Case Report

Coronary Artery Anomaly with Congenital Factor V Deficiency

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Abstract

The case of a 60 years old woman with coronary artery anomaly and factor V deficiency is reported. She presented with right radial artery pseudoaneurysm after coronary angiography. Her coronary angiography revealed aberrant origin of left anterior descending artery from right coronary cusp. On preliminary investigations her prothrombin time and activated partial thromboplastin time were prolonged. Her plasma factor V level was found out to be 4%, a moderate deficiency of factor V.

Introduction

Isolated factor V deficiency is a rare inherited coagulopathy. Homozygous factor V deficiency is rare, occurring in approximately one per million population.[1] A higher occurrence of congenital cardiovascular malformations (CCM) in patients with heritable coagulopathies has been reported.[2-4] However, no association has been seen with isolated factor V deficiency. We present a case with a unique combination of factor V deficiency and congenital coronary artery anomaly. This coexistence has important management implications as cardiac disease often requires major surgery and associated coagulopathies are likely to cause major bleeding problems.

Case History

A 60 years old woman was admitted for coronary artery bypass grafting. She had limiting angina for last two months. She had also developed a swelling at right wrist at the site of access for coronary angiography, which was carried out about a month ago. There was past history of post partum haemorrhage after childbirths and once blood transfusion was required due to massive bleeding. She also had history of menorrhagia and echymosis. Her father had ischaemic heart disease but there was no history of such bleeding diathesis in other family members. Her medication was amlodipine, bisoprolol, isosorbide mononitrate, simvastatain, and aspirin. She was not on anticoagulants. There was no history of drug allergy.

Table: Clotting profile of the patient.

TEST	RESULT
PT	28 sec
PT control	12 sec
INR	2.24
Correction with normal plasma	16 sec
Correction with aged plasma	19 sec
Correction with adsorbed plasma	16 sec
APTT	67.2 sec
APTT control	30 sec
Correction with normal plasma	33 sec
Correction with aged plasma	51.6 sec
Correction with adsorbed plasma	34.1 sec
Factor V level	4% (50-150) normal

Clinical examination of the patient revealed a swelling (3 x 2 cm) at the right wrist which was bluish in color, pulsatile, compressible with an audible bruit. There was also a small spontaneous haematoma on left thumb and multiple bruises on abdomen, thighs and buttocks. Rest of the examination was normal

The relevant blood tests showed haemoglobin of 12.4 g/dl, platelet count of 220,000/mm^3 and normal bleeding time. Prothrombin time (PT) and activated partial thromboplastin time (APTT) were markedly raised and further specialized hematological work up confirmed moderate factor V deficiency (Table). Her abdominal ultrasonography and echocardiography were unremarkable. The liver and renal function tests were normal as well.

Her coronary angiography showed anomalous origin of left anterior descending artery (LAD) along with right coronary artery (RCA) from right coronary cusp (Figure 1), whereas left main stem (LMS) bifurcated into left circumflex and a rudimentary LAD (Figure 2). There was a tight stenosis in middle of RCA, and significant disease in anomalous LAD and in an obtuse marginal branch

The right radial artery pseudoaneurysm was repaired without any adverse event. As her angina was controlled by medication, there was an increased risk of bleeding and her cardiac surgery was not considered essential for prognostic reasons, she was managed conservatively with dual antiplatelet regime of aspirin and clopidogrel, and a high dose statin.

