Diffuse large B-cell lymphoma associated with chronic inflammation in a patient with aneurysmal thrombus of the abdominal aorta

Lucile Perrin, ¹ Laure Gibault, ² Ismael Boussaid, ¹ Rudy Birsen ¹

Correspondence to Dr Rudy Birsen; rudy.birsen@aphp.fr

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DESCRIPTION A man in his 70s complained of abdominal pain, weight loss and night sweats. He had a medical history of type 2 diabetes, lower extremity arterial disease, chronic obstructive pulmonary disease, ischaemic stroke, abdominal aortic aneurysm known for 13 years (figure 1A), high blood pressure and ischaemic heart disease. Five months earlier, he presented with a partial thrombosis of his aneurysm of the diaphragmatic and abdominal aorta complicated by an occlusion of the coeliac trunk (figure 1B). Considering the prefissure aspect of the aneurysm, the patient benefited from an open aneurysm repair with replacement of the diseased aortic segment with a bifurcated prosthetic graft. In the following months, the patient presented with abdominal pain and diarrhoea. The biological workup showed an inflammatory syndrome with a C reactive protein of 30 mg/L (n<5 mg/L), a microcytic anaemia with a haemoglobin of 99 g/L and lactate deshydrogenase of 545 U/L (N=135-225 U/L). A thoracic and abdominopelvic CT scan was performed, allowing the discovery of a necrotic abdominal mass (figure 1C). There were local lymphadenopathies of the coeliac and mesenteric chains, without supradiaphragmatic lymph node involvement. Biopsy of the mass was performed through an echo-endoscopy. Pathology revealed a diffuse large B-cell lymphoma (DLBCL, figure 1D-F). A review of the pathology specimen of the aneurysmal thrombus taken 5 months earlier revealed a fibrin-associated DLBCL (figure 1G,I). An Epstein-Barr Virus (EBV) replication was detected with a blood viral load measured at 4.75 log (55 600 copies/mL). Given the acute and threatening symptomatology, the patient did not have time to undergo a positron emission tomography (PET) scan. There was no central nervous system invasion on the lumbar puncture and brain imaging. The patient was treated with a course of six R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) cycles, allowing a complete response. Notably, a PET scan at the end of treatment showed no fixation.

DLBCL associated with chronic inflammation is a lymphoid neoplasm occurring in the setting of long-standing chronic inflammation. The putative pathophysiological mechanism is a local inability of the immune system, due to long-standing chronic suppuration or inflammation, to eliminate polyclonal EBV-transformed B cells, which will ultimately lead to transformation into a monoclonal

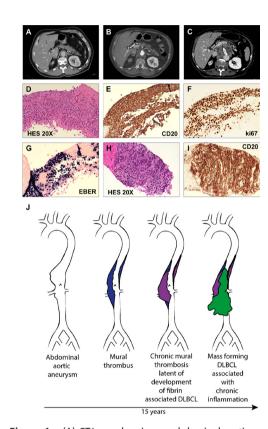


Figure 1 (A) CT/scan showing an abdominal aortic aneurysm with mural thrombus in 2009. (B) CT/scan showing abdominal aortic aneurysm with mural thrombus in 2022. This aneurysm was measured at 57 mm in axial sections and 56 mm in diameter in the transverse axis of the aorta, which extended approximately 170 mm in height. (C) CT/scan showing necrotic abdominal mass measuring 72×59 mm, infiltrating the mesenteric root and encompassing the previously stented superior mesenteric artery. (D-G) Initial thrombus with fibrin-associated DLBCL. (D) HES routine staining, ×20 magnification lens evidencing a focal proliferation of large lymphoid cells in fibrin. (E) B-cell lineage demonstrated by CD20 diffuse staining. (F) High proliferative index demonstrated by Ki67 staining, (G) EBV association demonstrated by Epstein-Barr encoding region (EBER)-positive in situ hybridisation. (H,I) DLBCL associated with chronic inflammation. (H) HES staining, ×20 magnification lens demonstrating the same large cell proliferation, this time forming a mass on imaging. (I) CD20 staining demonstrating B-cell lineage. (J) Development of DLCBL associated with chronic inflammation. IB and LG created figure 1A-H, and IB created figure 1J. DLBCL, diffuse large B-cell lymphoma; HES, H&E saffron.



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¹Hematology, APHP, Paris, France

²Pathology, Hopital Europeen Georges Pompidou, Paris, France

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B-cell proliferation such as DLBCL.¹ DLBCL associated with chronic inflammation is an aggressive lymphoma requiring chemotherapy and/or radiotherapy, with a 5-year survival of 20%–35%.²

Fibrin-associated DLBCL is an unusual form of DLBCL associated with chronic inflammation. This type of lymphoma is not mass forming and is usually asymptomatic. The discovery is often incidental on surgical specimens of various pathologies such as pseudocysts, subdural haematomas, hydroceles or materials of the cardiovascular system. Fibrin-associated DLBCL is presented as a different entity from DLBCL associated with chronic inflammation, mainly because of its incidental finding, and the generally favourable evolution after surgical resection alone.³

Abdominal aortic aneurysms are associated with atherosclerosis, transmural degenerative processes, neovascularisation, decrease in content of vascular smooth muscle cells and a chronic infiltration, mainly located in the outer aortic wall.⁴ In the case presented here, the chronic inflammation of the aortic

Learning points

- ➤ Fibrin-associated diffuse large B-cell lymphoma (DLBCL) is associated with chronic inflammation, does not form a mass and is most often found incidentally.
- Progression to an aggressive form with a poor prognosis, called DLBCL associated with chronic inflammation, is possible and should be suspected in case of progression to an infiltrative mass after surgical resection.
- Epstein-Barr Virus (EBV) is central to the pathophysiology of inflammation-associated lymphomas.

aneurysmal thrombus has probably created a local immunosuppression that led to the development of fibrin-associated DLBCL of slow and chronic evolution. However, a clone of this fibrin-associated lymphoma probably developed into a more aggressive entity before surgical treatment of the aneurysm was performed. Residual cells may have evolved secondarily into an infiltrative tumour, forming an abdominal mass with a clinical and pathological presentation compatible with the diagnosis of DLBCL associated with chronic inflammation as defined by the WHO 2016 classification (figure 1J).

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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