UNUSUAL PRESENTATION OF NON-HODGKIN LYMPHOMA IN YOUNG FEMALE. CASE REPORT.

Fatima Hydar Abo Ahmed Mohd[1], Asadullah Nawazani[2], Mohammed Kamal Mohammed Mahgoub[3], Lakshmiah G Raman[4].

ABSTRACT

We are presenting a case report of a young female patient, who presented with rapid enlargement of breasts associated with generalized lymphadenopathy, fever and signs of 3rd cranial nerve palsy. Radiological imaging revealed widespread organ involvement and subsequent lymph node biopsy confirmed Burkett's Lymphoma. This case reminds aggressiveness of this tumour and its capacity to invade multiple organ systems over a short period of time. Therefore, a high index of suspicion should be maintained in timely diagnosis of this rapidly developing and potentially curable disease.

Key words: Burkett's Lymphoma, Bilateral Breast Enlargement, Isolated Divisional 3rd Nerve Palsy.

1. Specialist Internal medicine, Internal Medicine Department. UAE.
2. SEC Endocrinology UK, Diploma in Diabetes Cardiff UK. Senior Specialist, Internal Medicine Department, Dubai Hospital, UAE.
3. Specialist internal medicine, UK.
4. Specialist Senior Registrar, Histopathology Department, Dubai Hospital, Dubai, UAE..

How to cite this article: Mohd FHA[1], Nawazani A[2], Mahgoub MKM[3], Raman LG[4]. **UNUSUAL PRESENTATION OF NON-HODGKIN LYMPHOMA IN YOUNG FEMALE. CASE REPORT.** *JPUMHS; 2021:11:01,142-146.*
http://doi.org/10.46536/jpumhs/2021/11.01.309

Corresponding author: Asadullah Nawazani, MRCP Ireland, FCPS (Medicine), SEC Endocrinology UK, Diploma in Diabetes Cardiff UK. Senior Specialist, Internal Medicine Department, Dubai Hospital, UAE.E mail: asadnawazani@hotmail.com

Received on: Dec 22, 2020, Accepted On 15 March 2021, Published On 31 March 2021

BACKGROUND

Burkett's lymphoma (BL) is a very aggressive type of B cell non-Hodgkin lymphoma, its doubling time range 24 to 48 hours, early diagnosis and prompt management is related to outcome in terms of morbidity and mortality.

CASE PRESENTATION

24-year-old expat female with no past medical history of significance, presented in an Emergency department (ER) of a hospital with ten days history of severe headache and blurring of vision. Prior to her presentation, she had visited another clinic with the same complains where headache was linked to a pain referred from a wisdom tooth.So she underwent a wisdom tooth extraction. However, headache persisted and she had developed blurring of vison as well with ptosis of left eye. On further questioning she revealed that, illness started 2 months prior to her presentation with inguinal swelling, which was gradual, non-tender associated with yellowish and itchy vaginal discharge, no genital ulcers. No fever or swellings elsewhere. The patient had received two courses of oral antibiotics without significant improvement. Shortly, after inguinal swellings, patient noticed gradual and progressive almost symmetrical increase in size of both breasts, reaching fourfold increase in two weeks' time, without tenderness, nipple discharge or any change in the overlying skin.At that time patient also started to have infrequent episodes of intermittent high-grade fever associated with sweating. It was not associated withweight loss or GI symptoms. The patient denied any previous history of hospital admission and chronic medical condition. She was not on regular medications and had no known allergy to drugs. She is married for one year and not pregnant, no history of abortion. She has irregular menstruation since menarche. She smoke regularly and consume alcohol socially. Examination of the patient at the time of admission revealed a fully conscious and oriented lady with intact higher mental functions and vitally stable. CNS examination showed, partial ptosis of left eye and dilated non-reactive pupil, restricted upward movement and diplopia. Other cranial nerves were intact and rest of neurological examination was normal. The patient had multiple enlarged left axillary lymph nodes, largest measure 3x3 cms, freely mobile, non-tender with no changes in the overlying skin. There were also enlarged fixed right inguinal lymph nodes nodes, matted together, hard but not-tender. Cervical lymph nodes were not palpable. Chest examination showed equal but reduced air entry bilaterally. No crackles or wheezes. The breasts were symmetrically and

