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NODAL T FOLICULAR HELPER CELL LYMPHOMA ANGIOINMUNOBLASTIC TYPE TREATED AS A CLASSIC HODGKIN LYMPHOMA

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INTRODUCTION

Nodal T follicular helper cell lymphoma, angioimmunoblastic type (nTFHL-AI), is a neoplasm of mature T cells with a TFH phenotype, characterized by systemic disease and a polymorphous lymphoid infiltrate, prominent proliferation of high endothelial venules and follicular dendritic cells.

AIM

There are a wide range of differential diagnoses including polymorphic lymphoproliferative disorder, large B-cell lymphoma, plasmacytoma, marginal zone lymphoma, and classic Hodgkin lymphoma.

Accurate diagnosis of nTFHL-AI can be difficult due to its diverse clinical symptoms and wide histopathologic changes.

METHOD

A 62 years old was diagnosed of mixed cellularity Hodgkin Lymphoma stage IIE A (infradiaphragmatic with psoas infiltration). The fifth lines of treatment consisted on: ABVD 6 cycles; ESHAP X 3 cycles, Autologous Transplantation, Radiotherapy and 16 sessions BV. PEC TC after every treatment line showed disease progression (Fig. 1a) . Due to the poor clinical evolution, a new biopsy was performed and reviewed by other pathologist. A nTFHL-AI be conclusive (Fig. 2). A 6th line: Belinostat started with a good response (Fig. 1b) (Table1).

RESULTS

nTFHL-AI is a disease of middle-aged and elderly individuals with a slight male predilection. nTFHL-AI accounts for 36% of the non-cutaneous T-cell lymphomas. Characteristic features at presentation include generalized lymphadenopathy (typically <1-3 cm), hepatosplenomegaly, skin rash and immune dysregulation, resulting in autoimmune complications and immunodeficiency with opportunistic infections. nTFHL-AI course is generally aggressive (median survival of 3 years and a 5-year overall survival rate of approximately 30%).

Accurate diagnosis of nTFHL-AI can be difficult due to its diverse clinical symptoms and wide histopathologic changes. One of the common challenges is identifying “early” nTFHL-AI cases (type I pattern) and separating them from reactive paracortical hyperplasia. B-cell proliferations that accompany nTFHL-AI may also pose a challenge in the differential diagnosis. The presence of HRS-like cells may lead to misdiagnosis as CHL. Distinguishing nTFHL-AI from other nodal PTCL-TFHs can be particularly difficult, although it currently has no impact on clinical management.

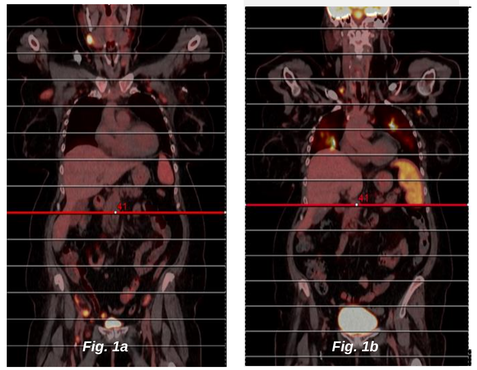


Figure 1: a) PET showed disease progression ; b) PET after 2 cycles of Belinostat and bilateral cytomegalo-virus pneumonia.

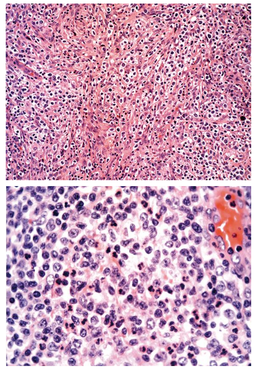
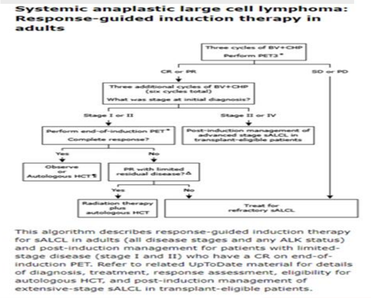


Figure 2: a) Lymph node biopsy shows an Angioimmunoblastic T cell lymphoma with arborizing blood vessels and large tumor cells with clear cytoplasm. H&E, x20; b) Angioimmunoblastic T cell lymphoma with polymorphous infiltrate. Higher magnification shows an area with mixed cellularity. H&E, x60.

Table 1: Treatment of relapsed or refractory peripheral T cell lymphoma.

Chemotherapy
1. **Traditional regimens:** patients who are transplant candidates may respond to traditional regimens used for - ICE (ifosfamide, carboplatin, etoposide) - DHAP (dexamethasone, high dose cytarabine, and cisplatin) - ESHAP (etoposide, methyl-prednisolone, cytarabine, and cisplatin - Single-agent gemcitabine - GDP (gemcitabine, cisplatin and dexamethasone) - Gemcitabine and oxiplatin
2. **Newer agents:** our choice among the newer agents is generally guided by the specific subtype of PTCL and the toxicity profile of the agents. - Pralatrexate - Belinostat - Brentuximab - Romidepsin
Transplantation
Autologous versus allogeneic HCT Allogeneic HTC Autologous HCT



CONCLUSIONS

Given the complex heterogeneous nature of nTFHL-AI, a multiparameter approach with careful integration of clinical history, morphology, and immunophenotypic findings, supplemented by molecular/genetic analysis as necessary, is essential to make an accurate diagnosis.

For pathologists, diagnosing nTFHL-AI may be challenging due to its wide clinical and histopathologic spectrum, which can mimic a variety of reactive and neoplastic processes.

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