Case Report

Chylous pericardial tamponade in a haemodialysis patient with catheter-associated thrombosis of internal jugular and subclavian veins

Najin Lee and Maria Coco

Department of Medicine, Renal Division, Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, NY, USA

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Introduction

Chylous pericardial effusion may occur after cardiothoracic surgery or in association with congenital lymphangiomatosis. Other causes may include chest trauma, mediastinal radiation, malignant neoplasm of the mediastinum or thrombosis of the subclavian vein and superior vena cava [1]. Idiopathic chylopericardium has also been described, most commonly in children and young adults [1]. Cardiac tamponade due to chylopericardium has been described in several of these cases.

Pericardial disease with effusion and tamponade is not a rare complication in end-stage renal disease (ESRD) patients [2]. The aetiology of pericardial effusions can be transudative with no known aetiology, exudative secondary to malignancy or infection, or haemorrhagic secondary to uraemia. Chylous pericardial effusion in patients with ESRD is distinctly rare, with one case referenced in the surgical literature [3]. There are no reports of pericardial tamponade due to chylous effusion in dialysis patients. We present a patient admitted with chylopericardium.

Case report

A 25-year-old African-American man, with ESRD secondary to hypertension, was admitted with a complaint of left neck and left arm pain and swelling for 3–4 days. He denied recent trauma, fever or chills. He had been on warfarin until 2 months earlier for deep vein thrombosis of a lower extremity. Medications at the time of admission were metoprolol, sevelamer, clonidine, fosinopril, simvastatin and erythropoietin. The patient had begun haemodialysis <1 year prior to the current admission via a left internal jugular vein tunneled catheter. This had been removed due to catheter-related sepsis 5 months before this presentation. A right internal jugular vein tunneled catheter was used for maintenance haemodialysis. A left forearm arteriovenous fistula created shortly after the initiation of haemodialysis had never matured and had thrombosed 1 month prior to this admission.

On physical examination, the patient was afebrile with normal blood pressure, heart rate, and oxygenation. He had diffuse oedema and mild tenderness to palpation over his left neck and upper arm. The thrombosed arteriovenous fistula was not infected. A tunneled catheter with a clean exit site was present in the right anterior chest wall. The remainder of his physical examination was unremarkable: no jugular venous distention, murmurs or rubs on cardiac examination, clear lungs, benign abdomen and no lower extremity oedema.

Laboratory evaluation included normal liver function tests, haemoglobin 15.4 g/dl, white blood cells 14 200/µl, normal platelets, and normal prothrombin and partial thromboplastin times. Chest X-ray showed increased density and swelling of the soft tissues overlying the left neck, chest and arm; a small left pleural effusion; cardiomegaly with mediastinal widening and a right internal jugular venous catheter with its tip in the right atrium. A computed tomography (CT) scan of the neck using intravenous contrast demonstrated thrombosis of the right internal jugular vein, extensive oedema of the soft tissues of the left neck, and lymphadenopathy of the left superior mediastinum and left supraclavicular regions. Venous ultrasound examination of the neck and upper extremities demonstrated diminished flow in the right and left internal jugular and subclavian veins. The patient was placed on intravenous anticoagulation. Dialysis was continued via femoral vein access.

On hospital day 3, the patient developed chest pain, dyspnoea, haemoptysis and hypotension. A helical CT scan of the chest with intravenous contrast...
and a repeat CT of the neck showed thrombosis of the left innominate vein, superior vena cava (Figure 1) thrombus in the right atrium adjacent to the dialysis catheter tip, prominent collateral venous circulation in the mediastinum and anterior chest wall, and pleural effusion (Figure 2), and a large pericardial effusion (Figure 3). Contrast was seen in the right internal jugular vein on this CT scan, implying partial recanalization of the vein (Figure 4). Transthoracic echocardiogram confirmed the large pericardial effusion with tamponade physiology.

The patient underwent emergency pericardial drainage and window formation. 550 ml of milky fluid were drained. Laboratory analysis reported a triglyceride level of 1313 mg/dl and lymphocyte predominance (78%) on the cell count differential, consistent with chyle.

Cultures of the fluid were positive for *Staphylococcus epidermidis*, but negative for fungus, acid-fast bacilli or viruses. All blood cultures were negative. Due to concern for possible infectious thrombophlebitis of the central veins with seeding of the pericardium, the patient was treated with intravenous vancomycin. Follow-up cultures of the pericardial fluid and peripheral blood were negative. Evaluation for hypercoagulability including assays for inherited thrombophilias, hyperhomocysteinaemia, antiphospholipid syndrome, and heparin-induced thrombocytopenia was negative. Pathological examination of the pericardial
tissue showed fibrosis and chronic inflammation; there were no granulomata or neoplasm. The patient tested negative for antinuclear antibodies, and his thyroid function tests were normal.