grossly enlarged with flat nipples, no nipple discharge, and no changes in the overlying skin. Both breasts were firm and smooth with mild tenderness. Abdomen was soft and not tenderness. Liver and spleen were not palpable. There was a palpable Left iliac fossa mass extending deep into the pelvis, fixed but non-tender. Biochemistry showed very high LDH (7346 U/L) and uric acid (7.4 mg/dl) together with high fibrinogen level (640.7 mg///dl). Liver functions test were mildy derangement. White cell count was normal at presentation but gradual decrease in platelets count was noted. Renal functions were normal and viral screening including HIV serology was negative. Whole body CT scan with contrast and MRI brain were requested. CT scan revealed enlarged thymus(figure 1), bilaterally enlarged axillary lymph nodes (LNs) with mild bilateral pleural effusion and underlying posterior basal partial collapse together with bilateral breast lesions (figure 2). There were also hepatic, renal and bowel infiltrations, celiac, para aortic and inguinal lymphadenopathy, pelvic masses, mild ascites and small bowel intussusception (figure 3). The liver was mildly enlarged with multiple variable sized hypodense hepatic focal lesions scattered in both hepatic lobes, the largest seen encasing the left portal vein, which shows intra luminal filling defects measuring about 5 x 6.5 cm along its maximum transverse dimensions (figure 4). Lymph nodes biopsy with histopathology and immunophenotyping, together with bone marrow aspiration and trephine biopsy were consistent with Burkett's Lymphoma (BL), (figures 5,6,7,8). Biopsiestaken from the caecum and rectum also showed infiltration with (BL). After confirmation of diagnosis of BL, patient was referred to haematology service. Next day, she receivedcombine hyper CVAD chemotherapy along with uricosuric drugs and intravenous hydration. She received total eight cycles of chemotherapy along with rituximab and intracranial methotrexate. Her condition improveddramatically after the first cycle of chemotherapy. She become afebrile, headache subsided and breast swelling regressed. Her hospital course was complicated with neutropenic sepsis, which was treated successfully with combination of broad spectrum antibiotics, and she was discharge from the hospital with anadvice to continue follow up with a haematologist in her home country.

