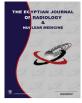
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Original Article

Complicated IVC anomalies: Are they more common than we thought? An experience of 100 MDCT venography examinations

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ABSTRACT

Objective: The aim of this study was to monitor the incidence of complicated inferior vena cava (IVC) anomalies and evaluate the role of Multidetector Computed Tomographic Venography (MDCTV) in diagnosis and assessment of associated venous collaterals, lower limb deep venous thrombosis (DVT) or varicose veins (VV).

Patients and methods: During two years duration 100 patients with clinical history and complains suggesting of DVT or VV were prospectively evaluated after performance of MDCTV examination. The images obtained were interpreted and reconstructed using dedicated software and work stations. Results were correlated with Color Doppler Ultrasound (CDUS) findings.

Results: Out of 100 cases, 9 cases (9%) were diagnosed to have complicated IVC anomalies while 91 cases (91%) had either well developed IVC or common anatomical variations. 6 cases (66.7%) had complicated IVC anomalies and 3 cases (33.3%) had associated complicated common iliac veins (CIV) anomalies. 8 cases (88.9%) had associated DVT and all cases (100%) had bilateral VV. 2 cases (22.2%) had associated varicocele and 1 case (11.1%) had associated KILT syndrome.

Conclusion: MDCT venography examination has a major role in diagnosis of complicated IVC anomalies and detection of associated venous collaterals, lower limb DVT or VV.

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1. Introduction

IVC anomalies are rare and under reported diagnosis that may predispose to DVT [1,2]. Formation of IVC occurs during 6th–8th weeks of embryogenesis. IVC is formed by continuous formation and regression of three paired embryonic veins: the posterior cardinal, subcardinal, and the supracardinal veins. IVC anomalies occur when there is an abnormal regression or persistence of any of these embryonic veins [3].

Several reports have reported absence of the whole IVC [4–6] or absence of infra renal part with preservation of supra renal part [7,8].

Absence of post hepatic IVC suggests that all three paired venous systems failed to develop correctly. Absence of infra renal IVC implies failure of development of posterior cardinal and supracardinal veins. Since it is difficult to identify a single embry-onic event that can cause one of these options, there is controversy

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as to whether these conditions are true embryonic anomalies or the result of perinatal IVC thrombosis [4,7,8].

IVC anomalies have an estimated prevalence of 0.5–0.6% in healthy individuals [9]. The most common congenital abnormalities of the IVC are duplication and retroaortic left renal vein. Absent IVC (segment or entire) has an incidence of 0.0005–1% in the general population [10]. Associated DVT may be present in more than 5% of the cases [11].

Despite being associated with DVT or chronic venous insufficiency, the correct diagnosis is often made late during the assessment of patients with suspected peripheral venous thrombosis [12].

While imaging is a reliable method to investigate IVC anomalies, most radiologists and clinicians may lack sufficient knowledge of these anomalies. This may lead to improper diagnosis in some cases [13].

The aim of this study was to monitor the incidence of complicated IVC anomalies and evaluate the role of MDCTV in diagnosis and assessment of associated venous collaterals, lower limb DVT or VV.

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2

2.1. Patients

cases.

2.2. Methods

2. Patients and methods

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2.3. Data analysis and interpretation

the end of injection.

of raw data including interactive viewing of multiplanar reconstructed images in axial source, 2D coronal maximum intensity projections (MIP), 3D MIP and volume rendering (VR) reconstructions.

The study was approved by the local ethical committee. 100 patients aged 22–40 years (with mean age of 31 years old) were encountered in this prospective study. The study was performed in a specialized medical center with an initial diagnosis of deep venous thrombosis (DVT) or varicose veins (VV) after performing Color Doppler study coming for confirmation. There were no set criteria for referral. Patients had clinical evaluation including medical and family history. Information regarding interventional procedures, surgeries and medical treatment was obtained from all

MDCTV examinations were carried out using an Aquilion One 320 scanner (Toshiba Medical Systems, New York, USA), with tube potential set at 140 kV, current at 300 mA, collimation at 3 mm and table movement at 4 mm/s. Two 20-gauge cannulas were placed one at each limb in a dorsal vein of the foot, with the legs adducted and extended. A tourniquet was tightly applied, one at each side above the level of the ankle. Total amount of 100 ml of 50% diluted (with normal saline) non ionic contrast material (iopromide, 300 mg iodine per ml, Ultravist 300; Schering AG, Berlin, Germany) was injected at each lower limb with an automatic injector at a flow rate of 3 ml/s. An operator initiated (>120 HU) Smartprep trigger was used to begin scanning from the level of the ankles up to the lower chest following short delay time of about 10–15 s after

All images were performed and evaluated by one staff radiologist with expertise in venographic imaging.

3. Results

100 consecutive patients were subjected to MDCTV, according to the study protocol aged on average 31 years (range, 22– 40 years). Patients were divided into two groups: Group 1 consists of 9 cases (9%) who were diagnosed to have complicated IVC anomalies and group 2 consists of 91 cases (91%) who had either well developed IVC or common anatomical variations as listed in Table 1.