The patient continued to drain about 250–500 ml of chylos fluid per day. A transoesophageal echocardiogram on post-operative day 7 showed minimal pericardial effusion, and the previously identified thrombi in the superior vena cava, right atrium and at the dialysis catheter tip. Also noted was a 1 cm mobile thrombus attached to the main thrombus at the catheter tip. Because of persistent pericardial fluid production despite drainage, the patient underwent surgical exploration of the central veins on hospital day 25. Thrombi in the right internal jugular vein, superior vena cava and left innominate vein were evacuated and the right internal jugular vein tunnelled catheter was removed. Repeat transthoracic echocardiogram showed no pericardial effusion. The chest tubes drained progressively smaller amounts of serosanguinous fluid with no evidence of chylos drainage, and were subsequently removed.

The patient was discharged home on post-operative day 6 in good condition, on anticoagulation with warfarin. He was returned 2 days later in a state of cardiac arrest with recurrent cardiac tamponade secondary to haemorrhagic pericardial effusion. Despite efforts at resuscitation and repeat pericardiostomy, the patient died. The effusion was not sent to the laboratory for further chemical analysis. Autopsy was not performed.

Discussion

We describe the first ever reported case of an ESRD patient, on haemodialysis, who presented with chylos pericardium and developed cardiac tamponade. This patient had extensive thrombosis of the central and peripheral veins associated with multiple vascular dialysis accesses, as well as a history of deep vein thrombosis of a lower extremity. The left innominate vein thrombosis, which was the probable aetiology of the chylopericardium, may have been related to the previous left internal jugular vein catheter. The superior vena cava thrombosis may also have formed at the same time, but the acute presentation of this patient suggests that complete venous occlusion and lymphatic obstruction did not occur until immediately prior to this admission. Alternately, the symptoms of pain, tenderness and swelling along with the finding of extensive lymphadenopathy may have been due to possible infection of previously asymptomatic venous thrombi.

There are a very few reported cases of chylopericardium in association with superior vena cava and/or subclavian vein thrombosis. Four were associated with venous catheters [4–7], two with Behcet’s disease [8,9], and one with a large anterior mediastinal tumour presumed to be Hodgkin’s lymphoma or thymoma [10]. There are four other reported cases where catheter-associated thrombosis was suspected but no venograms or other imaging were performed [11–14].

The hypothesized underlying cause of the chylopericardium is obstruction of the thoracic duct ostium. Pericardial lymphatics drain via mediastinal and tracheobronchial lymph nodes into the thoracic duct [15]. The thoracic duct then drains into the left subclavian or internal jugular vein or at the junction of the two, although considerable anatomic variability exists. With obstruction of the thoracic duct ostium, there is presumed reflux of chyle back into the pericardium [1]. However, the mechanism of chylopericardium formation is not entirely clear. The majority of cases in which lymphangiography was performed failed to show a macroscopic connection between the pericardium and the thoracic duct. In addition, valves in the lymphatic vessels prevent reflux of chyle unless the pressure in the lymphatics exceeds 15 cm of water [1]. Also, thoracic duct ligation in animals results in the collateral lymph drainage into the azygos venous system rather than the formation of chylos effusions [1]. Thus, events leading to chylopericardium require further study.

Treatment for chylopericardium varies with the aetiology. The best outcome in idiopathic chylopericardium has been achieved with pericardial window formation and low thoracic duct ligation [16–20]. The recurrence rate is about 50% if pericardial window alone is used [1]. The treatment of secondary chylopericardium is more conservative. Most cases are managed with drainage of the effusions via pericardiocentesis or pericardiostomy. There is one report of a pericardio–peritoneal shunting used in four paediatric cases—one post-surgical and three with lymphangiomatosis—with no recurrence in any of the four cases [21].

In cases of thrombosis-associated chylopericardium, treatment is similar to that of other cases of secondary chylopericardium. Additional treatment has included removal of the indwelling catheter if present, anticoagulation and immunosuppressive treatment for those cases associated with Behcet’s disease as well as stent placement within a thrombosed superior vena cava [4]. These cases reported no recurrence at follow-up ranging from 9 weeks to 4 years [4,5,7–10]. In our patient, pericardial window formation alone was insufficient. Surgical evacuation of the venous thromboses with removal of the catheter and anticoagulation therapy were necessary for the resolution of the chylopericardium. It remains unclear whether the subsequent haemorrhagic pericardial effusion was related to the original condition.

Despite the frequent use of central venous catheters for haemodialysis and the common occurrence of catheter-associated venous thrombosis [22,23] as well as pericardial effusions in dialysis patients [2], there has been only one other reported case of chylopericardium in a dialysis patient. The patient was treated with thoracoscopic clipping of the thoracic duct and creation of a pericardial window, with no recurrence after follow-up of 7 months. No further details regarding dialysis access or presence of thrombus were given in the report [3].
Dialysis catheters are very commonly used in maintenance haemodialysis, often with significant morbidity including thromboses of the central veins, intracardiac thrombi and infections. We suggest that chylopericardium, although rare, be included among the serious sequelae of catheter use.

Conflict of interest statement. None declared.

References


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