

Thrombosis of left Internal Jugular Vein in Nephrotic Syndrome in a child with congenital absence of right Internal Jugular Vein: An extremely rare presentation.

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Abstract

There is a known association of nephrotic syndrome with hypercoagulable states and increased risk of thromboembolic complications. Internal jugular vein thrombosis is a rare complication of nephrotic syndrome with only a few cases described in the literature. Moreover congenital absence of right internal jugular vein itself is a very rare vascular anomaly of major venous channels. Here we report the case of a male child, 8 years old, a known case of steroid responsive nephrotic syndrome, presenting with extensive swelling of face, neck and abdomen. On cervical ultrasound completely occluding thrombosis of left internal jugular vein extending to left brachiocephalic vein was detected. On the right side, the absence of internal jugular vein was seen. A high index of suspicion and radiological investigations are needed for prompt diagnosis of this condition and also it should be kept in mind that any kind of anomalies can be seen during vascular interventions which can lead to various severe complications.

Keywords: thrombosis, nephrotic syndrome, internal jugular vein, agenesis

Introduction

Nephrotic syndrome is a loss of plasma proteins in urine and is defined in literature by a urinary protein level exceeding 3.5 g/1.73 m²/day and characterized by edema, hypoalbuminemia, hyperlipidemia and infectious and thromboembolic complications.¹ Nephrotic syndrome is well known to be associated with a hypercoagulable state due to many factors like

increase in the levels of fibrinogen and coagulation factors V and VIII in plasma, loss of antithrombin III in urine, thrombocytosis and increased platelet activation and aggregation. Thromboembolic disease is an important complication in patients with nephrotic syndrome and can involve both arterial and venous systems.² The incidence of thromboembolic

complications is comparatively less common in children than adults. The most frequently affected vessels include the renal veins, deep veins of the lower limbs, the inferior vena cava and the femoral & iliac artery.³ Vascular anomalies of major venous vessels are rarely seen. Moreover, congenital absence of internal jugular vein is extremely uncommon.⁴ A high index of suspicion is required to detect these abnormalities and imaging can be very helpful in diagnosis and early management of these patients.

Case Report

An 8-year-old male child was admitted with relapse of nephrotic syndrome who was referred to radiodiagnosis department for neck and abdomen ultrasound due to excessive edema in the abdomen, face and neck region with a focal painful lump on left side of the neck. He presented to the pediatric department with gradually increasing swelling of the whole body starting from the abdomen and now involving face, neck and limbs. He was diagnosed to have steroid-responsive nephrotic syndrome at the age of 3 years. He was an infrequent relapser.

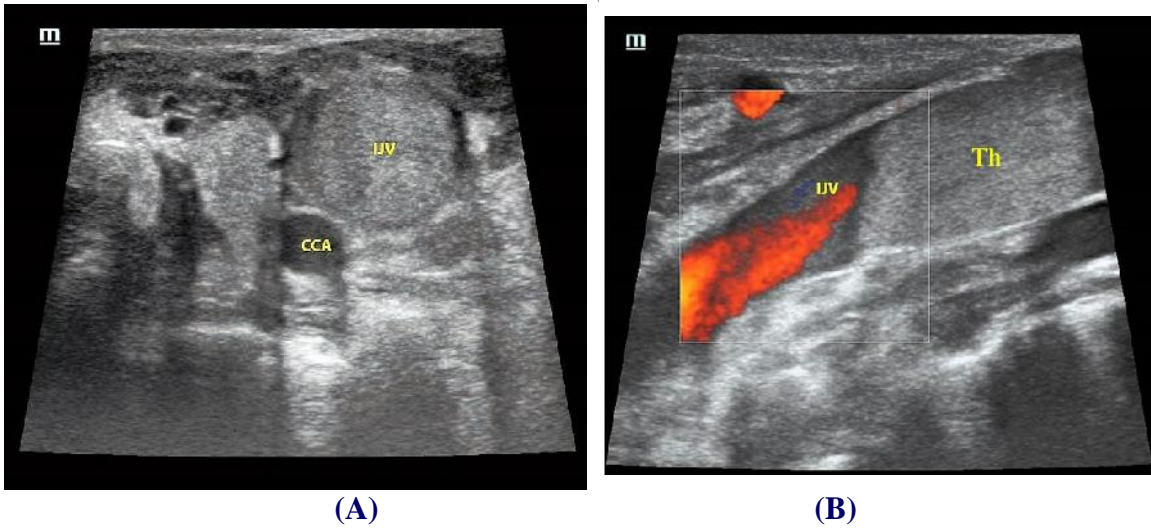
On examination, the child was irritable. Neurological examination did not reveal any signs of meningeal irritation, focal neurological deficits or papilledema. Height: 124 cm, Weight: 24.6 kg. Extensive edema involving abdomen, neck, face and limbs noted. Hb- 8.1g/dl, Urea - 42.7mg/dl, S.creatinine- 0.39mg/dl, ESR- 40, urine protein- 3+.



