REMOVAL OF RIGHT ATRIAL THROMBUS PROPAGATED FROM SUPERIOR VENA CAVA IN A CHILD WITH NEPHROTIC SYNDROME

Thromboembolic episodes occur frequently in patients with nephrotic syndrome. We report the first case who developed right atrial thrombus propagated from superior vena cava, and leading superior caval syndrome and massive edema. Removal of the organised thrombus was performed successfully under total circulatory arrest. She had also right femoral thrombectomy at the same operation. Postoperative mediastinitis, due to hypoalbuminemia, corticosteroid therapy and immune disturbances was treated successfully. She is still in good condition with remission of her renal disease, and without recurrent thrombosis 30 months after the operation.

Key word:

Various coagulopathies resulting in hypercoagulability occurs in the patients with renal disease. Thromboembolic events occur with a frequency of 3-5 % in children with nephrotic syndrome (NS) 1. Thromboembolism takes place more often in membranous glomerulonephritis type of NS and less in minimal change NS 2. Numerous abnormalities in all phases of coagulation have been described in NS 2.

Patients with NS are under risk of renal vein thrombosis. Many thromboembolic episodes relating pulmonary, femoral, coronary and mesenteric arteries as well as subclavian, axillary veins, inferior vena cava and superior vena cava (SVC) were documented 2,6. We could not detect another case who developed right atrial thrombus propagated from superior vena cava in NS in our literature study. Anticoagulants, thrombolytic therapy or surgical approach can be more appropriate depending on the localisation of thrombus and patients clinical condition.

CASE REPORT

A 4-year-old girl who has been followed up for 18 months with mesangial proliferative glomerulonephritis developed progressive
massive edema, superior caval syndrome, dyspnea and oliguria. She has been on cyclosporin and steroid therapy for 6 months (Fig. 1). She did not receive platelet inhibitor or anticoagulant therapy previously. Chylous fluid due to increased pressure in Ductus Thoracicus as a result of superior caval syndrome was drained from the right pleural cavity 2 weeks ago. On admission, she was dyspneic at rest. Her physical development was under the third percentile, arterial pressure was 80/60 mmHg, and heart rate was 130/min. The superficial neck and upper body veins were dilated markedly. There was extensive collateral circulation. Breath sounds were diminished at the lower zone bilaterally. Hepatomegaly was measured as 4 cm. subcostally. Minimal ascites was detected in abdomen. There was more prominent edema on the right leg (deep venous thrombosis). Serum BUN, creatinin and electrolyte levels were in normal ranges. Plasma albumin was 1.9 gr/dl, total plasma protein 4 gr/dl, Hb 10.5gr/dl, WBC 22,000/mm3, platelet 438,000/mm3, ESR:125 mm/hr, cholesterol 680 mg/dl, fibrinogen 480 mg/dl, PT %100. Urinalysis: Proteinuria: 3 gm/m2/day, Sediment: WBC>30, RBC>30 per field. Bilateral pleural effusion was detected on chest radiograms. Brachial vein angiography showed occlusion of axillary vein and increased collateral circulation. Echocardiography revealed a mass arising from superior vena cava and protruding into tricuspid annulus through the right atrium (Fig.2).

Patient was undertaken emergency operation. The only venous access could be provided through the left saphenous vein cut-down. After opening the sternum, large amount of chylous fluid was aspirated from pleural and pericardial cavity. Cardiopulmonary bypass was started with an aortic and single right atrial canula. SVC and innominate vein were dissected carefully during the cooling period. Total circulatory arrest at 16°C was started and right atrium was opened. The white colored organised trombus propagating from SVC (6x4 cm) was removed completely (Fig. 3, 4). SVC and innominate vein was cleared from thrombus with an embolectomy catheter.
Figure 3. Thrombotic mass protruding through the right atriotomy just before the removal under total circulatory arrest.

