Acute Superior Mesenteric Vein Thrombosis Associated with Factor V ‘Leiden’ Gene Mutation

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Abstract

Objectives: To study thrombophilia states in Indian patients with acute spontaneous superior mesenteric vein thrombosis (SMVT).

Methods: Two men with this condition, a 56 year old and a 31 year old presenting with acute SMVT, demonstrated on CT scan, were subjected to a thrombophilia screen consisting of Protein C, S, antithrombin levels, lupus anticoagulant, anticardiolipin antibodies, fibrinogen levels, factor VIII levels, factor V ‘Leiden’ gene mutation, and paroxysmal nocturnal hematuria screen.

Results: A thrombophilia screen showed factor V ‘Leiden’ gene mutation (heterozygous) in both cases. Additionally, the first patient had high fibrinogen levels and the second high factor VIII levels. Both patients are currently on long-term anticoagulation.

Conclusion: Factor V ‘Leiden’ gene mutation in association with other thrombophilic factors may predispose to spontaneous superior mesenteric vein thrombosis.

INTRODUCTION

Spontaneous acute superior mesenteric vein thrombosis is an unusual cause of acute abdominal pain.1 In 30-50% of these cases, investigations reveal an underlying thrombophilic state.2 The factor V ‘Leiden’ gene mutation is one such condition, which has been found to be of significance in venous thrombosis in the Western world2 as well as in hepatic venous thrombosis in the Indian population.3 We report two cases of superior mesenteric vein thrombosis detected to be heterozygous for the factor V Leiden gene mutation.

CASE REPORTS

Case 1

A 56-year old man presented with sudden-onset severe generalized abdominal pain. He had no previous medical complaints. He was a non-smoker. Examination revealed generalized abdominal tenderness and ileus. An abdominal CT scan showed thrombosis of the superior mesenteric vein (Fig. 1). He was managed conservatively and therapeutically anticoagulated, initially with heparin and then with warfarin. He improved and was discharged on a full diet with instructions to maintain anticoagulation therapy. A follow-up Doppler examination showed that the superior mesenteric vein had recanalised. However, he complained of persistent mild abdominal discomfort. Two months later, he suffered from another attack of severe abdominal pain. The CT scan on this occasion showed the superior mesenteric vein to be patent along its full length; however, there was stranding of mesenteric fat and some jejunal loops were

Fig. 1: CT scan showing thrombus in the superior mesenteric vein (arrow).

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Following amplification of the gene of interest, the factor V 'Leiden' mutation (G1691A) mutation. In addition, Case 1 had a high factor V 'Leiden' gene mutation. In patients with acute thrombosis the presence of this mutation in patients with acute thrombosis of the portal-mesenteric axis. It is interesting that both patients in the current series are Maharashtrians; a high prevalence of this mutation has been reported in the Parsi population.

Recent data from the West have suggested that venous thrombosis is a ‘multicausal’ and ‘multigenic’ disorder. Multiple factors, inherited as well as acquired, may play a role in the pathogenesis of venous thrombosis in patients with inherited thrombophilia. This is borne out in both our patients where, in addition to the factor V Leiden gene mutation, hyperfibrinogenemia existed in one and increased Factor VIII activity in another, both of which are independent thrombophilic factors. A combination of factors is likely to increase the risk of recurrent venous thrombosis.

Spontaneous acute superior mesenteric and portal vein thrombosis is a difficult clinical problem to diagnose and treat. Conservative treatment is effective in the absence of transmural gangrene or perforation. The duration of which anticoagulation needs to be administered in these cases is unclear. It has been observed that the thrombus may lyse spontaneously; anticoagulation may be discontinued in such cases. However, late complications, in the form of ischaemic strictures, rethrombosis and perforation may occur as seen in the first case, necessitating careful follow up. A schedule of 3-6 months of anticoagulation is recommended for those with a single attack of deep vein thrombosis. However, in the presence of thrombophilia states, there is a high possibility of recurrence, even at sites remote from the index attack, as seen in the second case, and long-term anticoagulation may be preferred in such patients, especially when more than one thrombophilic condition is detected.

REFERENCES


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**Book Review**

**Pulmonary Function Tests in Health and Disease**

Edited by: Prof. PS Shankar

This excellent monograph is a collective work of various authors from different parts of India. There are 23 chapters. The opening chapters provide basis of respiratory mechanics of breathing and pulmonary function equipment. Subsequently the tests for ventilation, airflow, lung volumes, airway resistance and compliance, distribution and small airways have been described in detail. There are chapters on classification of abnormal patterns of pulmonary functions bedside pulmonary function tests, preoperative assessment of pulmonary function, evaluation of PFT by radiology, regional lung functions, PFT during sleep, arterial blood gases and broncho, provocation tests. A small eye opener to all practising physicians with interest in pulmonary medicine.

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The "Silver Jubilee Conference Issue of Indian Society of Electrocardiology" is available on the Website. Feature of "ECG of the Fortnight" has also been instituted. You are requested to visit the website : www.iseindia.org

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Sd/-

SB Gupta

Hon. Secretary ISE