CASE REPORT

Fibrin Associated Epstein—Barr Virus Positive Large B Cell Lymphoma as a Complication of a Repaired Thoraco-abdominal Aortic Aneurysm

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WHAT THIS PAPER ADDS
The case of a patient with an untoward outcome after repair of his thoraco-abdominal aneurysm is reported. He was treated for an anastomotic pseudoaneurysm and for multiple visceral aneurysms of new onset. Infection was suspected however, it could not be confirmed. Finally, the post-mortem revealed a fibrin associated Epstein—Barr virus positive diffuse large B cell lymphoma, a very rare malignancy involving the aorta. Although infection is common in patients with a bad outcome after aneurysm repair, other diagnoses must be considered.

Background: Malignancies involving the aorta are extremely rare.
Case report: A 62 year old man with a history of open repair of a thoracic aortic aneurysm developed new visceral aneurysms and an anastomotic pseudoaneurysm. Fludeoxyglucose positron emission tomography/computed tomography, performed for suspicion of graft infection, found abnormal uptake around the bypass, the visceral aneurysms, and the femoral arteries. The patient was treated by embolisation of the visceral aneurysms and a thoracic endovascular aortic repair. Several bleeding complications occurred, and the patient died. Post-mortem revealed a fibrin associated Epstein—Barr virus positive diffuse large B cell lymphoma.
Conclusion: Patients with aortic aneurysms can develop malignancies including fibrin associated lymphoma. Clinical manifestations may be similar to those of an infectious process; timely diagnosis is uncommon.

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INTRODUCTION
Malignancies involving the aorta are extremely rare. They occur primarily in the abdominal aorta in association with an aneurysm or mimicking one. A case of a fibrin associated Epstein—Barr virus positive diffuse large B cell lymphoma (FA-DLBCL) is presented in a patient with history of thoraco-abdominal aortic aneurysm that had been treated by open repair. Written consent was obtained from the patient.

CASE REPORT
The patient was a 61 year old man, a former smoker, with a history of hypertension, dyslipidaemia, and a duodenal ulcer treated by partial gastrectomy. He was referred with a 60 mm thoraco-abdominal aortic aneurysm and a thrombosed coeliac trunk aneurysm.

An end to end interposition graft was performed from the descending thoracic aorta to the aorta above the superior mesenteric artery using a 30 mm Dacron graft. The coeliac trunk was ligated.

The patient remained asymptomatic for six months. He then developed fever, asthenia, and had high levels of C

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reactive protein. He was extensively evaluated for all possible infectious sources. Serologies were negative for antineutrophil cytoplasmic antibodies (ANCA), antinuclear antibody (ANA), and rheumatoid factor. A thoracic computed tomography (CT) scan revealed thickening around the graft. Because of the suspicion of graft infection, a fludeoxyglucose-positron emission tomography/computed tomography scan was performed, revealing abnormal uptake by the bypass graft, coeliac trunk, splenic artery, and both femoral arteries (Fig. 1). Samples of the left femoral artery and surrounding lymph nodes were tested. These were negative for abnormal cells and for bacteria. Empirical antibiotic treatment (amoxicillin and clavulanic) was prescribed for six weeks.

The patient remained pyrexial and asthenic, and finally, fifteen months after the surgery, he presented with haemoptysis. CT revealed new aneurysms: a 26 mm right upper pole renal artery aneurysm, a pseudo-aneurysm in the proximal aortic anastomosis, a 23 mm left common femoral artery aneurysm, various <10 mm pancreatic artery aneurysms, and a 15 mm splenic aneurysm with a large haematoma around the spleen (Fig. 2).

In an initial endovascular procedure, Onyx 34 and coil embolisation of the right upper renal pole aneurysm and coils and Glubran 2 embolisation to the splenic artery were performed. For the aortic pseudoaneurysm, two thoracic endografts, Bolton Relay Plus 36 × 36 × 250 and Bolton Relay Plus 36 × 36 × 100 (Bolton Medical, Sunrise, FL,

Figure 1. Fludeoxyglucose-positron emission tomography/computed tomography was performed, showing abnormal uptake of the bypass, coeliac trunk, splenic artery, and both femoral arteries.

