



VARICOSE VEINS WITH IVC THROMBOSIS: A RARE CLINICAL PRESENTATION

Surgery

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ABSTRACT

Developmental Inferior Vena Cava (IVC) hypoplasia is rare and associated varicose veins, is a rarer combination. Here we report a case of a 38-year-old gentleman presenting with signs and symptoms of lower limb varicose veins. The exceptional finding noted on examination was the prominent tortuosity of the superficial circumflex iliac vein, with reversal of flow. Hypercoagulability testing detected raised homocysteine levels suggesting a possible proclivity to deep vein thrombosis. Non-visualisation of common iliac veins and lower part of IVC on CT and MRI abdomen pointed towards congenital hypoplasia/aplasia of the veins. Long-term Warfarin therapy, along with stockings and life-style modification, were initiated. This case report brings forth a rare clinical entity and emphasises that surgical intervention may not always be appropriate for varicose veins, even when otherwise indicated. Anticoagulation therapy remains the mainstay of treatment of congenital anomaly of veins as well as DVT, irrespective of coagulation profile.

KEYWORDS

Congenital hypoplasia-Deep Vein Thrombosis- Inferior Vena Cava-Varicose Vein

Case Report:

A 38-year-old non-smoker, non-alcoholic, normotensive, non-diabetic, male, presented with the chief complaint of prominent varicosities in calf and medial side of thigh of the left leg. The prominences of the veins were painful, bled from multiple sites along the course of the vein, reportedly increased on standing and did not reduce completely even in the recumbent position. He had no history of dyspnoea or chest pain or any significant previous medical history or family history.

Clinical examination revealed a prominent great saphenous vein, moderate varicosity, with blackish discolouration and small ulcerations of overlying skin, at the sites of bleeding complained of (Fig 1a). An exceptional finding noted, was the excessively tortuous and prominent, superficial circumflex iliac vein (Fig 1b) with reversal of the direction of blood flow. Even in standing position, the direction of blood flow was from below upwards. Peripheral arterial pulses were normal and there was no apparent neurological deficit.

Routine hemogram, serum electrolytes, blood sugar, liver function tests, renal function tests and thyroid profile were unremarkable. BT and CT were within normal limits, at 2min 10secs and 5min 20secs, respectively. INR off Warfarin(>10days), was 0.97 while APTT was 28.5 secs (Control 33secs). Hypercoagulability testing panel detected only raised Homocysteine levels (Value: 40.27 μ mol/L, Normal range: 3.7–13.9 μ mol/L). Levels of other tested parameters were normal, namely, Anti-Cardiolipin antibodies IgM (2.954MPLU/ml, Considered Positive if >18 MPLU/ml), Protein C (79%, Normal range:70–140%), Protein S (74.9%, Normal range:74–147%) and Anti-Thrombin III levels (0.287gm/dl, Normal range: 0.19–0.31). Factor VIII activity assay revealed a value of 134% (Normal range: 50–150%). Factor V Leiden (FVL) mutation and Prothrombin gene G20210A mutation studies were unavailable, and would anyway, be too expensive to afford.

B-mode Duplex Ultrasound Scan did not reveal any thrombosis in the limb veins or distal, significant perforator reflux. The report of the CT abdomen was not explicit, but review suggests absence/hypoplasia of the segment of IVC below the renal veins (Fig 2). No significant anomaly could be detected in other organs on CT Abdomen. MRI

reported non-visualisation of common iliac veins and lower part of IVC (Fig 3 & 4) suggestive of DVT and/or Congenital Hypoplasia of the veins. The collateral venous drainage through the circumflex iliac vein was also well delineated (Fig 4). Routine surgery for limb varicosities was out of



Fig 1a Blackish discoloration of the skin overlying the varicose Great Saphenous Vein



