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Retroperitoneal extra-adrenal non-Hodgkin lymphoma: An uncommon presentation

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ABSTRACT

Primary retroperitoneal non Hodgkin lymphoma is extremely rare, its diagnosis is often difficult and it may requires a time consuming and a costly diagnostic workup. We report the case of a 46-year-old patient complaining of abdominal fullness and dorsal pain, who was diagnosed with an extra-nodal non-Hodgkin lymphoma presenting as a unique and large retroperitoneal mass. The suggested diagnosis was a malignant retroperitoneal tumor and the patient underwent an excision of the tumor throw a lombotomy followed by an R-CHOP chemotherapy regimen with good outcome.

Introduction

Non-Hodgkin lymphoma is considered to be one of the most common malignancies of the retro peritoneum, but this location has been very rarely reported as the primary site of the disease. Here, we present the case of a male patient with a retroperitoneal extra adrenal non Hodgkin lymphoma.

Case presentation

A 46-year-old diabetic man, presented to our department with progressively increasing pain and fullness in the left upper quadrant of his abdomen. On examination, he had a blood pressure of 130/80 mmHg with no postural drop, a pulse rate of 86 beats/minute, with no palpable mass on abdominal examination, there was no hepatomegaly, or splenomegaly and no palpable lymph nodes. No alteration was found in the standard hemato chemical investigations. CT scan revealed a heterogeneously enhanced soft tissue retroperitoneal mass, it measured 13 × 12 cm. The kidney and left adrenal gland were normal, there was no splenomegaly or abdominal lymphadenopathy (Fig. 1). In front of tomographic characteristics of the tumor and its size, the suggestive diagnosis was of malignant retroperitoneal tumor, we decided to excise the tumor throw a lombotomy, per operative findings revealed an adherent large tumor to the adrenal gland, thus we decided to perform a left adrenalectomy with excision of the tumor. Histopathological analysis revealed diffuse large B-cell lymphoma, immunohistochemical stains were strongly positive for CD20 (Fig. 2).

For further management, the patient was referred to our hematology clinic and was planned for R-CHOP chemotherapy regimen. After 6 cycles of chemotherapy he was in good condition with no evidence of relapsing lymphoma after 12 months follow up on scan.

Discussion

Extra nodal lymphoma occurs in approximately 40% of all patients with lymphoma, extra nodal disease is more common with non-Hodgkin lymphoma (NHL). It causes many deaths worldwide, and its incidence is increasing. It represents a heterogeneous group of neoplasms originating from lymphocytes. Diffuse large B-cell lymphoma (DLBCL) is the most common histological NHL subtype in adult patients.[1] Although lymphomas are considered to be one of the most common malignancies of the retro peritoneum, this location has been very rarely reported as the primary site.[2]

Due to the uncommon anatomical location and the lack of symptoms in many of them for a longtime, the diagnosis is usually late and the management of this patient is difficult. Although CT scan is the diagnostic modality of choice,[3] magnetic resonance imaging (MRI), offers superior soft tissue contrast in comparison with CT, they are necessary for the diagnostic because they allow the lesion localization and characterization. Biopsy or tumor resections are the tools that allow the definitive diagnosis.[4] Indolent NHL is generally considered incurable. Several regimens have been commonly used; however, treatment

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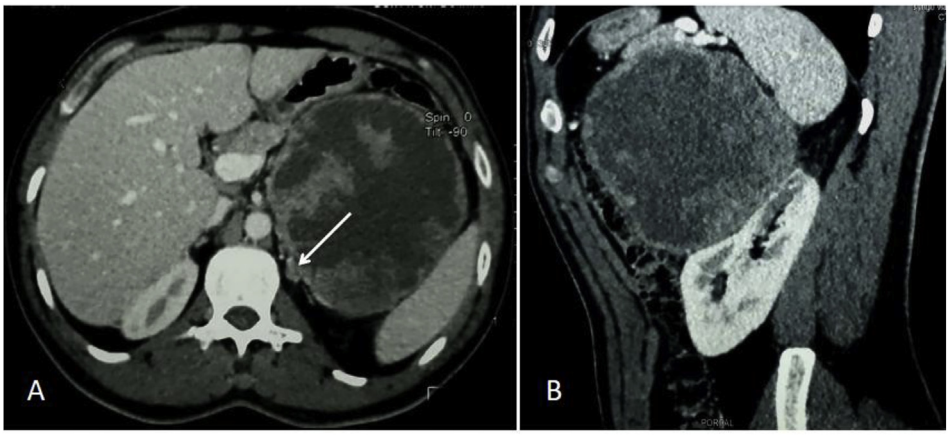

Fig. 1. A: Abdomino pelvic CT scan showing a well delineated heterogeneous left suprarenal retroperitoneal mass and the left adrenal gland (arrow) that adhering to the tumor B: sagital section shows the tumor that was pressing on the homolateral kidney and the adrenal gland.

