JACC: CASE REPORTS VOL. 29, 2024

CORONARY, PERIPHERAL, AND STRUCTURAL INTERVENTIONS

CASE REPORT: CLINICAL CASE

Chylopericardium and Superior Vena Cava Syndrome Caused by Central Venous Occlusion From Indwelling Hemodialysis Catheters

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ABSTRACT

Chylopericardium is a rare condition. Causes include superior vena cava syndrome resulting from indwelling catheters. We present a case of this condition in a 42-year-old man with end-stage renal disease treated with hemodialysis through a right subclavian vein catheter. He underwent successful endovascular stenting with resolution of his symptoms and chylopericardium. (JACC Case Rep. 2024;29:102483) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 42-year-old man presented with neck swelling, throat tightness, and dyspnea. He was hemodynamically stable and afebrile. On examination, he had face and neck swelling, elevated jugular venous pressure, diminished breath sounds, and a right-sided chest tunneled hemodialysis catheter. Laboratory testing revealed the following: white blood cell count, 6,000/L; hemoglobin, 10.7 g/dL; creatinine, 9.22 mg/dL; and blood urea nitrogen, 31 mg/dL. An electrocardiogram demonstrated normal sinus rhythm. Chest computed tomography (CT) revealed large pericardial and bilateral pleural effusions, diffuse lymphadenopathy, retropharyngeal inflammation, chronic left subclavian vein occlusion, and superior vena cava (SVC) narrowing at the cavoatrial junction surrounding his dialysis catheter (**Figure 1**). An echocardiogram revealed moderate circumferential pericardial effusion with tamponade physiology.

TAKE-HOME MESSAGES

- Hemodialysis catheters can lead to the development of SVC syndrome and chylopericardium, which is diagnosed by pericardial fluid analysis.
- Endovascular stenting is an effective treatment for this condition.

PAST MEDICAL HISTORY

His past medical history included end-stage renal disease (ESRD) treated with long-term hemodialysis through a right subclavian vein catheter (maturing right arm arteriovenous fistula [AVF]), hypertension, type 2 diabetes, psoriasis, and obesity.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received June 3, 2024; accepted July 9, 2024.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2024.102483

ABBREVIATIONS AND ACRONYMS

AVF = arteriovenous fistula

CT = computed tomography

CVC = central venous catheter

ESRD = end-stage renal disease

SVC = superior vena cava

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of his pericardial/pleural effusions included purulent (from bacterial or tuberculous infection), reactive from viral infection, chylopericardium/chylothorax, malignant, autoimmune-mediated, or idiopathic. Given his central venous occlusion/stenoses, SVC syndrome was considered; causes included both malignant and benign (eg, fibrosing mediastinitis, thrombosis, or stenosis/sclerosis secondary to long-term central venous catheter [CVC] placement) conditions.

INVESTIGATIONS

Pericardiocentesis yielded 358 mL of white turbid fluid (Figure 2); analysis revealed a triglyceride level of 1,296 mg/dL, 81% lymphocytes, positive chylomicrons, and no bacteria or acid-fast bacilli. Cytologic examination was negative for malignancy. Thoracentesis revealed a pleural fluid triglyceride level of 284 mg/dL with negative bacterial culture results. A left supraclavicular lymph node biopsy and flow cytometry revealed benign lymphoid tissue.