Group 1:

Complicated IVC anomalies were diagnosed in 6 cases (66.7%) while 3 cases (33.3%) had associated complicated common iliac veins (CIV) anomalies (Figs. 1A, 3B and C).

Table 1			
The two groups	encountered	in tl	he study.

	Well developed IVC	Anatomical variations	Complicated IVC anomalies		No. (Total)	%
Group 1	-	-	9		9	9
			Without CIV anomalies 3	With CIV anomalies 6		
Group 2	85	6	-		91	91
All	85	6	9		100	100



Fig. 1. 3D VR images of the lower limbs, pelvis and abdomen for two patients having deep venous collateral pathways secondary to complicated IVC anomalies. (A) Multiple varicosities along both legs more pronounced and larger along the anterior and medial aspects of the right leg. Non opacified right superficial and common femoral veins. Attenuated right external and common illac veins. Multiple venous collaterals at the sacral and para vertebral plexuses. Enlarged azygos and hemi azygos veins. (B) Few bilateral varicosities. Non opacified right common femoral vein, left popliteal, superficial femoral and common femoral veins. Multiple venous collaterals along the para vertebral plexuses.

DVT was present in 8 cases (88.9%) and single case (11.1%) was free. 5 cases (55.6%) had bilateral DVT and 3 cases (44.4%) had unilateral affection. The femoro-popliteal segment was involved in all cases.

All cases (100%) had bilateral VV along both legs, more pronounced along the medial and anterior aspects following the courses of the great saphenous veins and their tributaries and could extend along the thighs.

Regarding the venous collateral pathways, 2 cases (22.2%) had the deep pathway (ascending lumbar veins and vertebral plexuses to anastomose with azygos and hemiazygos veins) (Fig. 1), 3 cases (33.4%) had the superficial pathway (inferior epigastric veins, internal mammary veins and abdominal wall veins) (Fig. 2) and 4 cases (44.4%) had combined deep and superficial pathways (Fig. 3).

Special software and workstations were used in reconstruction

Associated bilateral varicocele more on the left side was diagnosed in 2 cases (22.2%) (Figs. 2A and fig3C).

One case (11.1%) had associated Kidney and IVC abnormalities with Leg thromboses (KILT syndrome). The right kidney was accidentally discovered to be absent (with compensatory hypertrophy of the left kidney) and this patient suffered from extensive right lower limb DVT (Fig. 3A).

Group 2:

Well developed IVC was present in 85 cases representing 85% of all cases and 93.4% of group 2 population. Common anatomical variations were present in 6 cases representing 6% of all cases and 6.6% of group 2 population including left IVC, double IVC and retro aortic renal veins.

DVT was diagnosed in 77 cases (84.6%) and 14 cases (15.4%) were free. 14 cases (15.4%) had bilateral DVT and 63 cases (69.2%) had unilateral involvement. IVC and iliac veins were affected in 32 cases (35.1%), femoro-popliteal segment was affected in 29 cases (31.9%) and infrapopliteal segment was affected in 16 cases (15.4%) as listed in Table 2.

76 cases (83.5%) had bilateral VV along both legs and 15 cases had normal superficial veins. 10 cases (11%) had associated superficial thrombophlebitis of the great saphenous vein.

4. Discussion

Complete or partial absence of IVC is asymptomatic in most of the cases and is diagnosed incidentally by imaging. The most common clinical symptoms are related to venous insufficiency of lower extremities and/or idiopathic deep venous thrombosis [14].

Usually the most evident morphological changes seen with imaging techniques are related to the redistribution of the venous flow and the development of collateral pathways [15].

The ideal imaging modality to diagnose an IVC anomaly must have high diagnostic accuracy, and be safe and reproducible. The most reliable non invasive method for diagnosing IVC anomalies is CTV [16].

Halparin et al. [17] diagnosed 5 patients having underlying congenital abnormalities of IVC out of 25 patients who presented with spontaneous DVT of the proximal lower extremities in four years duration. This was surprising and the authors mentioned that their case series suggest that the presence of IVC abnormalities as a risk factor for DVT has been under-recognized. The current study included 9 patients suffering complicated IVC anomalies out of 100 patients in two years duration with higher incidence than expected.

In a study performed by Ruggeri et al. [18] four cases of complicated IVC anomalies were discovered over a five year period presenting with idiopathic DVT in those below 30 years of age. This was estimated to represent 5% of cases. Chee et al. [19] similarly stated that up to 5% of 20–40 year olds presenting with DVT had an IVC anomaly. The current study showed 8 cases with DVT associated with IVC anomaly in 22–40 years old patients representing 8% of patients. This agrees with both authors' opinion that IVC anomalies are more common than initially estimated and may be underdiagnosed particularly in patients under 40 years old. This matches results of another study performed by Sagban et al. [20] who said that younger males (mean age of 28 years) with atypical DVT are more often affected by absent IVC presenting with signs and symptoms of an acute DVT without previous evidence of risk factors.

