

# Common variable immunodeficiency causing granulomatous disease of the abdominal aorta with aneurysm formation

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## Abstract

In 1995 a 42-year old patient with common variable immunodeficiency associated with granulomatous disease had an elective repair of a granulomatous abdominal aortic aneurysm. Five years later he presented with a ruptured false aneurysm of the left common iliac artery. Granulomata may cause aneurysm formation and weakens surgical anastomoses.

## Case history

In 1983 a thirty-year-old male was referred to the Immunology Department with recurrent episodes of multiple-site lymphadenopathy. Lymph node biopsy revealed granulomata containing Langhans type giant-cells. Autoantibody screen, Kveim tests, acid-fast-bacilli stains and cultures were all negative. Further immunology screening showed IgA deficiency and associated systemic antibody deficiency. The diagnosis of Common Variable Immunodeficiency (CVID) with granulomata / sarcoid like disease was made and the patient was started on intravenous human immunoglobulin.

Four years after treatment was commenced, the patient's renal function showed slight deterioration. An abdominal ultrasound scan (USS) revealed an enlarged spleen, cystic kidneys and a 9.5 cm infrarenal abdominal aortic aneurysm (AAA). A CT scan indicated that the right common iliac artery (CIA) was also aneurysmal, with a transverse diameter of 4.5 cm. The tissue planes around both aneurysms were poorly defined suggesting a significant inflammatory reaction.

The AAA was repaired with a rifampicin soaked aorto-bi-iliac dacron graft. Specimens from the aneurysm wall revealed non-caseating granulomata. The patient made an uneventful postoperative recovery. Three years later the patient had a haemodynamic collapse with abdominal pain. There was lower abdominal peritonitis; the left leg was mottled and cold with an absent femoral pulse. Laparotomy revealed a large haemoperitoneum, a ruptured left CIA false aneurysm and ischaemic changes to the left colon. Control of haemorrhage required oversewing of the internal and external iliac artery origins. Distal flow was restored with an 8 mm dacron graft from the left limb of the original graft to the femoral bifurcation. An extended left hemicolectomy and splenectomy were performed with transverse end-colostomy and stapled closure of the rectal stump. Specimens from the aneurysm, colon and spleen were sent for histopathology analysis with no significant findings reported. Cultures from the aneurysm wall were sterile.

The patient's recovery proved slow and difficult due his significant co-morbid conditions. On postoperative day 12, the patient began spiking a fever. Abdominal USS revealed fluid collections in the pelvis. Blood cultures grew *Pseudomonas aeruginosa*, coliforms and anaerobes. Laparotomy with drainage of intra-abdominal fluid collections was performed. There was no evidence of graft exposure and the retroperitoneum was intact. The rectal stump staple-line was found to have disrupted. Pelvic drains were placed and the patient was commenced on broad spectrum antibiotics. The patient was discharged six weeks after admission following a prolonged period in Intensive Care.

## Clinical Evidence

Common variable immunodeficiency is a primary immunodeficiency in which B lymphocytes produce few or no immunoglobulins. Of unknown aetiology, it affects both males and females with onset occurring at any age (1). Affected individuals suffer from recurrent respiratory and gastrointestinal infections and,

paradoxically, autoimmune diseases (2). Many individuals also have disorders of cell-mediated immunity. In severe disease, treatment is intravenous gammaglobulin (1).

### Unusual features

The occurrence of non-caseating granulomatous lesions in patients with CVID (GD-CVID) has occasionally been described (3,4). Although similar to Sarcoidosis, important clinical and immunological differences suggest that GD-CVID should be classified and treated as a separate clinical entity. The presence of vasculitides involving the abdominal aorta and subsequent aneurysm formation has occasionally been described in Sarcoidosis (5). To our knowledge, this is the first report of large vessel aneurysms complicating GD-CVID.

### Lesson

- Granulomatous formation may be associated with CVID;
- Affected individuals may be prone to aneurysm formation;
- Granulomatous tissue weakens the integrity of surgical anastomoses;
- Reoperation in affected individuals is difficult due to dense scar tissue formation;
- Early screening of GD-CVID patients for aneurysm formation may be warranted.

### References

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