Clinical Image

A Double Whammy of Mycotic Aneurysms and Acquired Dysfibrinogenemia in a Patient with Septicaemia

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A 65-year-old woman with IgA nephropathy presented with a two-day history of fever and right lower limb swelling. She had been treated with cyclophosphamide for a recent relapse five days preceding this current presentation. Apart from a body temperature of 38°C, her other vital signs were fairly stable. Her right lower limb, in particular the thigh, was swollen, erythematous and mildly tender. Review of other systems was unremarkable. Blood tests revealed elevated septic markers and group A beta haemolytic streptococci from the blood culture. She was treated with intravenous ampicillin/sulbactam for bacteraemia. On the 12th day of hospitalization, her right thigh became more swollen and painful. The computed tomography (CT) showed a large heterogeneous retroperitoneal haematoma measuring 4.1 cm x 14.2 cm x 17.2 cm, and another haematoma within the right gluteus medius muscle measuring 8.5 cm x 14.6 cm x 30 cm (Figure 1). The conventional angiogram showed an aneurysm from the right inferior pancreaticoduodenal artery with active bleeding, which was successfully secured by arterial embolization (Figure 2A, B). The right gluteal region was not intervened in the hope of achieving haemostasis through the tamponade effect of the haematoma. The embolization procedure was complicated with persistent oozing from the femoral puncture site. Repeated activated partial thromboplastin time (aPTT) was prolonged (> 180 seconds), which was not corrected by the mixing test. Coagulopathy work-up showed markedly prolonged thrombin time (TT) of 216.8 seconds, fibrinogen level of 2.14 g/L (normal range: 1.36 g/L - 4.65 g/L), elevated levels of factors VIII and IX of 384 % and 185 % respectively. Acquired dysfibrinogenemia was highly suspected. The fibrinogen activity-antigen ratio was not performed as she was not keen. She was then treated with multiple transfusions of fresh frozen plasma (FPP), cryoprecipitate, packed cells and desmopressin to achieve haemostasis. On the 16th day, she was found to have active bleeding from an aneurysm arising from a branch of the right inferior gluteal artery, which was then successfully treated with arterial embolization (Figure 2C, D). She was discharged on the 26th day of admission without any major complications. Written informed consent was obtained from the patient.

Staphylococci and streptococci are the most common causative pathogens for mycotic aneurysms.[1] Aneurysms due to group A streptococcal septicaemia usually affect the large vessels such as the aorta.[2] Our patient developed two aneurysms along the right inferior pancreaticoduodenal and right inferior gluteal arteries, which were relatively uncommon. The management of an infected aneurysm is

individualized, and largely depends upon the characteristics of the aneurysm and patient. Treatment options include open surgery, endovascular stent placement, endovascular embolization, medical therapy, or a combination of any of these.[3,4] Any abnormality of fibrinogen can cause haemorrhage, thrombosis or both. Dysfibrinogenemia is a condition associated with prolonged TT or low fibrinogen level. If TT is prolonged, fibrinogen activity–antigen ratio is performed to diagnose dysfibrinogenaemia.[5] Acquired dysfibrinogenemia occurs most often in patients with severe liver disorder producing abnormal fibrinogen molecules. Dysfibrinogenemia may also be associated with cancer, most commonly being liver tumours. Autoantibodies inhibiting specific functions of fibrinogen have been described in systemic lupus erythematosus, ulcerative colitis and multiple myeloma.[6] Our patient likely had autoantibodies interfering with the fibrinogen activity due to her recent relapsed IgA nephropathy. This was further supported by her prolonged aPTT, which was not corrected with the mixing test. Concurrent occurrence of these two medical conditions in a patient is extremely rare, and to our knowledge, has never been reported. This case highlights the importance of suspecting aneurysm related bleeding in uncommon locations to avoid treatment delay in potentially reversible and life-saving conditions.

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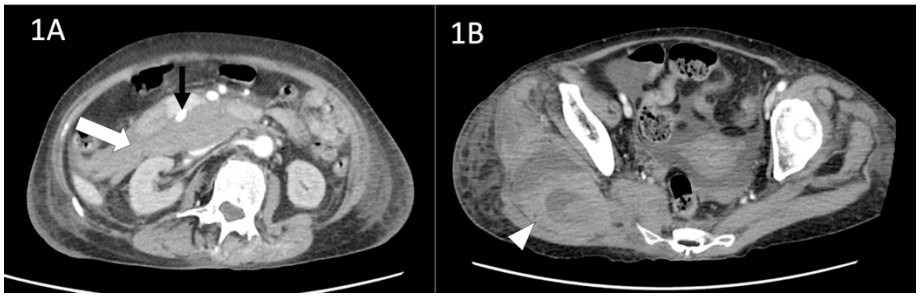

FIG. 1. Contrasted computed tomogram (CT) of the abdomen and pelvis in the portal venous phase. Figure 1A shows a large retroperitoneal haematoma (white arrow) and aneurysm (black arrow) just posterior to the uncinate process of the pancreas. Figure 1B shows a large heterogeneous haematoma (white arrow head) within the right gluteus medius muscle.

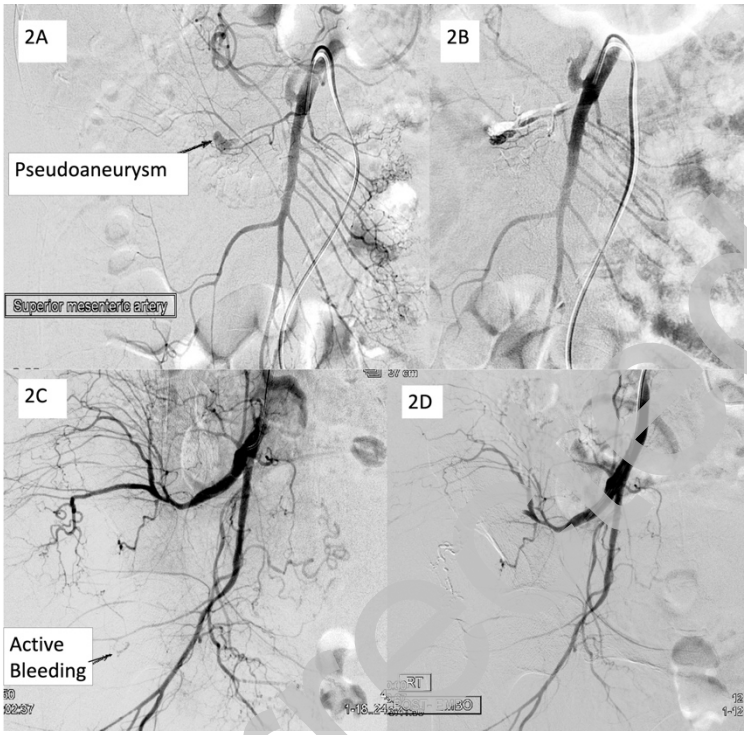

FIG. 2. A-D. Digital subtraction angiogram (DSA) of the superior mesentery artery shows a pseudoaneurysm arising from the right inferior pancreaticoduodenal artery (black arrow). (B) Post-embolization superior mesentery angiogram shows no contrast flow into the pseudoaneurysm. (C) DSA of the right common iliac artery shows active contrast blush (black arrow) seen from a branch of the right inferior gluteal artery. (D) Post-embolization angiogram shows no active bleeding.