Bilateral ovarian Burkitt's lymphoma

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Summary

Primary ovarian lymphoma is a rare entity. We submit a case of a 34-year-old black patient presenting with a bilateral adnexal tumor. She underwent hysterectomy with double salpingo-oophorectomy followed by polychemotherapy treatment. Histology confirmed Epstein-Barr virus-positive bilateral Burkitt's lymphoma. The patient died from septic shock after a month of treatment. Endemic Burkitt's lymphoma has a predilection for the female genital tract, manifesting itself clinically as a pelvic mass and less frequently as a menstrual disorder. It is a rare entity in our environment but should be kept in mind when treating patients of African origin.

Key words: Ovary; Lymphoma; Burkitt.

Introduction

Burkitt's lymphoma is a specific type of poorly differentiated malignant lymphoma. It is a variation of non-Hodgkin's lymphoma which manifests in children – predominantly between the ages of four and seven although young adults can also be affected.

Certain geographical areas in East and West Africa as well as Papua, New Guinea are regarded as endemic given the relative frequency of the tumour – though non endemic cases have also been described [1, 2].

Ovarian involvement by lymphoma is a rare entity. It may be primary or secondary, although the latter is more frequent.

One of the characteristics is its unspecific symptoms, which on most occasions leads to casual diagnoses.

Case Report

A 34-year-old black patient from Senegal with an unremarkable history and medical obstetrical history reflecting an eutocic delivery was admitted to the emergency ward due to month-long abdominal pain that had worsened over the last few days, accompanied by a general state of deterioration.

Exploration revealed a large uterus-dependent abdominal mass that extended up to the navel.

Complementary vaginal ultrasound (US) was also performed, revealing a large complex cystic mass that exceeded the uterine bottom; it measured 190 x 115 mm and dense-looking content filled about two-thirds of it.

Abdominal scan detected the presence of grade 2/3 bilateral hydronephrosis secondary to the previously described mass and a small quantity of ascites. Thoracic X-ray showed left pleural effusion.

Analyses showed the presence of microcytic anemia and increased creatinine and tumor markers CA 125 (205 IU/ml) and HDL (1375 IU/l).

Given the patient's origin a serological study for hepatitis B, hepatitis C, human immunodeficiency virus and syphilis were requested with negative results.

During her admission there was a sudden deterioration of her general condition, together with a fall in hemoglobin and hematocrit levels; following the diagnosis of acute abdomen an exploratory laparotomy was urgently performed. After entering the cavity a 1000 cc hemoperitoneum was detected, and a large tumor with an irregular surface that blocked the pouch of Douglas was found adhering to the anterior face of the rectosigmoid. Hysterectomy and double salpingo-oophorectomy were performed, as well as removal of a large mass (adenopathy) at the level of the left common iliac vein; the extension study was completed with peritoneal biopsies and appendicectomy.

Anatomopathological macroscopic examination revealed bilateral ovarian involvement by two tumoral masses (19 and 21 cm), of solid appearance and soft tissue consistency (Figure 1).

Histologically, a lymphoid proliferation of small round cells of diffuse growth was observed, with a high nucleo-cytoplasmatic ratio. The mitotic index of the macrophages was high, creating the typical starry-sky pattern (Figure 2).

Immunohistochemical data evidenced B-cell p53 and Epstein-Barr virus-positive lymphomas (Figure 3).

Having reached a definitive diagnosis of Burkitt's lymphoma with bilateral ovarian involvement, an extened study by means of abdominopelvic computerized axial tomography showed a large mass that spread cranio-caudally at the level of the celiac trunk, rounding the mesenteric and renal vessels, aorta and inferior vena cava, as well as the iliac vessels, to a greater degree on the left side.

Bone biopsy and lumbar puncture were negative.

Postsurgical treatment was started with polychemotherapy, with the patient displaying a good response to the cytoreducing treatment.

The patient finally died after a month of treatment, having displayed symptoms of septic shock secondary to mediastinitis due to esophageal perforation.

Discussion

Burkitt's lymphoma was described by Burkitt [1] in Uganda in 1958. He described it as an endemic disease

