Agenesis of Inferior Vena cava in complicated by recurrent Deep venous thrombosis and Pulmonary embolism in a tertiary hospital , Sudan

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Abstract

A 47 year-old man who was successfully treated for PE . later on Discovered to have left sided venous engorgement in the abdomen and left lower limb . He was later discovered to have congenital absence of inferior vena cava and was prepared for investigations including venography and coagulation profile

Key Clinical Message : Congenital absence of Inferior Vena cava is a rare anomaly . As the case for other congenital anomalies , it should be suspected at all ages in any patient who presents with recurrent complications of venous stagnation such as PE and recurrent DVTs . Key Clinical findings are : recurrent DVT and/or PE , Stigmata of Venous engorgement as a result of development/enlargement of collateral veins .

Background **:** Congenital absence of Inferior Vena Cava (CAIVC) is a rare anomaly in which the lower-body venous return is impaired . There is usually engorgement of other veins especially Azygos and Hemiazygos systems.

Ethical Clearance : A full Informed consent was taken from the patient . Permission was taken from the patient for publication.

Case summary **: a 47 year-old man presented who was successful treated for PE . Later discovered to have left sided venous engorgement in the abdomen and left lower limb . He was later discovered to have congenital absence of inferior vena cava and was prepared for further investigations including venography and coagulation profile .**

Conclusion **: CAIVC could present later in life with complications as a result of Blood stagnation including DVT and PE .**

Introduction **: Agenesis of the inferior vena cava (IVC) is a rare abnormality that occurs in less than 1% of the population. The origin of the inferior vena cava is a complex process that occurs between the sixth and eighth week of gestation. When the process of formation of the vena cava incompletely occurs or it does not occur, we have a compensatory dilatation of the azygos system to help the venous drainage of the lower segment of the body.(1,2)**

Case presentation :

a 47-year-old male come to our clinic in 07/02/2019 complaining of,chest pain, shortness of breath and cough for the previous 5 days . The Chest pain was retrosternal , sudden in onset and was not radiated or referred

. In addition , the pain was severe , dull in character and there were no reported exacerbating factors . It was associated with shortness of breath at rest and cough . On examination , the patient was ill- looking , dyspneic with a respiratory rate of 30 . He was pale , and axillary temperature was 38.1 . Blood pressure was 110/73 and the Mean Arterial Blood Pressure (MAP) was 82 . Finally , SpO2 was 90 % on room air improving to 94% with high-flow oxygen via mask .

Regarding cardiovascular examination , there was pulsations in the neck which was found to be an arterial pulsation . JVP was not raised . Pulse was 139 , sinus rhythm , good volume , normal character and there was no radio-radial or radio-femoral delay . Precordium examination was normal . Chest examination was also normal apart from some transmitted sounds and possible fine crackles over lower zones . Abdominal examination revealed tortuous , snake-like protrusions extending from the epigastrium down about 2-3 cm from the midline in the left side radiating away from the umbilicus . In addition , there are few scattered swellings in the left lumbar and the left iliac regions . (Figure 1) . These swellings were found to be engorged superficial veins . Otherwise , the rest of abdominal examination was unremarkable .

On Examination of the lower limbs , the left lower limb was swollen up to the knee and it was darker in color as compared to the other limb . Also , there were multiple crustations and scratch marks more prominent on the medial ankle extending medially just below the knee but there were no ulcers . The limb was pitting with normal temperature . Later on , a diagnosis of DVT was confirmed using Doppler U/S scan . The right lower limb was completely normal .

After that , a set of Investigations were requested . CTPA confirmed a diagnosis of pulmonary embolism (PE) . CXR was normal . CT chest revealed dilated thrombosed azygos veins , subpleural honeycombing , early traction bronchiectasis , reticular interstitial thickening and lung architectural distortion . (Figure 2) . CT angiography for the lower limbs was completely normal .The patient has a previous venography that was done many years ago but unfortunately , it was not available at the moment . It showed complete **absence of inferior vena cava and dilated azygous and hemi-zygous venous systems more in the right side** .This finding was also noted in the abdominal CT (Figure 3)

BNP was high a level of 700 pg/mL .He received fractionated heparin 60 mg and warfarin 5 mg was thereafter continued in addition to analegesia . The patient's condition was improved , SOB and chest pain disappeared and repeated BNP was normal at a level of 10 pg/ml . The patient has had several episodes like this before and he was admitted at least once to the hospital for treatment of Pulmonary embolism . The diagnosis is Congenital absence of Inferior vena cava complicated by recurrent lower limb DVT and PE . Further investigations were requested to rule out other causes of VTE including Coagulation factors . Unfortunately , we could not be able to view the results of these tests because the patient did not bring them back later .

Author Contribution : Mohammed I Alfaki and Asma Abdallah : Were involved in selection of the case . In addition , they worked together in writing and editing the manuscript . Samoal Dafallah : He supervised the case presentation and management . He guided the process of selection and involved in final approval of the manuscript . All authors were involved in the editing and final approval of the manuscript.

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