Figure 2. Computed tomography scan: 3D reconstruction. (A) 26 mm right upper pole renal artery aneurysm. (B) Pseudoaneurysm in the proximal aortic anastomosis. (C) 23 mm left common femoral artery aneurysm.
USA), were placed from below the subclavian ostium to the supramesenteric portion of the aorta (Fig. 3).

Genetic analysis revealed the presence of the NOTHC 1 gene that is related to Marfan syndrome. Nevertheless, because there were no other pertinent genetic findings and no phenotypic associations, this diagnosis was ruled out.

Empirical endovenous antibiotic treatment with meropenem and vancomycin was given for four weeks and oral levofloxacin and clindamycin were given for three months. Thereafter, the patient was admitted to the emergency room in haemodynamic shock. A new CT scan showed major haemorrhage after rupture of the previously treated renal aneurysm, requiring massive blood transfusion and repeat embolisation. In the intensive care unit, he developed intestinal bleeding from the right colic artery, firstly treated by embolisation and finally by caecectomy and ileostomy. The patient recovered however he suffered a left femoral aneurysm rupture. Ligation and femoropopliteal bypass with a reversed saphenous vein were performed. Pathological findings from the bowel specimen showed arterial thrombosis. The femoral specimen showed cells positive for the CD20 receptor. Despite all these treatments, the patient died.

The autopsy revealed a fibrin associated Epstein–Barr virus positive diffuse large B cell lymphoma. Large B cells were found in relation to the aortic prosthesis, the visceral aneurysms, and the renal arteries. In situ hybridisation on lymphoma cells showed positivity for Epstein–Barr virus and the diagnosis of a FA-DLBCL was made (Figs. 4 and 5).

**Figure 3.** Computed tomography reconstruction of thoracic aorta with two Bolton Relay Plus® thoracic endografts of 36×36×250 cm and 36×36×100 cm.

**Figure 4.** Fibrin associated diffuse large B cell lymphoma arising in an endovascular aortic prosthesis (A, haematoxylin and eosin (H&E) ×40). The prosthesis is surrounded by fibrinous material and some atypical lymphoid cells (B, H&E ×100). Secondary branches are also affected by the lymphoid proliferation admixed with fibrin (C, H&E ×200). Neoplastic cells are large, with vesicular chromatin, prominent nucleoli and amphophilic cytoplasm (D, H&E ×400).
DISCUSSION

Lymphomas are believed to arise from lymph nodes or lymphoid tissue associated with other organs. Large B cell lymphoma is the most prevalent. Lymphoma in unusual clinical scenarios has been reported including cases of DLBCL arising in prosthetic heart valves, metallic implant, surgical mesh implants, and chronic skin ulcers.

DLBCL comprise a group of relative common haematological malignancies. They may be associated with chronic inflammation, now considered a rare EBV associated subtype in immunocompetent individuals.

EBV positive DLBCL have also been reported as an incidental finding in the setting of chronic haematomas, atrial myxomas, and pseudocysts. FA-DLBCL tends to be confined to the primary disease site, warranting separate classification. The World Health Organisation describes FA-DLBCL as a non-mass forming malignancy found in the walls of pseudocysts. Most cases behave indolently with the potential for cure by surgery alone. Nevertheless, primary cardiac or vascular disease may pose a higher risk of recurrence despite systemic chemotherapy. Embolisation of malignant cells could have played a role in the untoward outcome of this case.

Given the patient’s non-specific presentation, graft infection was presumed, because this is the most frequent complication. FA-DLBCL is a very rare pathology, and timely diagnosis is not common. These patients typically succumb to the lymphoproliferative process and its complications.

CONFLICTS OF INTEREST
None.

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