Figure 1: An 8-year-old child, k/c/o steroid-dependent nephrotic syndrome, presented with facial and neck swelling with a focal lump on the left side of neck and abdominal distension.

Ultrasound abdomen study using curvilinear transducer (Mindray ultrasound machine) showed massive ascites, both enlarged and echogenic kidneys with normal renal vessels as well as inferior vena cava. Doppler study showed no evidence of thrombus in them.

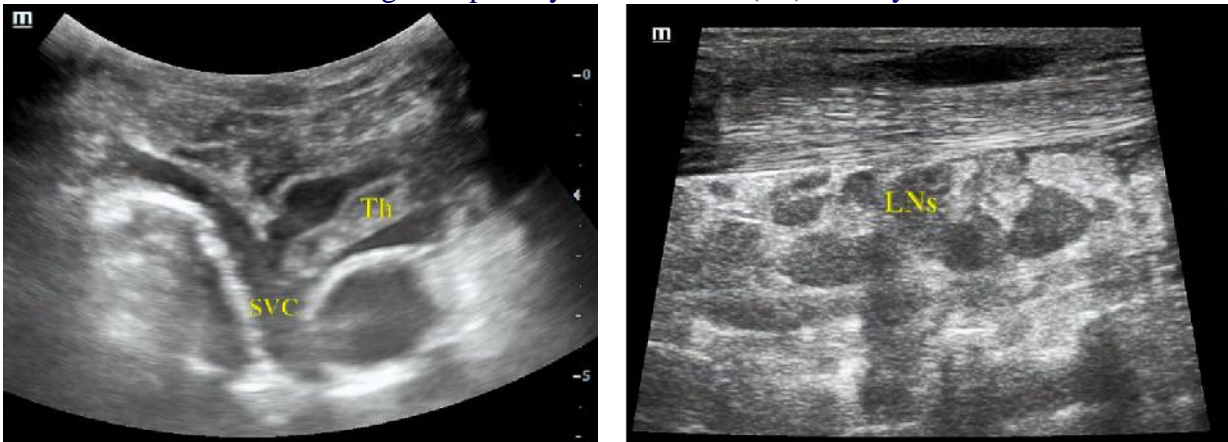
Ultrasound and Doppler study of the neck with linear high-frequency transducer revealed extensive subcutaneous edema with oedematous superficial parotid glands on both sides. Left internal jugular vein lumen was dilated, non-compressible, containing homogeneously echogenic completely occluding thrombus, which was also seen extending to left brachiocephalic vein up to the origin of superior vena cava. Thrombus also involved some proximal part of the left subclavian vein. Left internal jugular vein showed thickened wall, absent venous pulsations, the absence of normal Valsalva maneuver effects with left cervical chain lymphadenopathy along it. The flow was seen in the proximal lumen, which was being disrupted by the thrombus. Left common carotid artery was normal in caliber and flow. On Right side, a single pulsatile vessel was seen, which was not compressible suggestive of the right common carotid artery and was normal in course, caliber and characteristic pulsatile blood flow and spectral pattern. However, no evidence of right internal jugular vein was seen. Care was taken to apply minimal pressure on the probe to prevent the collapse of the IJV. No history of any neck surgery or vascular intervention was present. This was an incidental finding and suggests the congenital absence of the right internal jugular vein.



