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CASE REPORT

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Acute mesenteric ischemia in an African American patient with heterozygous factor V Leiden deficiency

Hadiatou Barry [a], Ali Ghani[a] and David Gary Smith[a]

[a]Internal Medicine, Abington Hospital Jefferson Health, Abington, PA, USA

ABSTRACT

It is well documented that factor V Leiden mutation (FVL) is a common hypercoagulable risk factor in the Caucasian population. Patients with homozygous FVL mutation have an increased risk for venous thromboembolism. However, there have been few cases of heterozygous FVL mutation associated with arterial thrombosis described in the literature. Our case report presents an African American (AA) female with heterozygous FVL mutation who presented with acute arterial mesenteric ischemia.

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1. Introduction

Factor V Leiden is a mutation form of factor V which makes factor V insensitive to activated protein C [1]. This mutation makes patients hypercoagulable [2]. FVL is the most common inherited thrombophilia in the Caucasian population [1,2]. Patients tend to develop venous thromboembolism (VTE) commonly (DVT) and pulmonary embolism (PE) although, only a small percentage of these patient will have a VTE [2]. For patients who are carriers of more than one thrombophilic defect, the risk of thromboembolism is increased [1]. Patients with this mutation can suffer unprovoked, life-threatening DVTs, PEs, or VTE at an unusual site such as in the mesenteric or portal vein and tend to be on lifelong anticoagulation.

Occlusive arterial obstruction is due to an acute embolism or thrombosis and most commonly affects the superior mesenteric artery (SMA) [3,4]. Interestingly, arterial thromboembolism associated with factor V Leiden mutation has been controversial if not, rare. There are case reports that show patients with factor V Leiden mutation and arterial thromboembolism, however, there has not been any clear association found in the literature between FVL and arterial thrombosis [1,2]. We present a case report of a patient with heterozygous factor V Leiden deficiency and arterial thrombosis.

2. Case presentation

42-year old African American female with a past medical history of morbid obesity, heterozygous FVL mutation, DVT and PE, splenic infarction, descending aortic thrombi, insulin dependent diabetes and hypertension. Hypercoagulable workup was otherwise unremarkable except for FVL. Her family history is significant for a mother decreased at age 35 from PE and a father deceased from myocardial ischemia. Due to the patient's prior history of thrombosis and hypercoagulable state, she was on lifelong anticoagulation with warfarin.

Patient was admitted with a sudden on-set of acute left-upper-quadrant (LUQ) abdominal and epigastric pain on the morning of admission. Pain was crampy, severe, constant, and non-radiating associated with nausea and two episodes of bilious vomiting. Pain was worse with movement and associated shortness of breath. On examination, patient was tachypneic and tachycardic, in acute distress from pain with dry mucous membranes. Abdominal exam was notable for tenderness on the LUQ and epigastric region with guarding. Blood pressure was well maintained. Patient was found to have a subtherapeutic International Normalized Ratio (INR) 1.6, white count of 19.6 K/UL, elevated d-dimer of 2889 NG/ML DDU, and a lactic acid of 3.4 MEQ/L.

The patient had a prior history of allergy to contrast dye where she developed desquamation after exposure.

She required pre-medications with IV methylprednisolone and diphenhydramine. CT angiogram was performed which revealed an abrupt occlusion of the SMA (distal to its origin) with associated small bowel dilatation consistent with ischemia. Fortunately, this was without associated bowel perforation.

Patient was placed on Intravenous (IV) heparin, IV hydration, IV board-spectrum antibiotics and taking to the operation room (OR). Patient underwent exploratory laparotomy with superficial mesenteric artery

CONTACT Hadiatou Barry Hadiatou.barry@jefferson.edu Internal Medicine, Abington Hospital Jefferson Health, 1200 Old York Road, Abington, PA 19001, USA

thrombectomy and temporary abdominal negative pressure wound therapy (VAC) placement, complicated with hypotension. Consequently, the patient was placed on norepinephrine for pressor support. She underwent a second look and re-exploratory laparotomy on day 2. Over the course of 2 weeks, she underwent multiple surgeries including exploratory with resections and wash out. The patient received multiple pack red blood cell transfusions throughout the course of her complicated surgeries and had a total of 119 cm of small and large bowel resection. She required total parenteral nutrition (TPN). She had an interventional radiology embolization of bleeding in the abdominal wall. After a one-month hospitalization, the patient was discharged to inpatient rehabilitation in stable condition.

3. Discussion

Factor V Leiden mutation is strongly associated with VTE, prevalence can range from 2–7% in patients without a history of thrombosis to 20–50% in patients with venous thrombosis [5]. However, there have been some case reports in the literature of FVL and arterial thrombosis. FVL alone does not predispose to clot formation in the absence of other risk factors such as stasis and endovascular injury. A case report of a heterozygous FVL mutation in a smoker with elevated homocysteine levels and peripheral arterial disease concluded that FVL associated with other prothrombotic risk factors may predispose patients to arterial thrombosis [1]. Furthermore, FVL enhances the risk for thrombosis in those patients with other thrombophilic risk factors i.e. protein C and S deficiencies, hyperhomocysteinemia, and patients on oral contraception or estrogen replacement therapy [2]. There is a report of a 24-year old female with FVL mutation and acute artery thrombotic occlusion on the digital artery of the thumb and index finger; only additional risk factor was oral contraception [6].

Another study presents a case of a young patient with a history of heterozygous FVL who developed lower limb arterial, ascending aorta and aortic arch thrombosis and subsequent emboli to the eye [7]. There were four patients with heterozygous FVL mutation who developed visual field defects due to arterial thrombosis, two of these patients had no other coagulation abnormality other than FVL mutation. The study concluded that heterozygous FVL mutation might predispose some patients to arterial occlusion [8]. Therefore, it is essential to evaluate other factors that can predispose patients to arterial and venous thrombosis such as hyperhomocysteinemia, obesity, oral contraction, and peripheral arterial disease.

Clearly, there are multiple cases that have demonstrated that FVL and arterial thrombosis require further investigation to identify association or causation. Since there is no clear association that is well established in the literature between FVL and arterial thrombosis, many of the case reports noted here have other comorbidities or factors that may predispose the patient to arterial thrombosis. Our patient had a history of prior arterial thrombosis and multiple risk factors for atherosclerosis and vascular pathology (hypertension; obesity; poorly controlled and prolonged diabetes; and a family history of heart disease). FVL in this case may be an additional risk factor for the patient. She had no other hypercoagulabilities except for FVL. In these patients, it is important to mitigate the modifiable risk factors that increase risk of arterial thrombosis. Furthermore, every effort should be made to maintain adequate anticoagulation, advocating for Direct Oral Anticoagulant (DOAC) instead warfarin.

It is essential to carefully review patients' personal and family history, and any history of prior embolic events. Prior embolic events and personal or family history of thrombosis presents in many patients with acute mesenteric thrombosis [9]. This case report added to other prior case reports that suggest that FVL mutation may contribute to arterial thrombosis however, other risk factors may also play a role. Therefore, more research studies are needed in this area to establish a clear association.

4. Conclusion

This report outlines a few case reports in the literature that present patients with a history of FVL mutation and arterial thrombosis. Our case report adds to this growing literature and supports the need to establish an association and looking at other synergetic factors that lead to arterial thrombosis in this patient population.

Disclosure statement

No potential conflict of interest was reported by the authors.

ORCID

Hadiatou Barry http://orcid.org/0000-0003-2352-4061

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