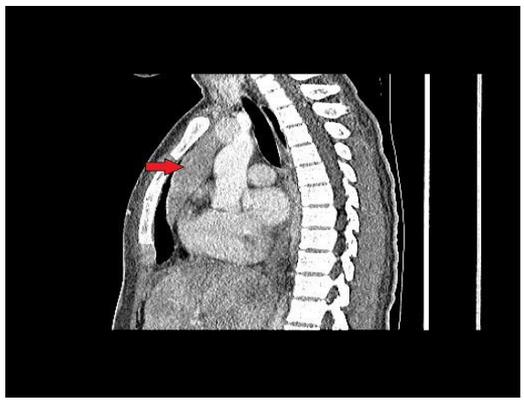

Figure 1

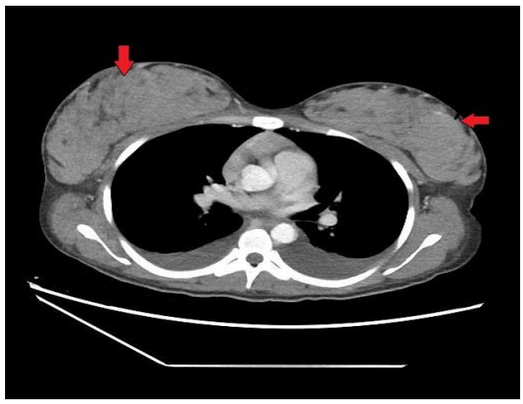

Figure 2

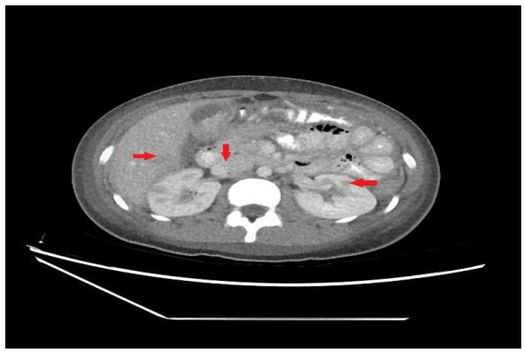

Figure 3

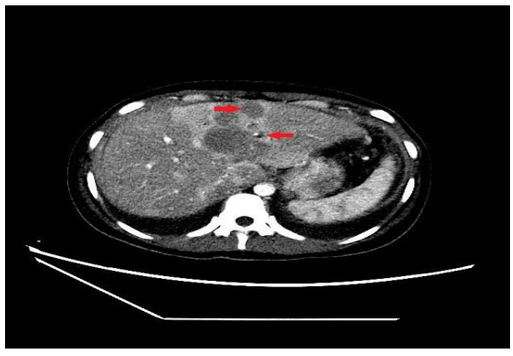

Figure 4

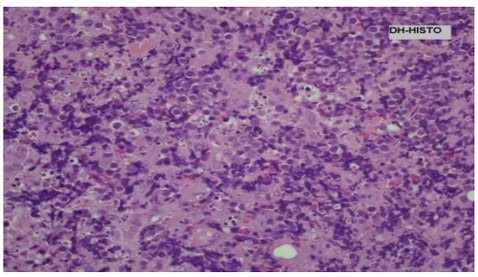

Figure 5

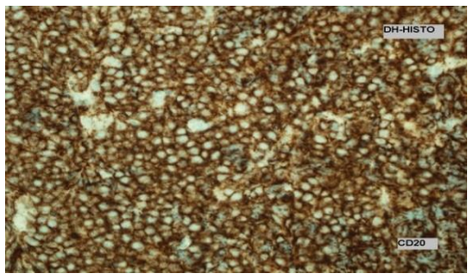

Figure 7

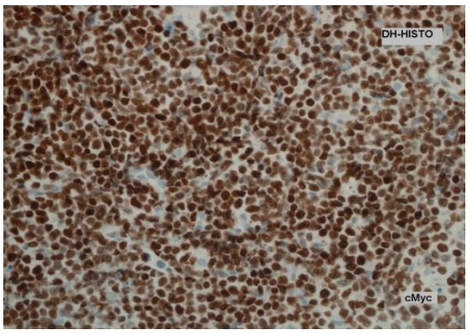

Figure 6

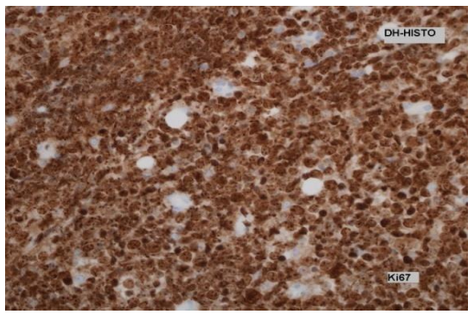

Figure 8

Figure 1: Enlarged thymic gland measuring 2.5 x 5.2 cm along its maximum transverse dimensions presenting heterogeneous CT density.

Figure 2: CT scan thorax: Bilateral lobulated breast densities more evident on the right side associated with skin thickening.

Figure 3: CT scan Abdomen: Hepatic, renal and bowel infiltrations. Celiac, para aortic lymphadenopathy, mild ascites and small bowel intussusception.ith skin thickening.

Figure 4: CT scan abdomen: The liver is mildly enlarged with multiple variable size hypodense bilateral hepatic focal lesions, the largest seen encasing the left portal vein with intra luminal filling defects.

Figure 5: Histopathological section of Lymph node biopsy, showing monotonous population of lymphoid cells with starry-sky appearance. (Haematoxylin and eosin stain) H&E x400)

Figure 6: Histopathological section of Lymph node biopsy section showing diffusely positive cMyc in all lymphoid cells (x400) Figure 7: Histopathological section of Lymph node biopsy is diffusely positive of CD20 in all the lymphoid cells (x400) Figure 8: Histopathological section of Lymph node biopsy showing high proliferative Ki 67 index approaching (100%x400)

TREATMENT: Include pharmacological and non-pharmacological, e.g. surgery, physiotherapy, supportive care. Cyclophosphamide, Cytarabine, Doxorubicin, Dexamethasone, Rituximab, Intracranial methotrexate. Antibiotics', anti-Fungal and other supportive medicines.