Fig 1b Tortuous and prominent superficial circumflex iliac vein

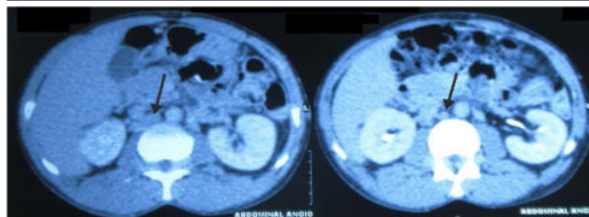


Fig 2 Loss of IVC Diameter Below Renal Hilum

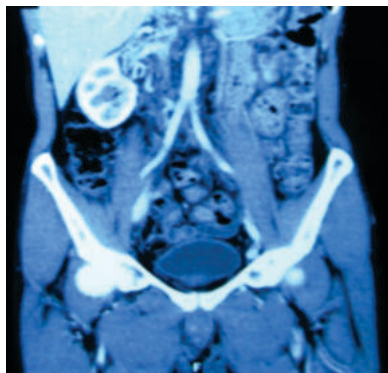


Fig 3 Absence of IVC Shadow Beside Aorta

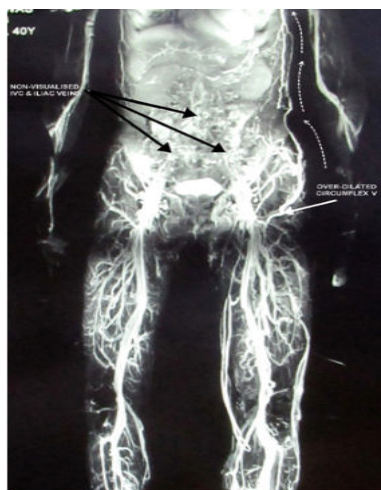


Fig 4 Non-visualisation of IVC and Iliac Veins and Circumflex Iliac Vein Reflux

Question, as the main route of venous drainage of the limb was through the superficial veins, into the circumflex iliac vein. The patient was started on Warfarin and was advised stockings and life-style modification, to avoid progression of limb symptoms.

DISCUSSION:

Inferior Vena Cava (IVC) hypoplasia is a rare phenomenon with an extremely low incidence (<1% in the general population, and 5% in young patients with DVT without other predisposing factors) [1]. The IVC develops from formation, regression and fusion among three pairs of veins namely, posterior cardinal, sub-cardinal and supra-cardinal veins. Numerous transformations during this complex embryological evolution may result in different anomalies in its final form (e.g., IVC hypoplasia, left-sided IVC, double IVC, and agenesis of infrarenal IVC).

Different studies [2],[3] reported a co-occurrence rate of approximately 5% between anomalous IVC and 'spontaneous unprovoked DVT' with 10 times increase in incidence of anomalies of IVC from 0.3%-0.5% in young healthy individuals to 5-6.7% in young adults with spontaneous DVT [2]. Stasis due to hindered blood return through anomalous IVC probably predisposes to venous thrombosis [4,5].

The possibility of co-existence of IVC anomalies and DVT prompted us to conduct laboratory investigations for detection of hypercoagulability, as far as available in our set-up. In our case, increased Homocysteine levels suggest co-existent proclivity to DVT.

However, in absence of consensus guidelines from academic societies, there is significant variability in the indications, nature and interpretations of the thrombophilic testing in a clinical practice [6]. In spite of significant addition to treatment expenses (in our case, Rs 28,000, even without genetic testing), their role in further management decisions is minimal [6]. Irrespective of results of thrombophilic screening, lifelong administration of warfarin is justified in this case due to the evidence in favour of IVC anomalies alone, leading to increased risk of DVT and its serious sequelae [7]. 1q

Though low back pain and abdominal pain have been reported as common presenting symptoms of co-existent IVC thrombosis and IVC anomaly, in our case, these symptoms were distinctly absent [8]. Although reported earlier [3] extensive literature search revealed that a presentation of varicose veins, among patients with IVC thrombosis in a setting of congenital anomaly, is rare.

We conclude by highlighting certain issues, that we feel make this case worthy of presentation. First, co-existent congenital IVC hypoplasia and DVT is a rare clinical condition, with varicose veins being an uncommon presentation of the same. Secondly, although varicose veins are encountered quite frequently in clinical practice, its aetio-pathogenesis demands careful attention. Otherwise, routine surgical intervention may lead to disastrous consequences. Thirdly, in the presence of IVC anomalies, coagulation-profile testing probably makes little difference in patient management, and hence, is, arguably, an unnecessary luxury in resource-poor patients. Lastly, anticoagulation therapy may be the mainstay of management of such patients, along with supportive measures necessary for amelioration of local symptoms.

Ethics approval: Not applicable

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