reported/published. Although lymphomas represent an important cause of FUO, we are not aware of previously published cases of primary ovarian lymphoma presenting with FUO as the primary presentation (Figures 2,3).

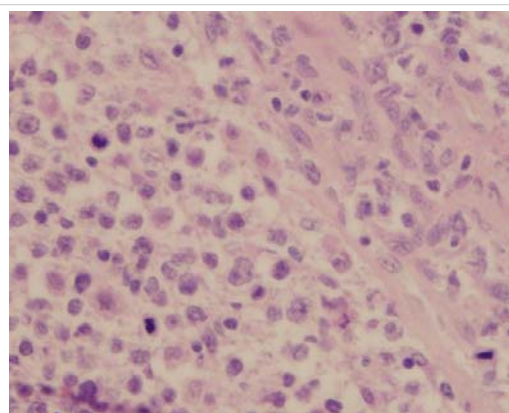


Figure 2: Diffuse large cell lymphoma (40x H&E) showing sheets of atypical cells with numerous mitoses.

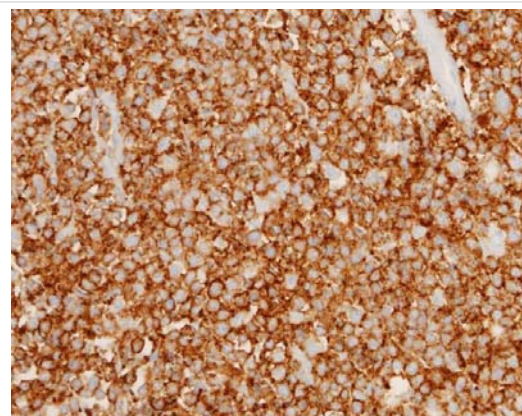


Figure 3: CD 20 immunostaining showing monoclonal diffuse membranous staining.



Conclusion

Although uncommon, it is advisable to consider ovarian lymphomas in the differential diagnosis of FUO. Moreover, to our knowledge, this is the first case report of primary ovarian lymphoma presenting as FUO. Therefore, we suggest considering primary ovarian lymphoma in the differential diagnosis of FUO in women presenting with a pelvic mass. This would help with prompt diagnosis and is likely to save the patient further intervention, e.g., antibiotic treatment, as well as health care resources by avoiding numerous other diagnostic and treatment modalities.

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