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SUPERIOR VENA CAVA SYNDROME AS A RESULT OF THROMBOSIS IN A CHILD WITH NEPHROTIC SYNDROME*

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SUMMARY: Çalışkan S, Sever L, Sarıoğlu A, Sarıoğlu T, Kasapçopur Ö, Arısoy N. (Department of Pediatrics, İstanbul University Cerrahpaşa Faculty of Medicine and Institute of Cardiology, İstanbul, Turkey). Superior vena cava syndrome as a result of thrombosis in a child with nephrotic syndrome. Turk J Pediatr 1997; 39: 561-564.

A three-year-old nephrotic girl is presented with vena cava superior syndrome. Angiography showed obliteration of the distal ends of both axillary veins and echocardiographic examination revealed a mobile mass in the right atrium. A thrombus originating from the vena cava superior with a stalk extending to the right atrium was surgically removed. To our knowledge, vena cava superior thrombosis in nephrotic syndrome has not been reported previously. *Key words: nephrotic syndrome, vena cava superior, thrombosis, thromboembolic complication.*

Increased tendency to hypercoagulability in nephrotic syndrome leads to thromboembolism, one of the syndrome's most serious complications. To date various localizations of venous and arterial thromboembolism have been reported. We present a nephrotic child with thromboembolism of the vena cava superior, a localization not previously reported, which led to vena cava superior syndrome.

Case Report

A three-year-old girl with nephrotic syndrome was admitted to our institution in September 1991. She was put on prednisolone (2 mg/kg/day, in divided doses) but there was no improvement after two months of treatment. The renal biopsy specimen showed segmental capillary wall collapse with sclerosis and adhesion to Bowman's capsule in two of nine glomeruli, which was consistent with focal segmental glomerulosclerosis. She was then treated with cyclosporine and low dose prednisolone for eight months, again with no response.

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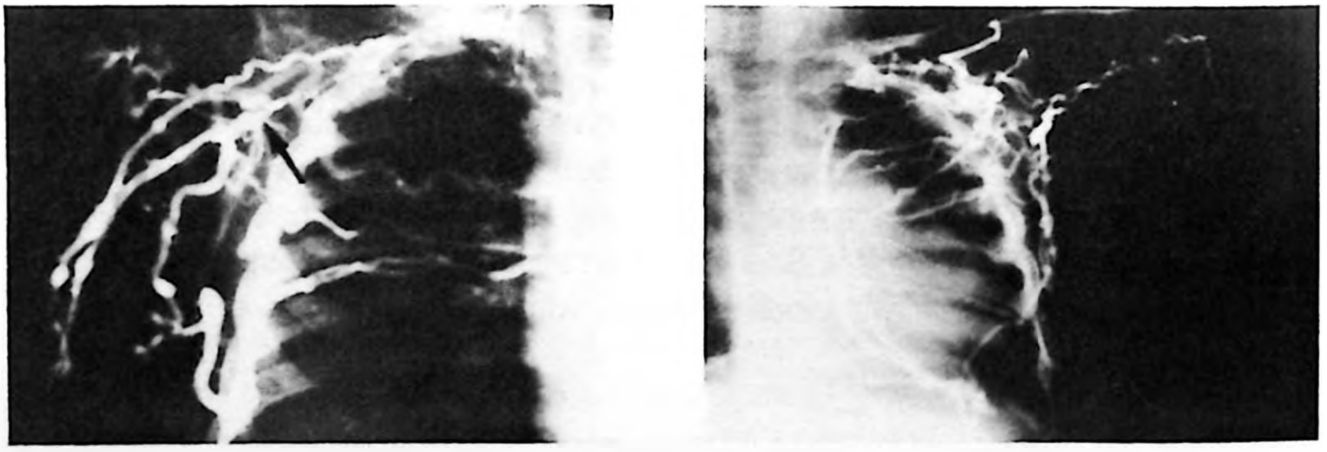
Having received prednisolone (5 mg/day) and cyclosporine (5 mg/day), the patient was readmitted in September 1992 with dyspnea and marked edema around the neck and shoulders. She did not receive diuretics and had no history of diarrhea. Collateral circulation was observed in the upper chest. Bilateral periorbital puffiness and pedal edema were also present. Blood pressure was 105/80 mm Hg, heart rate 120/min and respiration rate 34/min. Chest was dull to percussion on the left side with diminished breath sounds over the lower half of the same side; no rales were detected. Cardiac sounds and pulses were normal. A three centimeter hepatomegaly and minimal ascites were noted. Other clinical findings were within normal limits. Laboratory findings were as follows: serum protein 3.3 g/dl, albumin 1.7 g/dl, total lipid 2250 mg/dl, cholesterol 691 mg/dl, BUN 9 mg/dl, creatinine 0.45 mg/dl, sodium 143 mmol/L, potassium 3.6 mmol/L, Hb 11.5 g/dl, Hct 35%, WBC 12.000/mm^3, ESR 117 mm/h, GOT 26 IU/L, GPT 28 IU/L, PT 17" (53%), PTT 41". The urinary protein concentration was 0.8 g/dl (urinary protein/creatinine ratio 3.1). Chest x-ray revealed bilateral pleural effusion. On the second day of admission, the patient's right leg became swollen. It was painful and warm. Anti-aggregant therapy (salicylic acid 80 mg/day) was started. Echocardiographic examination revealed pericardial effusion but no other abnormality; however, the vena cava superior could not be detected. Thoracentesis yielded chylous fluid. Angiographic examination revealed extensive collateral circulation. The vena cava superior as well as both brachiocephalic and subclavian veins were not visible (Figs. 1a, 1b). A second echocardiographic examination, made 15 days after the



