A rare presentation of Posttransplant Lymphoproliferative Disease (PTLD) in a pediatric liver transplantation patient: Plasmablastic lymphoma complicated with hemophagocytic lymphohistiocytosis

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Introduction

Background

- Plasmablastic lymphoma (PBL) is a very rare cause of PTLD.
- It was first described as a EBV-associated B-cell neoplasm in (HIV) patients
- PBL was also reported as a rare cause of PTLD

Importance

- We present a 3 year old female patient diagnosed as PBL after liver transplantation which makes our case the second pediatric case of PBL as PTLD
- Furthermore our case was complicated with hemophagocytosis

Case presentation

Patient profile

A three year old female- liver transplanted from her father for biliary atresia 1 year ago
Admission with fever and pancytopenia
From hx: EBV PCR (+) 3 months ago and RIS was administered (tacro…sirolimus)

Clinical findings

Fever
Cervical,axillary,inguinal LAP 2 cm
Splenomegaly of 5 cm
Hepatomegaly of transplanted liver

Diagnostic workup

Hb: 6,8 g/dL, WBC: 3,24x10 ^{3}/µL, Plt: 68x10 ^{3}/µL
LDH: 1623 U/L uric acid: 8.5 mg/dl.
EBV PCR - 5 X10^{4} copies/ml.
Ferritin: 5291 µg/L
TG: 240 mg/dL

Case presentation

Radiology

- Lympadenopathies in bilateral parailiac, mesenteric, cervical, axillary, paratracheal, pretracheal, subcarinal and hilar regions with a maximum diameter of 3x3 cm
- PET-CT: Multiple lymph nodes showing increased FDG uptake were observed in the bilateral cervical chain, left supraclavicular region, more pronounced on the left side, bilateral axillae, left retropectoral area, bilateral paramammary region, mediastinum, bilateral hilar regions, left anterior peridiaphragmatic region, and abdominopelvic region
- Bone marrow aspiration: Increased histiocytes and hemophagocytosis

Case presentation

Pathology

LN bx: Monomorphic post-transplant lymphoproliferative disorder with plasmablastic lymphoma phenotype’ with neoplastic cells exhibiting widespread strong positivity with CD138 and CD79a, focal positivity with EBER, Ki-67 proliferative activity of 90% and negativity for CD20, Pax 5, CD68, CD30, CD15, CD3, CD2,CD1a, CD4, CD8, CD34,CD56 and TdT.
Lambda clonality +
t(11/14) and t(14/18) (-)

Discussion

Diagnosis

She was diagnosed Plasmablastic Lymphoma Type-Monomorphic Post Transplant Lymphoproliferative Disorder (PTLD) leading to secondary Hemophagocytic Lymphohistiocytosis (HLH)

Treatment

RIS: Tacrolimus to sirolimus
4 cycles of Rituximab +6 cycles of CHOP chemotehrapy
4 doses of IVIG for HLH therapy every 21 days
In remission after 6 cycles
PET CT at cessation of chemotherapy:CR

Discussion

PBL is very rare after SOT
It is even more rare in children with SOT
We found only one similar case after small bowel & Liver transplant in an infant.
Furthermore, our patient also had HLH clinics at admission
This is a new clinical situation not presented previously

Conclusion

Plasmablastic lymphoma (PBL) is a rare and aggressive type of Non-Hodgkin lymphoma typically seen in the settings of an immunocompromised state, classically associated with HIV infection. Very rarely, plasmablastic lymphomas may be seen as a type of post-transplant lymphoproliferative disorder

Furthermore, our patient is also admitted with secondary HLH complicating PTLD more. Despite the aggresiveness of PBL-PTLD in adults, there is no report showing prognosis of pediatric patients. The rapid response to therapy made us think positive for the prognosis of the patient, however it is not easy to forsee the prognosis in pediatric patients.More stuides on PTLD-PBL pediatric patients are warrented.

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