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Fig. 1

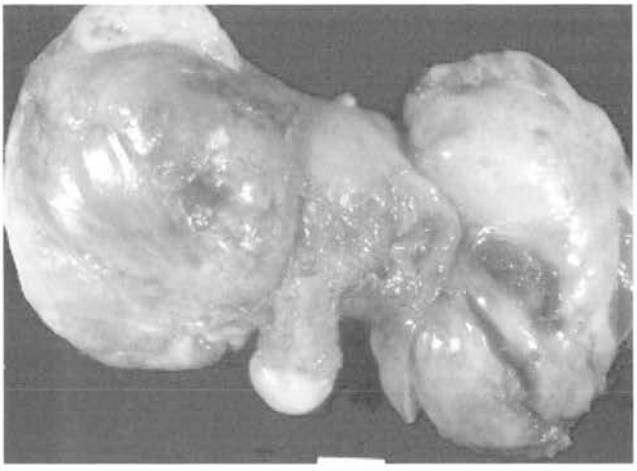

Fig. 2

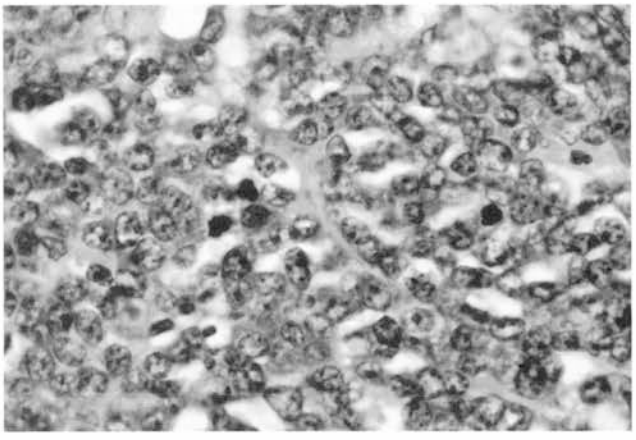

Fig. 3

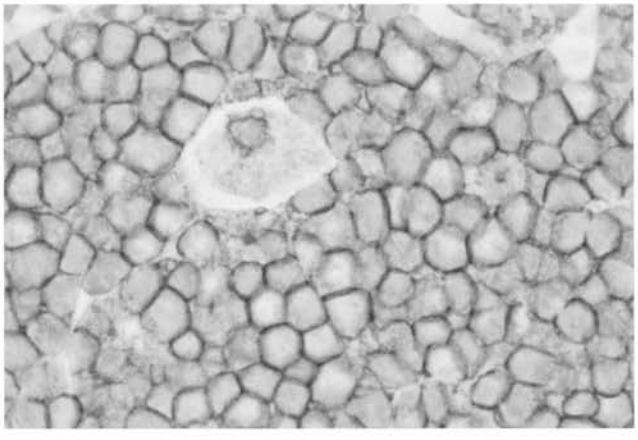

Figure 1. — Anatomopathological macroscopic examination: two tumoral masses 19 cm and 21 cm in size with solid appearance and soft tissue consistency.

Figure 2. — Histological examination: A lymphoid proliferation of small round cells of diffuse growth is observed with a high nucleo-cytoplasmatic ratio. The mitotic index of macrophages was high, creating the typical starry-sky pattern.

Figure 3. — Immunohistochemical data showing B-cell p53 and Epstein-Barr virus-positive.

characterized by the appearance of a tumour in the jaw and abdomen of children in Central Africa, although later cases were described with a similar histology in other parts of the world [2]. Ovarian involvement by lymphoma is rare; it may be primary, usually bilateral and associated with ascites [3] or secondary – the latter being more frequent [4].

The medical literature describes few cases of primary ovarian lymphomas [5, 6]. Between 7% and 26% of patients with malignant lymphomas suffer from ovarian involvement, although only 1% displays initial ovarian involvement [7].

There are three epidemiological clinical forms [8, 9]:

– *African or endemic variety* (our case): occurs more frequent among children, with seven being the average age. It is related to the Epstein-Barr virus and is frequently located in the mandible and abdominal viscera such as kidneys, ovaries and retroperitoneum.

– *Non-endemic variety:* average age is 10.2 and it affects the abdominal viscera. There is rarely mandibular involvement [10] and a lesser association with the Epstein-Barr virus.

– *Immunodeficiency-associated epidemic variety:* mainly nodal [12].

The most frequent signs and symptoms are abdominal pelvic pain, abnormal vaginal bleeding and a fast growing abdominal mass [2, 13].

A differential diagnosis should be made with granulose tumors, dysgerminoma, small-cell carcinoma of the hypercalcemic type, granulocytic sarcoma, poorly differentiated carcinoma, and metastasis, but mainly with epithelial ovarian neoplasias [14].

The definitive diagnosis of this entity should be undertaken by means of histopathological and immunohistochemical studies as well as of a genetic study that should include c-myc detection. Translocations t (8:14), t (2:8), and t (8:22) have also been used to confirm the diagnosis [3].

One of the pathological characteristics – though not pathognomonic – has been the starry-sky pattern caused by extensive mitoses of macrophages. The fast growth of these tumors raises serum LDH levels.

Ovarian lymphoma can be treated by means of surgery and polychemotherapy, the latter being the treatment that might potentially lead to recovery [10, 11].

Prognosis depends on the stage at diagnosis, acute presence, histological type and response to treatment. The rate of recovery among patients with localized disease is 90%, and among those (as in our case) with disseminated disease the rate of recovery is 30%. The most common cause of death is cerebral involvement [5].

In conclusion, endemic Burkitt's lymphoma has a predilection for the female genital tract, manifesting itself clinically as a pelvic mass and less frequently as a menstrual disorder. It is a rare entity in our environment and should be kept in mind when treating patients of African origin.

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