DISCUSSION

Burkett's lymphoma (BL) is a very aggressive type of B cell non-Hodgkin lymphoma, its doubling time range 24 to 48 hours[1]. Three clinical variants of the disease are described; the endemic form of Burkett lymphoma (eBL), most commonly seen in equatorial Africa where EBV and malaria are endemic [2], with jaw and facial bone (orbit) involvement.[3] The sporadic form (sBL) most often present with abdominal tumors and bone marrow involvement occurs in more than 50% of cases. However, generalized lymphadenopathy is rare. Other clinical presentations including abdominal masses (ileal, cecal, etc.), as well as ovarian, gonadal, skeletal, and breasts involvement have also been documented in both types.[3] Immunodeficiency-related Burkett lymphoma usually presents as nodal involvement with bone marrow involvement occurs frequently.[3]

The remarkable features in this patient is a combined involvement of CNS involvement in a form of isolated superior divisional 3rd nerve palsy and symmetrical involvement of bilateral breasts which is uncommon in BL. Generalized lymphadenopathy is also not common in sporadic form of BL, which usually presents with abdominal and bone marrow involvement.[3] In this case extensive involvement of the abdominal viscera, including both kidneys, bowel infiltration, with the multiple pelvic masses and sparing of the spleen is an unexpected finding. Breast lymphoma may be a rare disease, either as a primary website or as a secondary involvement, representing zero.04-0.5% of malignant breast growth [4]. the bulk of breast cancer gift as a unilateral painless breast

lots in AN older girl, average age at identification 55-60 years.[4] Most of the reported cases of bilateral breast involvement seen with Burkett's Lymphoma are in pregnant or lactating women. This sugges that a tumor growth can be influenced by hormonal stimulation.[5] The disease in this patient was not associated with pregnancy or lactation. Massive bilateral breast involvement in the course of a Burkett's lymphoma was also reported in pediatrics patients. Plantaz D el at reported a case of Burkett's lymphoma as an exceptional occurrence in a 13 year-old girl, in early puberty.[6] In 1994; J. L. Fahmy el at , reported a case of 16 year old female diagnosed as Burkett's lymphoma with bilateral breasts involvement together with involvement of abdominal viscera and abdominal lymph nodes, disseminated bone lesions and multiple extra-dural masses.[7] CNS involvement in Burkett's Lymphoma (BL) is common with Cranial nerves involvement occurring in cases of gross tumor invasion to the facial or skull bones, and soft tissue, with regional cranial nerves involvement, or with tumor growth along the affected nerve, which is not present in this patient. Cavernous sinus invasion as associate initial presentation of (BL) has been rumored solely seldom. The clinical signs ar those of a cavernous sinus syndrome (CSS), characterised by ocular motor nerves palsies involving the third, fourth and sixth nerves, moreover because the 1st or second divisions of the nervus trigeminus in numerous combos, and orbital pain. The pupil could also be concerned or spared.[8] However, isolated ocular nerve palsy related to cavernous sinus is uncommon.[9] Lesions involving the fascicular, cisternal, sinus cavernosus or orbital parts of the third nerve may result in an exceedingly divisional CN III paralysis. Specifically, superior divisional CN III palsy has been delineate in association with surgical procedure, basilary apex, superior neural structure, and posterior arteria cerebri aneurysms, meningitis, diabetes, sphenoid bone redness, ophthalmoplegic cephalalgia, and a post infectious agent sickness. [10] Isolated 3rd nerve palsy associated with Burkett's lymphoma is extremely rare with only one reported case of Burkett's lymphoma (BL) presenting as a complete third nerve palsy in a 34 year old HIV-positive patient.[11] In my knowledge isolated superior divisional 3r nerve palsy in (BL) has not been reported in litrature before.

CONCLUSION:

1. Burkitt's Lymphoma is a very aggressive type Lymphoma.
2. It can involve multiple body systems simultaneously over a short period of time.

