CASE REPORT

Hughes syndrome and splenic infarction as an unusual cause of acute left sided abdominal pain

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Back Ground:
In 1983 Graham Hughes described a condition of antiphospholipid syndrome in which there is a danger of thrombosis. This condition can be readily detected by blood tests and once diagnosed the risk of further thrombosis can be markedly reduced by anticoagulation. This group of patients can be distinguished by a specific blood test, the detection of antiphospholipid antibody (Ref-1). We report a case of a young woman presenting with acute left sided abdominal pain. On investigation she was found to have splenic infarction. Haematological investigations were positive for anticardiolipin antibody. A diagnosis of Hughes syndrome complicated by splenic infarction was made and she was treated with anticoagulation. Patients with Hughes syndrome have hypercoaguuable state with a markedly increased risk of both arterial and venous thrombosis. Splenic infarction is a rare presentation.

Case Report:
44 year old woman was admitted under the acute surgical “take” with left sided abdominal pain radiating to her back. She was a dental hygienist who lived with her husband and her two children. She smoked 15 cigarettes a day and there was no previous history of venous thrombosis. She had history of border line thyrotoxicosis in early 1990s and underwent Tension free vaginal tape for stress incontinence in September 2003. She was on etonogestrel-releasing implant for contraception. She was locally very tender over the left side of the abdomen but rebound tenderness was absent. Rectal examination was unremarkable. Investigations showed haemoglobin of 13.2 g/dl, white cell count -19.9 10^9/L, platelets 214 10^9/L with neutrophilia. Amylase and renal function tests were normal. Liver
function tests were deranged with Gamma GT 244 u/l (twice normal). Abdominal Ultrasound scan suggested a possible splenic infarction which was confirmed by a CT scan of abdomen. Tests were carried out to investigate the possibility of a post thrombotic state. Hams test was negative, but anticardiolipin antibody was positive. IgM level was 52 (normal up to 10) and IgG was 18.8 (up to 10). She also had border line APC Sensitivity 1.9 (2 to 4.3).Kaolin time 49 sec (70-120) Ktnix 64 sec (70-120), thyroid function test revealed TSH 0.32 mu/L ,fT4 20.2 pmol / L (10-25) .Her symptoms settled with simple analgesia and she was discharged home with long term anticoagulation.

Discussion:
Antiphospholipid syndrome (APS) represents a hypercoaguable state with increased incidence of both arterial and venous thrombosis. The characteristic features include vascular thrombosis, pregnancy loss, presence of antibodies, neurological disease, thrombocytopenia and lividoreticularis. The antibodies are directed against phospholipids proteins and there is a linear association between the tendency to clot and the level of phospholipids antibodies (Ref-2). Most of the patients are young women and potential causative factors are contraceptive pills, smoking, and hormone treatment. Sex ratio is 9:1 female: male. Cerebrovascular symptoms are mainly migraines, transient ischaemic attacks to full blown strokes, memory loss, choreo-athetosis, myelopathy, transverse myelitis and epilepsy. Cardiovascular involvement includes risk of fatal and non-fatal heart attacks in young women as a result of inflammatory thrombotic processes. Some develop accelerated atheromatous disease and there is high prevalence of echocardiographic valve problems. Thrombotic complications can present with visceral infarctions. In kidneys micro thrombi can form and also there can be evidence of renal vein thrombosis. Case studies showed higher prevalence of renal artery stenosis (26 % ) in patients with APS who have difficult to control hypertension than in a hypertensive group and otherwise healthy potential renal donors(Ref-8).APS is also found to be a cause of Budd chiari syndrome .Addison’s Disease due to acute infarction of Adrenal glands has been reported. E.Coli infections / surgery can precipitate wide spread thrombosis in these patients. One large study from USA did CT scan on 215 patients with antiphospholipid syndrome and found splenic infarction in 6 patients (Ref-9). Current therapy to prevent recurrent thrombosis is controversial. Anticoagulant treatment is a better option than anti-aggregants alone .But there is a risk of bleeding with anticoagulant treatment, and the need for frequent monitoring of the INR (International normalized ratio)to measure the anticoagulant effect of warfarin concern patients and physicians. Acute thrombosis is treated by heparin infusion followed
by warfarinisation. Use of low molecular weight heparin reduces the risk of thrombocytopenia. Most data support long term oral anticoagulation. Prospective studies suggested that an INR between 2.0 – 2.8 might be adequate prophylaxis for venous thrombosis while retrospective studies suggest the need for a higher intensity anticoagulation to prevent recurrent arterial events. (Ref-4)

**Comment:** Unusual surgical presentation of a thrombotic abnormality as abdominal pain due to splenic infarction. The surgeon always has to be “on his toes”!

**REFERENCES:**
1. GRV Hughes: The antiphospholipid syndrome a historical review. Lupus (1998) 7, suppl 2, s 1-4
2. E C G Grant: The Hughes (Anti-phospholipids) syndrome. Journal of nutritional and environmental medicine; Jun 1998, 2; proquest nursing journals page 153
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