Baeshko et al. [21] reported that incidence of bilateral DVT and chronic venous insufficiency is more than 50% of patients with IVC abnormalities. This matches the results of present study as 5 out of 9 cases had bilateral DVT representing 55.6% of cases and all cases

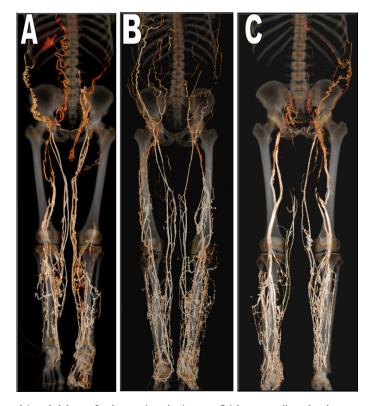


Fig. 2. 3D VR images of the lower limbs, pelvis and abdomen for three patients having superficial venous collateral pathways secondary to complicated IVC anomalies. (A) Multiple bilateral superficial varicosities. Non opacified right common femoral vein, left popliteal, superficial femoral and common femoral veins. Multiple venous collaterals along right lateral and anterior abdominal walls. Left sided varicocele. (B) Multiple bilateral superficial varicosities. Non opacified bilateral common femoral, superficial femoral and popliteal veins. Multiple venous collaterals along the anterior abdominal walls. (C) Multiple bilateral superficial varicosities. Non opacified superficial femoral vein, Multiple collateral veins along the left anterior abdominal wall and left intercostal veins.

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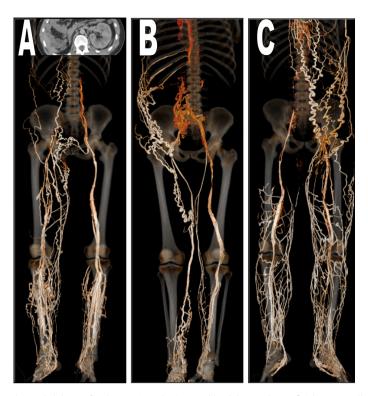


Fig. 3. 3D VR images of the lower limbs, pelvis and abdomen for three patients having combined deep and superficial venous collateral pathways secondary to complicated IVC anomalies. (A) Multiple bilateral superficial varicosities. Non opacified right superficial and common femoral veins. Multiple venous collaterals along the para vertebral plexus, inguinal regions, right anterior and lateral abdominal walls. Absent right kidney and compensatory hypertrophy of the left kidney (KILT syndrome). (B) Multiple bilateral superficial varicosities along medial aspects of the legs and right thigh. Non opacified right infrapopliteal, popliteal and deep femoral veins. Multiple venous collaterals at the lumbar veins, para vertebral plexus, inguinal regions, right anterior and lateral abdominal walls, inter costal veins. (C) Multiple bilateral superficial varicosities. Non opacified left common femoral and external iliac veins. Multiple venous collaterals along left para vertebral plexus, anterior left abdominal wall and intercostals veins. Right sided varicocele.

Table 2					
Deep venous thrombosis	incidence in	all c	ases,	n (%).

	Deep venous thrombosis			No DVT
	IVC and iliac veins	Femoro-popliteal	Infrapopliteal	
Group 1 (9 patients)	_	8 (88.9%)	_	1 (11.1%)
Group 2 (91 patients)	32 (35.1%)	29 (31.9%)	16 (17.6%)	14 (15.4%)
Total (100 patients)	32	37	16	15

(100%) had bilateral VV along both legs and sometimes extending to the thighs. IVC anomalies predispose patients to DVT; therefore, patients with a diagnosed IVC anomaly should be advised against excessive exercise or demanding physical activity [22]. DVT recurrence rate in this specialized population is presumably greater than in the general population with DVT because of flow stagnation and lifelong anticoagulation is typically recommended [23].

The common collateral pathways in disturbance in drainage of IVC are deep pathway (most common), intermediate pathway (rare) and superficial pathway (common) [24]. This slightly differs from the results of the current study as the superficial pathway was the most common in 33.4% of cases followed by deep pathway in 22.2% of cases while 44.4% had both pathways.

Our study included one patient with absent right kidney. Van Veen et al. [25] proposed the clinical constellation of kidney and IVC abnormalities with associated DVT to be named KILT syndrome.

MDCTV suffers certain technical limitations. In patients with severe edema of the lower extremities venous puncture may be difficult. Further problems which may take place are beamhardening artifacts due to the inflow of contrast and flow artifact due to the inflow of unopacified blood. In order to overcome these problems, diluted contrast material was used with scan delay time not more than 10 s. It is important to be distinguished from other causes of IVC occlusion like chronic thrombosis or compression/ invasion by retroperitoneal tumors. The correct radiological diagnosis would give the clinicians awareness of the condition, possible complications and alternatives for treatment.

In conclusion, complicated IVC anomalies should be suspected in young patients presenting with venous flow abnormalities or thrombosis with lack of classical risk factors. MDCTV is essential for the diagnosis, being able to not only diagnose the condition but also assess the related abnormalities as collateral venous pathways, lower limb DVT and VV.

Conflict of interest

The authors declared that there is no conflict of interest.

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