Early diagnosis and treatment can decrease morbidity, mortality and even cure most of patients from this otherwise fatal disease.

CONSENT TO PARTICIPATE: consent was taken from subjects and next of kin

FUNDING: No Funding

ACKNOWLEDGEMENTS: We would like to thank the all contributors and staff and other persons for providing useful information.

AUTHORS' CONTRIBUTIONS: All persons who meet authorship criteria are listed as authors, and all authors certify that they have participated in the work to take public responsibility of this manuscript. All authors read and approved the final manuscript.

CONFLICT OF INTEREST: No competing interest declared.

REFERENCES:

1. D Burkitt . Etiology of Burkitt's lymphoma—an alternative hypothesis to a vectored virus. Natl Cancer Inst, 42 (1969), pp. 19-28
2. D Burkitt A "tumour safari" in east and central Africa. Br J Cancer, 16 (1962), pp. 379-386
3. Ali H. Kanbar, Burkitt's Lymphoma and Burkett's -like Lymphoma, Medscape, Apr. 08, 2016, author:MD; chief editor: Emmanuel C. Besa, MD.
4. Ghasem Janbabai, Shaghayegh Kayedimajd, Shahriar Alian, Farshad Naghshvar, Masoumeh Rashidi, and Touraj Farazmandfar,Bilateral breast swelling in a 23-year-old woman with Burkett's lymphoma. Department of Hematology and Oncology/Cancer Research Center, Mazandaran University of Medical Science (MUMS), Sari, Iran. 1Department of Infectious and Tropical Disease, Mazandaran University of Medical Science (MUMS), Sari, Iran.2Department of Pathology, Mazandaran University of Medical Science (MUMS), Sari, Iran.
5. 5.Tsuyako Saito and Hirokuni Taguchi, Department of Medicine Kochi Medical School, Kochi, Japan. Kensuke Yamamoto, Department of Surgery, Nagasaki University School of Medicine, Nagasaki, Japan. American Journal of Hematology 81:147–148 (2006). Burkitt Lymphoma of the Breast, Isao Miyoshi,
6. Plantaz D, Bachelot C, Dyon JF, Frappat P, Schaerer R, Bost M. Arch Fr Pediatr. 1987

Mar; 44(3):199-200.Massive breast involvement in Burkitt's lymphoma.

7. J.L.Fahmy, B.P.Wood, J.H.Miller, bilateral breast involvement in a teenage girl with Burkitt's Lymphoma, Department of Radiology, Childrens Hospital Los Angeles, Los Angeles, CA 90054, USA, Pediatr Radiol (1995) 25:56-57.
8. Burkitt's Lymphoma with Bilateral Cavernous Sinus Involvement: A Case Report, IBIMA Publishing JMED Research, Vol. 2016 (2016), Article ID 253994, 7 pages, DOI: 10.5171/2016.253994. Monique Boukobza, Department of Neuroradiology and interventional therapeutic, Hôpital Lariboisière, Paris. France. Jean-Philippe Brouland, Hopitaux de Paris; Université Paris Sorbonne Cité,Groupe hospitalier Lariboisière, Paris. France.
9. 2.Seong-Min Choi*, Joon-Tae Kim, Seung-Han Lee, Man-Seok Park, Byeong-Chae Kim, Myeong- Kyu Kim and Ki-Hyun Cho, Chonnam Isolated Oculomotor Nerve Palsy due to Non-Hodgkin's Lymphoma Demonstrated by Serial MRI,Medical Journal, Vol. 44, No. 2, pp. 109~112.
10. M. Tariq Bhatti, MD; Stephen Eisenschenk, MD; Steven N. Roper, MD; et al.Superior Divisional Third Cranial Nerve Paresis Clinical and Anatomical Observations of 2 Unique Cases, Arch Neurol. 2006; 63(5):771-776. doi:10.1001/archneur.63.5.771.
11. Levy J1, Kratz A, Lifshitz T. Burkitt's lymphoma presenting as oculomotor palsy in an HIV-positive patientDepartment of Ophthalmology, Soroka University Medical Center, Ben-Gurion University of the Negev, Beer-Sheva, Israel. ljaime@bgu.ac.il