Figs. 1a, 1b: Angiographic frames showing obliteration of right axillary vein (arrow) and extensive collateral circulation. Note the vena cava superior, both brachiocephalic and subclavian veins, and left axillary vein were not visible.

first one, revealed a mobile mass (2.1 x 3.5 cm) in the right atrium. It was partly penetrating the right ventricle during diastole (Fig. 2). In the following days, a thrombus originating from the vena cava superior with a stalk extending to the right atrium was surgically removed. A thrombectomy as also performed on the

vena femoralis. Following initial anticoagulation with heparin (100 IU/kg/day) for three weeks, the patient was treated with Coumadin (1.25 mg/day). On the 17th post-operative day, fever and chest pain appeared; the physical findings were consistent with mediastinitis. Culture from the wound revealed plasma coagulase (-) staphylococcus. A surgical review was performed and the patient was treated with appropriate antibiotics. One month after thrombectomy, a control echocardiographic examination revealed no abnormality. She received a course of pulse methylprednisolone (30 mg/kg) weekly for three months, followed by alternate day prednisolone therapy (2 mg/kg/day) for two years. Proteinuria had disappeared completely by March 1994 and anti-coagulant and anti-aggregant therapies were discontinued.

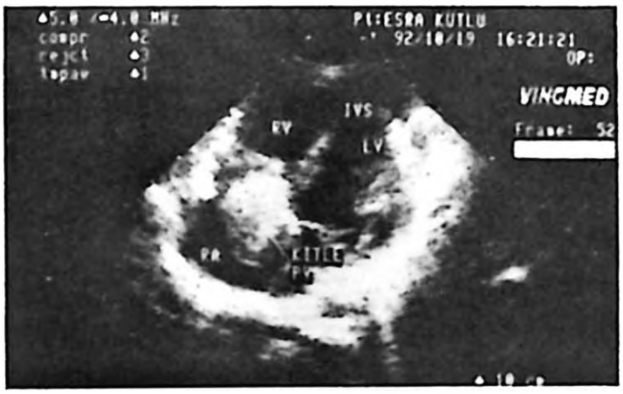


Fig. 2: Echocardiogram denoting the large thrombus in the right atrium partly penetrating the right ventricle.

At the time of her last visit in April 1995, the patient was doing well, and was active and healthy in appearance. Her physical examination was normal. Urinalysis and echocardiographic examination revealed no abnormality.

Discussion

Various factors, such as hypoalbuminemia; hemoconcentration; increased platelet aggregation; hepatic over-synthesis of fibrinogen, cofactor and lipoproteins; urinary loss of clotting inhibitors (especially anti-thrombin III); functional deficiency of protein C and protein S; and steroid and diuretic therapy, are responsible for hypercoagulability in nephrotic syndrome. It is assumed that this hypercoagulable state causes thromboembolic complications[1]. In spite of the low incidence of thromboembolic conditions in childhood[2], prospective studies showed a significant number of asymptomatic cases. This could contribute to an underestimation of the true incidence[3].

Deep vein thrombosis of the extremities and pulmonary embolism are most commonly encountered. A significant number of renal vein thromboses, with or without vena cava inferior thrombosis, have been reported[1]. Subclavian, axillary,

external jugular, splenoportal, mesenteric, hepatic and cerebral sinus veins and intracardiac spaces are uncommon sites for venous thromboembolism[4]. Arterial thrombosis is much less common than venous thrombosis and is observed mainly in children. Thrombosis of mesenteric, axillary, femoral, ophthalmic, carotid, cerebral, renal, pulmonary and coronary arteries has been reported[4]. To our knowledge, vena cava superior thrombosis in nephrotic syndrome has not been reported previously, but it has been encountered after interventions (venous catheterization, implantation of pacemaker and ventriculoatrial shunting), and in the course of systemic diseases (polycythemia, anti-phospholipid syndrome, Behçet's syndrome). It also occurs as the result of external compression (Hodgkin's and non Hodgkin's lymphomas)[5].

Another interesting point in this case is presentation of the thrombus as a right atrial mass in the second echocardiographic examination. The delay in diagnosis, due in part to technological inadequacy, may have caused the thrombus to grow and extend into the right atrium. Babcock et al.[6] reported the superiority of Doppler echocardiography of the superior vena cava, with its high frequency transducer for the diagnosis of thromboembolism, in which case an angiographic examination may be unnecessary.

Today, thromboembolic complications are successfully treated with thrombolytic agents like streptokinase and urokinase. In view of the stalkiness of the thrombus and the serious risks of pulmonary embolism, open-heart surgery was the preferred method of treatment for our patient.

In conclusion, vena cava superior thrombosis is a life-threathening complication that cannot be overlooked in nephrotic syndrome.

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