Profuse telangiectasias in an immunocompetent patient misleading presentation revealing a hepato-splenic-Tγδ-cell lymphoma

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Abstract

Here we present the case of an hepato-splenic-Tγδ-cell lymphoma interestingly occurring in a non-immunocompromised patient, with profuse telangiectasias giving originally misleading orientation towards the diagnosis of B angiotropic lymphoma.

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Authorship Contributions:

AD, BC, LT, KL and MMF managed the patients, and provided samples and clinical data. RD end SP performed anatomopathological and biological analysis. AD, and MMF wrote the manuscript, which was approved by all the authors.

Consent & Agreement:

The patient was informed and gave permission to publish the case.

Conflict of interest :

The authors declare that they have no conflict of interest.

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Case report

A 50-year-old female with no significant past medical history presented with hectic fever, weight loss, asthenia and abdominal pain 3 weeks before admission in our department. Closer examination revealed multiple telangiectasias located on the chest and upper back, with proximo-distal extension (Fig.1). Physical examination displayed a voluminous hepato-splenomegaly. The biological analysis showed a hemophagocytic syndrome (HS) with inflammatory syndrome (CRP 41 mg/L). A small population of Double negative abnormal T population CD2+ CD3+ CD4- CD8- CD5-CD56- CD16+ CD17+ accounting for 27.9% of total lymphocytes, presenting γδTCR was identified using flow cytometry. Medullar karyotype identified 44,X,-X,+8,add(10)(q26),-11,-21[6]/46,XX[14]. Plasma Vascular Endothelial Growth Factor (VEGF) level was significantly elevated (1140 pg/mL, normal value inferior to 500 pg/mL). A mutation of*STAT5B* T628S was identified by molecular biology. Computed tomography (CT) scan showed voluminous hepatosplenomegaly. Positron emission tomography-CT revealed diffuse hypermetabolism in the hepatosplenomegaly and regarding the osteomedullary area. The course was marked by a worsening of the abdominal pain revealing a subcapsular intraparenchymal spleen rupture. The splenectomy associated with liver biopsy concluded Hepatosplenic T CD4-/CD8- lymphoma stage IVBb (Fig 2.). Remarkably, the patient's global state improved after splenectomy. After 2 lines of chemotherapy, she received allogeneic stem cell transplantation from an unrelated donor.

Here we present the case of an hepato-splenic-Tγδ-cell lymphoma interestingly occurring in a non-immunocompromised patient, with profuse telangiectasias giving originally misleading orientation towards the diagnosis of B angiotropic lymphoma. We observed an increased blood VEGF level that can be possibly correlate with clinical telangiectasias. Gamma-delta-hepato-splenic T-cell lymphoma is a rare entity of primary extranodal disease and represent less than 5% of all PTCL cases. The prognosis of this lymphoma is poor requiring multiple lines of chemotherapy and allogeneic stem cell transplantation.

References

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Figures legends :

Fig 1. Profuse Telangiectasias on the patient's chest

Fig 2. (A) Spleen involvement with sheets of small neoplastic lymphoid cells involving cords and sinuses of spleen (hematoxylin and eosin [H&E] stain; original magnification ×600). (B) TiA1 immunohistochemical stain in spleen, highlighting the nonactivated cytotoxic phenotype of neoplastic T cells (original magnification ×600). (C) CD3 immunohistochemical stain in liver biopsy, highlighting the sinusoidal expansion by neoplastic T cells (original magnification ×600).

Fig 3. Perioperative splenectomy (9.8 * 7.9 inches)