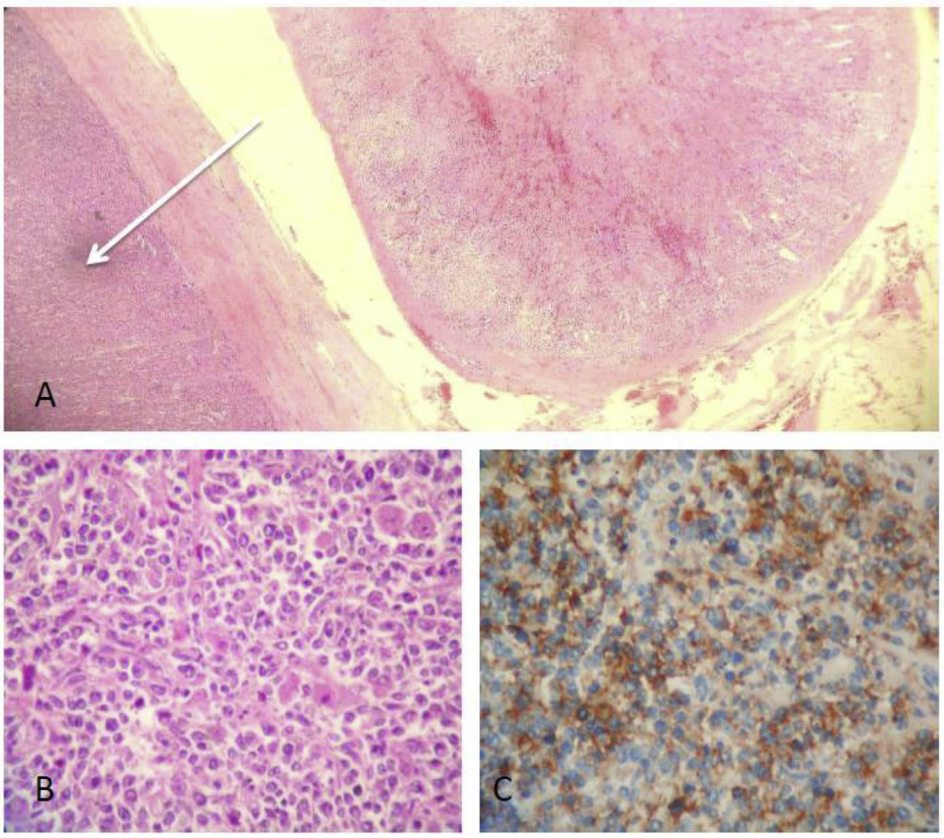

Fig. 2. A: (HES X 4) Showing an encapsuled lymphoid proliferation with diffuse growth pattern and large cells (arrow), the adrenal gland is normal (on the right). B: (HES x 10) The malignant lymphocytes are very large with a moderately abundant cytoplasm, and the nuclei are round to ovoid with prominent nucleoli and occasional mitoses. C: Immunohistochemical staining strongly positive for B-cell marker CD20.

has never been shown to extend overall survival. The poly-chemotherapy with CHOP represents the standard chemotherapy regimen for NHL treatment with good outcome, and few and acceptable side-effects.[2] New strategies with retuximab, a chimerical anti CD20 IgG1 monoclonal antibody which is a cell surface protein that occurs almost exclusively in mature B-cells, are used to improve the prognosis of these patients.[5] The prognosis has improved in recent years owing to the development of various aggressive chemotherapeutic regimens depending on the histological type, stage and age of each patient. Complete response is obtained in about 45–53% of cases with long term survival of 30–37%.[2]

Conclusion

Primary retroperitoneal DLBCL is extremely rare, and has a variable and non specific presentation and many resemble other neoplasic or inflammatory conditions. Obtaining a definitive histological diagnosis by biopsy or surgery resection and using and using appropriate chemotherapy are essential for recovery and long term survival of these patients.

Conflicts of interest

The authors declare that they have no conflict of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2018.12.003.

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