(A)

(B)

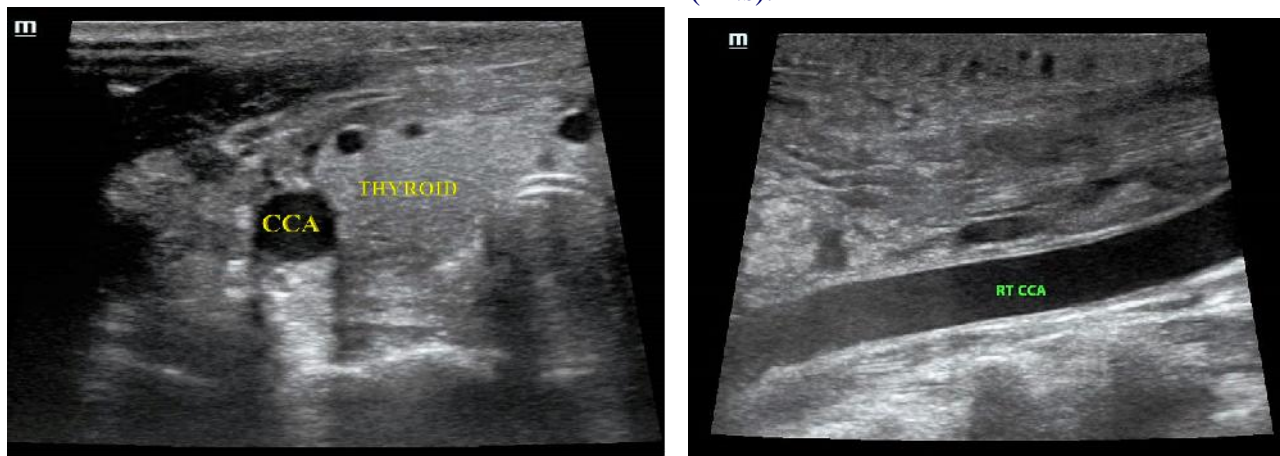
Figure 2: Ultrasound and study on the left side of the neck revealed, on the Axial scan (A), dilated internal jugular vein (IJV) containing completely occluding thrombus in it. Left common carotid artery (CCA) is normally visualized. On Longitudinal scan (B), flow can be seen in the normal proximal part of IJV, which is being disrupted by the thrombus (Th) distally.



(A)

(B)

Figure 3: (A) Thrombus can be seen extending distally into the left brachiocephalic vein, partially occluding it, and reaching up to the origin of Superior vena cava (SVC). (B) Enlarged chain of jugular lymph nodes (LNs).



(A)

(B)

Figure 4: Ultrasound study of right side of neck revealed: On Axial (A) and Longitudinal scans (B), only single vessel in the carotid sheath adjacent to the right lobe of the thyroid gland, which was noncompressible, pulsatile s/o right common carotid artery. No evidence of right Internal jugular vein is seen.