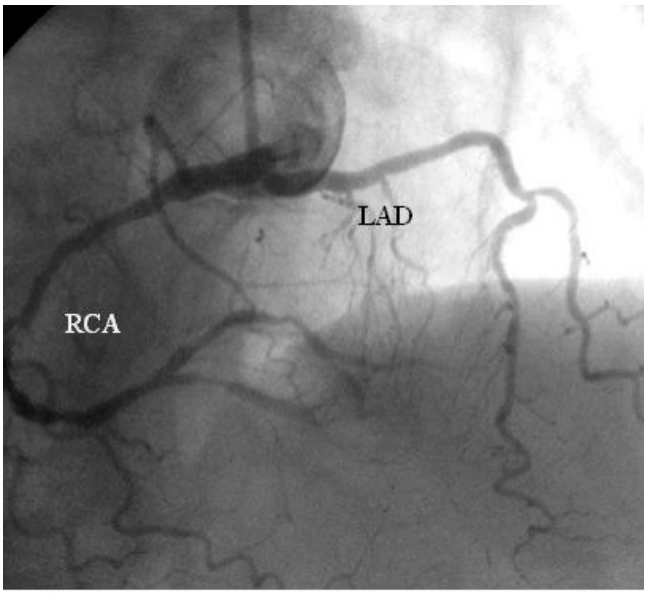

Figure 1: Posteroanterior (cranial) view showing, LAD (arrow) arising from RCC alongwith RCA.

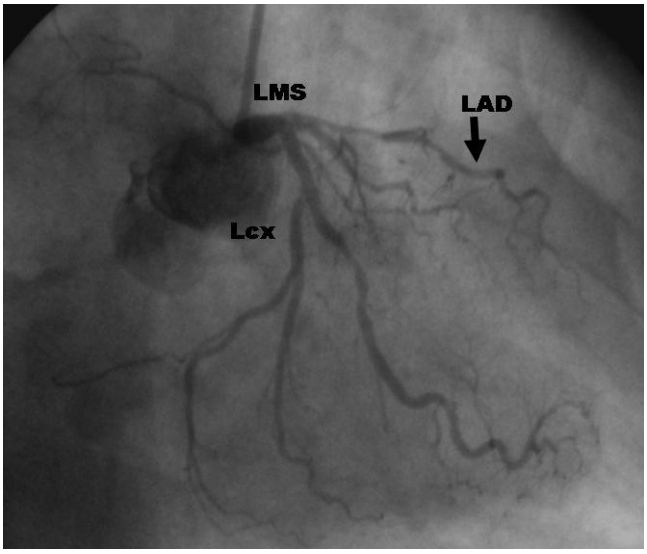

Figure 2: Posteroanterior (caudal) view showing left main stem dividing into left circumflex artery and a rudimentary LAD (arrow).

Discussion

Factor V deficiency is a rare coagulation disorder which is inherited in an autosomal recessive manner. It is also known as Owren disease or parahaemophilia. This defect was first described in Norway in 1947 by Paul Owren.[5] Because of the rarity of the disease so far no real associations have been identified. There are a few studies showing increased occurrence of CCM in babies born to the parents with heritable coagulopathies.[2,3] Similarly Jedele et al reported an increased frequency of congenital heart defects in patients of haemophilia.[4]

Embryologically the association is possible as the heart and blood arise from common angiogenic cells; the endothelial cells, which are the first components of primitive heart, and these cells also synthesize coagulation factors. But further studies are needed to find out the definite association of CCM with the coagulopathy. Generally there are reports of isolated factor V deficiency[6,7] or various surgical problems dealt in patients with factor V deficiency.[8-10] In this case we observed factor V deficiency along with congenital coronary anomaly that is LAD arising from RCC along with RCA.

Bleeding is a major risk in patients with factor V deficiency undergoing surgery. The concentrates of factor V are not commercially available, so fresh frozen plasma (FFP) is used to correct the deficiency. The loading dose of FFP is 15-20 ml/kg to be followed by 3-6 ml/kg daily. Subsequent dosages depend upon monitoring the factor V level by obtaining the peak and trough levels, with the aim having a factor V level of 25% before surgery. Postoperatively, FFP should be administered daily for 3-10 days, with careful observation of bleeding.[1]

In summary, we have described a rare case of aberrant origin of LAD with isolated factor V deficiency. To the best of our knowledge, there has been no similar reported case of coronary anomaly with heritable coagulopathy.

References

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