

“Right atrial-jugular vein graft bypass” for treatment of superior vena cava syndrome caused by protein S deficiency

*Protein S yetmezliğine bağlı gelişen vena kava süperiyör sendromunun tedavisi için
“sağ atriyo-jugüler ven greft bypass”*

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Superior vena cava syndrome from protein S deficiency was diagnosed in a 32 year-old female. A three month period of medical treatment did not relieve her symptoms. We performed a “right atrial-jugular vein graft bypass” which relieved her symptoms within 10 days. The patient did not develop any complications in the 18 month postoperative period. We present this case with a review of the relevant literature.

Key words: Protein S deficiency; superior vena cava syndrome; venous surgery.

Superior vena cava (SVC) thrombosis is a rare condition caused by coagulation disorders. Protein S is a cofactor of activated protein C. Spontaneous venous thrombosis may occur in the central veins of a patient with protein S deficiency.^[1]

Initial medical treatment of these patients includes thrombolytic, anticoagulant and steroid drug therapies. If the patient’s symptoms do not improve with medication, surgical or endovascular interventions are performed.

We present a case in which right atrial-jugular vein prosthetic graft bypass was performed successfully for surgical treatment of SVC syndrome caused by protein S deficiency.

CASE REPORT

A 32 year-old woman with SVC syndrome presented with facial, neck and bilateral shoulder edema, dyspnea, chest pain, and headaches for three months. She had a history of facial flushing when in the supine position. There was a history of bilateral deep vein thrombosis (DVT) one year ago.

Otuz iki yaşında kadın olguya protein S eksikliğine bağlı olarak gelişmiş vena kava süperiyör sendromu tanısı konuldu. Üç aylık tıbbi tedavi sürecinde hastanın semptomlarında gerileme sağlanamadı. Sağ atriyo-jugüler ven greft bypass uygulanan hastanın yakınmaları 10 günde geriledi. Ameliyat sonrası 18 aylık takip süresince herhangi bir komplikasyon gelişmedi. Olgu ilgili literatür eşliğinde sunuldu.

Anahtar sözcükler: Protein S yetmezliği; vena kava süperiyör sendromu; venöz cerrahi.

Computed tomography (CT) of her chest, Doppler ultrasound examination of her neck veins and bilateral upper extremity venography revealed a thrombus in the superior vena cava, innominate vein and proximal part of subclavian veins. Dilation of both internal jugular veins were also observed (Fig. 1). The thrombus was detected in the deep venous system of the lower extremities by Doppler ultrasound.

Initial hematological values included: white blood cell count of 9.400/mm³, hemoglobin 12.4 g/dl, platelets 214.000/mm³, and reticulocytes 13%. Liver and renal function test results, amylase and lipase values were normal. We found a deficiency of protein S activity (32%, normal range within 55%-160%), but antithrombin III activity and protein C activity were normal.

At the beginning of the treatment, heparin and steroids were administered and warfarin was given as the maintenance therapy. The patient’s complaints did not resolve with medical treatment.

We performed an 8 mm ring-enforced expanded polytetrafluoroethylene graft (ePTFE) bypass from

Received: November 10, 2006 Accepted: November 16, 2006

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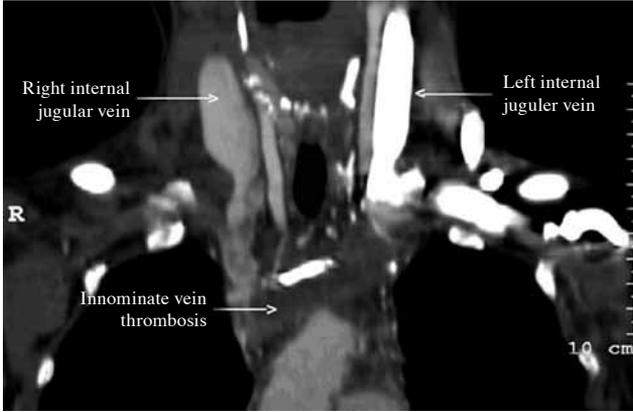


Fig. 1. Computed tomographic scan of thorax with contrast showing a thrombotic occlusion of innominate vein and dilation of both internal jugular veins.

the left internal jugular vein to the right atrium (Fig. 2). The operation was performed through a median sternotomy. A 100 U/kg heparin sodium solution was administered intravenously after opening the pericardium. Thymic tissue and its vessels were not ligated. The ePTFE graft was passed through the surgically prepared tunnel from the left neck region to the mediastinum. A side-biting Satinsky clamp was placed on the right atrial appendage, and was opened longitudinally. Some trabecular muscles were excised, and the anastomosis with the ePTFE was performed. The proximal and distal anastomoses of the graft were performed to the right-atrial appendage and the left internal jugular vein respectively with the end-to-side technique.