Figure 4. Thrombotic mass had the similar shape of the right atrial cavity.
There was no thrombus in the inferior vena cava. The patient could be weaned from CPB without any problem. Anticoagulant therapy with heparin and corticosteroid therapy was started immediately after the operation. Oral anticoagulation was started postoperative second day. Generalised edema was resolved rapidly in the first few days (Fig. 5). In the postoperative second week, reoperation was undertaken because of mediastinitis and sternal dehiscence. Povidone iodine solution (1%) irrigation was continued for 2 days after the reoperation. Then after, the patient had a remission of her renal disease with diminishing proteinuria and normal urine output. The patient was discharged at the 31th day after the operation in good condition. Prothrombin time was kept at two fold of the initial value. She is still in good condition with remission of the NS and without recurrent thrombosis 30 months after the operation under anticoagulant and corticosteroid therapy.

**COMMENT**

The definite increase in thromboembolic episodes in NS depends on several factors: hepatic synthesis of fibrinogen, prothrombin, F VII, F VIII and F X is increased; while F XII, F XI and F IX synthesis is decreased. Antithrombin III, as a natural inhibitor of coagulation, inhibits F XII, F XI, F IX, F X, F VII and thrombin. Plasma antithrombin III level is directly related to plasma albumin level, and inversely related to amount of proteinuria. Protein S activity is also diminished in patients with NS. Total protein S level may be increased, but free active protein S level is always low in these patients. Thrombocytes may be normal or increased and functional defects can be detected. Both spontaneous and collagen, ADP induced aggregation is augmented. Hypoalbuminemia in NS causes an increase in free arachidonic acid, and thromboxane synthesis is increased. This induces platelet aggregation. It has been declared that increase in plasma lipids change thrombocyte membrane structure and augment aggregation. Fibrinolytic activity is generally reduced in patients with NS due to increased level of alpha-2 antiplasmin and inhibition of plasminogen activator, and also due to loss of fibrinogen as a fibrinolytic activator with urine. High triglyceride level and use of steroids also diminish fibrinolytic activity. In addition, hypovolemia and hemoconcentration may lead to thromboembolic episodes. In our patients, low antithrombin III level due to massive proteinuria, corticosteroid use, thrombocytosis and dehydration were the known thereby risk factor for thrombosis. Thrombosis occurs more often in mesangial proliferative glomerulonephritis than other types of NS. For the most part, this rare type of NS also has a poor prognosis with slow progression to renal failure. Our patient still shows deterioration of her renal disease with episodically increased albuminuria, and needs intermittent corticosteroid therapy 2.5 years after the operation. Despite the lower incidence, thromboembolic complications tends to be more severe in children than adults. One of the differences in thromboembolism in adults and children is localisation. Adults frequently have venous, while children usually have arterial thrombosis. SVC thrombosis has been reported in two children with NS due to a plasma exchange catheter and otitis media. No other cause other than NS could be found in our patient in...
whom we detected right atrial thrombosis propagated from SVC.

Addition of albumin and high dose corticosteroid in CPB prime fluid of our patient influenced the good outcome according to our decision. A short period of total circulatory arrest is very useful, and makes possible the complete removal of the thrombus in superior vena cava and its branches. Diuretic therapy in the early postoperative period also helped to reduce generalised oedema.

Routine prophylactic use of antiaggregant drugs are advised usually for the adult patients with NS. Pediatric use of antiaggregant drugs are only indicated in patients with documented thrombosis.\textsuperscript{12} Half dose protamin was administered at the end of CPB. Intravenous heparin followed by coumadine was administered as in the early postoperative period and used for 6 months. Continuous low dose acetyl salicylic acid therapy was started at the same time.

Excessive tissue oedema due to hypoalbuminemia and superior caval syndrome, and cyclosporin use caused defective wound healing and reduced immune resistance. Staphylococcus aureus mediastinitis occurred and was treated with surgical debridement, 1 % povidone iodine irrigation and vancomycin therapy. Remission of the disease with increased diuresis and decreased proteinuria also played a promising role in this period.

As a result, we described a case of SVC and right atrial thrombosis in a child with NS who has been followed up for 30 months without recurrence echocardiographically after complete surgical removal of the organised thrombus under total circulatory arrest.

REFERENCES