Discussion

The overall incidence of thromboembolism in nephrotic syndrome is about 3% in children and about 25% in adults.⁵ Maintenance of hemostasis involves a number of processes including platelet activation and aggregation, activation of the clotting cascade, termination of clotting cascade through a variety of inhibitors and dissolution of the clot by plasmin.⁶ Thrombosis in nephrotic syndrome can occur due to many factors like loss of proteins involved in the inhibition of systemic hemostasis or increased synthesis of factors promoting thrombosis or by activation of the hemostatic system by the glomerular disease. Other factors predisposing to thrombosis are intravascular volume depletion, exposure to steroids, increased blood viscosity associated with hemoconcentration due to use of diuretics.^{1,6} From various studies, it is found that the median time to thromboembolic events is about 71 days after the diagnosis of nephrotic syndrome.⁶ The most frequently affected vessels include the renal vein, the pulmonary artery, the deep veins of the lower limbs, the inferior vena cava, the femoral and iliac arteries.⁷ Our patient had left internal jugular vein thrombosis which is a rare site to be involved in this process. Lilova et al retrospectively studied 447 children with nephrotic syndrome for thromboembolic complications. The most commonly affected vessels were deep leg veins, followed by inferior vena cava. Other rarely involved locations observed were superior vena cava, mesenteric artery, IVC, and hepatic veins.⁸ Vyas et al reported a case of IJV thrombosis in a patient with nephrotic syndrome.⁹ Onishi et al reported a case of deep venous thrombosis in the upper extremities in a patient with nephrotic syndrome.¹⁰ Torres et al reported a case of venous sinus thrombosis in a patient with nephrotic syndrome.¹¹ Many other causes of IJV thrombosis include complication of surgical procedures, the presence of an indwelling venous catheter, tumor invasion, hypercoagulability, caudal extension of sigmoid sinus thrombosis, compression from adjacent tumor or nodes, a complication of adjacent infectious process, direct venous injection and after deep vein thrombosis of upper limbs.¹² IJV is a rare site to be involved in this process.

Moreover, IJV thrombosis has subtle and nonspecific clinical findings, So high clinical suspicion is required for the early diagnosis of this potentially life-threatening condition in the patients with nephrotic syndrome especially children. In the past, contrast venography was considered the standard diagnostic test but has now been replaced by ultrasonography which is a noninvasive and easily available modality.

Vascular anomalies result from embryological developmental deformities. Although vascular malformations exist at birth but they may not be detected until adolescence or adulthood.¹³ Venous malformations are mostly asymptomatic and are commonly located at head and neck region.⁴ IJV runs downwards through the neck along the carotid artery within the carotid sheath and behind the sternal end of the clavicle. It then unites within the subclavian vein to form the brachiocephalic vein, which enters the thorax to join superior vena cava which then drain into right atrium. In our patient, the possible absence of the vein can be explained by agenesis of IJV, which represents a form of truncular venous malformation due to a developmental arrest during the later stages of embryonic development, which could eventually lead to either aplasia or hypoplasia of the vein.¹⁴ Denys and Uretsky studied 200 patients undergoing IJV cannulation under US guidance and found that in 2.5% of the patients, the IJV was not visualized.¹⁵ Miller reported a case of absence of the right-sided IJV in a 12-year-old boy during US evaluation prior to attempted cannulation.¹⁶ We could not find any case in the literature with the combined presentation of congenital absence of IJV on one side and completely thrombosed IJV on another side, as in our case. The information of anatomical variations of IJV is clinically essential for venous applications especially in developing countries like India where central venous cannulation using the landmark technique is practiced commonly as an ultrasound machine may not be available in all centres. This case enforces the use of ultrasound-guided vascular access to prevent complications, which can arise due to the absence of a vein or other venous anomalies.

Conclusion

Thromboembolic disease is a life threatening complication observed in patients with nephrotic syndrome. Pulmonary embolism, renal vein thrombosis and deep vein thrombosis are the most common venous thromboembolic diseases seen in patients with nephrotic syndrome. Thrombosis of internal jugular is a rare complication of nephrotic syndrome and can present as painful neck swelling. A high index of suspicion is required to rule out this in patients with nephrotic syndrome. Doppler ultrasonography is the preferred method for the screening of deep vein thrombosis and should always be preferred to rule out thrombosis. Moreover, congenital absence of internal jugular vein itself is very rare. Practitioners are advised for prior USG confirmation for any vascular anomaly or variants before any vascular intervention without any complication and prolongation.

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