The postoperative period was uneventful. The swelling in the patient's face and neck began to subside during the first postoperative day. The patient was symptom-free within 10 days after operation. The patient was discharged with warfarin. Graft patency during follow-up

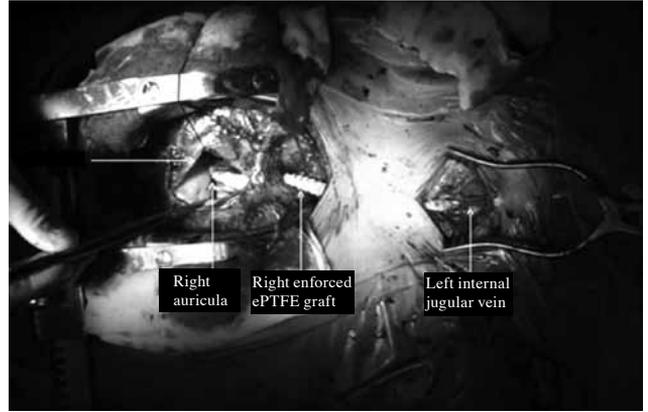


Fig. 2. Operation view.

was controlled by Doppler ultrasound. Follow-up after 18 months showed no complications (Fig. 3).

DISCUSSION

Superior vena cava syndrome is clinical diagnosis of arm and facial swelling with distension of the neck veins. The common causes of SVC syndrome are malignancies such as medullar or follicular carcinoma of the thyroid, mediastinal lymphoma, thymoma, teratoma, angiosarcoma, and synovial cell carcinoma.^[1] Benign disease is a rare cause of SVC syndrome, accounting for only 22% of cases.^[2] The most frequent benign causes of SVC syndrome are mediastinal fibrosis, granulomatous disease and coagulation disorders. Recently, the incidence of iatrogenic SVC syndrome caused by using central venous catheters and pacemaker lines has increased.^[1] Right subclavian vein thrombosis has been observed in a patient with end-stage renal disease due to a pericatheter thrombosis after the permanent catheter placement.^[3]

Abnormal thrombosis may occur secondary to deficiencies of protein S, protein C and antithrombin III.^[4]



Fig. 3. (a) Preoperative and (b) postoperative view of the patient.

Protein S is a co-factor for the natural anticoagulant protein C which primarily inhibits factor V. Protein S synthesis mainly occurs in the liver, but also occurs in endothelial cells and megakaryocytes. Protein S deficiency is an autosomal dominant trait. Spontaneous venous thrombosis commonly occurs in protein S deficiency while arterial thrombosis is rare.^[1,4] Engesser et al.^[5] reported that of patients with protein S deficiency; 71% had a DVT, 72% had superficial thrombophlebitis, and 38% had pulmonary embolus. Bilateral DVT, inferior vena cava, renal vein and isolated internal jugular vein thrombosis caused by protein S deficiency have been reported in the literature.^[6,7] Kalra et al.^[8] reported only one out of 32 patients who has a hypercoagulable state with SVC caused by benign disease. In our case, there was a thrombus in SVC caused by protein S deficiency, innominate vein and proximal part of subclavian veins.

A clear treatment algorithm has not evolved for this patient group. Recently, the evolution of the role of angioplasty, stents and covered stents has provided an alternative to surgical treatment. These interventions have been used successfully, especially in cancer patients with a life expectancy of less than one year.^[1,8,9]

Spiral saphenous vein graft and ePTFE grafts are commonly used for surgical treatment of SVC syndrome. Although spiral venous grafts are the best autologous conduits with low thrombogenicity, results of several series of implanted ePTFE grafts showed excellent patency, being the least thrombogenic prosthetic graft for SVC reconstruction.^[1,10] Ring-enforced ePTFE grafts are less collapsible than other synthetic and saphenous grafts.^[11] Wisselink et al.^[12] reported 100% patency at one year for ePTFE bypass grafts placed to treat central vein occlusion in six patients. We wanted to use a spiral venous graft, but we could not because of the patient's previous deep venous thromboses in both lower extremities. A ring-enforced ePTFE graft was preferred for right atrial-jugular vein bypass grafting procedure in our case because of its protection of the graft from kinking or compression. For instance, Panneton et al.^[13] used a modified sapheno-jugular bypass procedure with saphenous vein graft tunneled through a PTFE. Unilateral reconstruction is sufficient to relieve the symptoms in most patients with SVC syndrome because collateral circulation in head and neck is almost always adequate for venous return.^[1] For these reasons, we preferred the left internal jugular vein for the proximal anastomotic side.

In conclusion, a right atrial-internal jugular graft bypass with a ring-enforced ePTFE graft in patients

with SVC syndrome caused by protein S deficiency may be performed successfully.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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