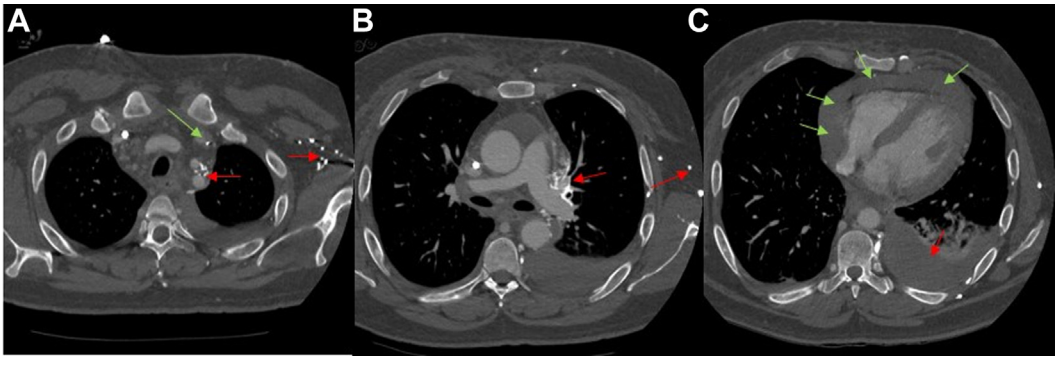
MANAGEMENT

The patient was initially treated with antibiotics out of concern for purulent pericarditis; these drugs were ultimately discontinued given his clinical stability. Results of multiple bacterial blood cultures were negative for growth. Once his pericardial/pleural fluid analyses were completed, his presentation was thought to be most consistent with chylopericardium/chylothorax, and he was started on a low-fat diet. Venography showed SVC occlusion and moderate bilateral brachiocephalic and subclavian vein stenoses with prominent collateral drainage pathways. SVC syndrome was thus diagnosed. SVC balloon venoplasty was attempted but resulted in minimal improvement in SVC narrowing. He was discharged according to his preference with plans for venous reconstruction. He presented again with recurrent chylopericardium and tamponade several weeks later, however, and underwent balloon venoplasty up to 8 mm in the bilateral brachiocephalic veins and SVC (Figure 3), followed by placement of 3 Medtronic Abre venous stents in a "kissing" formation in the SVC and bilateral brachiocephalic veins under intravascular ultrasound guidance. The stents were dilated to 8 mm in the SVC, 10 mm in the right brachiocephalic vein, and 12 mm in the left brachiocephalic vein after placement. Finally, a new hemodialysis catheter was placed in the initial location with tip termination in the right atrium. This procedure resulted in restoration of in-line venous flow from the subclavian veins to the right atrium on venography (Figure 4), rapid improvement in pericardial drain output, and symptom resolution. He was started on apixaban for 6 months.

DISCUSSION

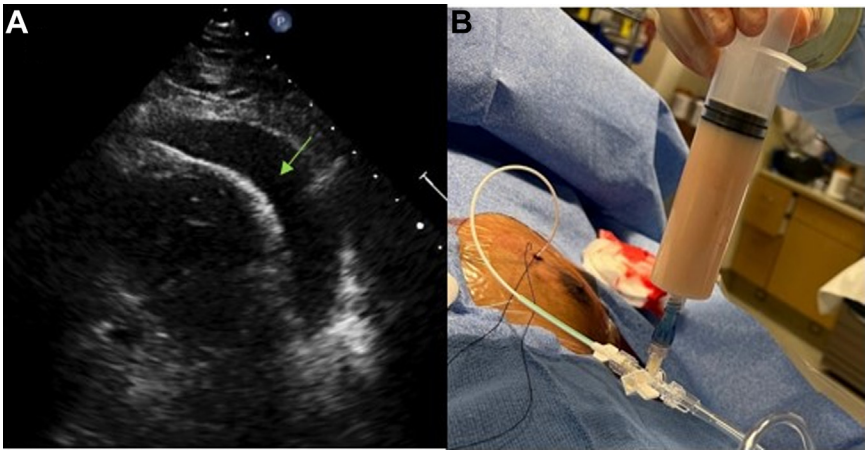
Chylopericardium is a rare condition characterized by pericardial chyle accumulation. Causes include trauma, idiopathic conditions, cardiothoracic surgery, mediastinal neoplasms, tuberculosis, and rarely anatomic causes of elevated central venous pressure (eg, central venous thrombosis/stenosis begetting SVC syndrome). A total of 64% of patients with

FIGURE 1 Initial Computed Tomography Scan



The scan revealed (A) brachiocephalic vein occlusion (green arrow) and collateral vein formation (red arrows); (B) collateral vessels are also shown by red arrows. The scan also showed (C) a circumferential pericardial effusion (green arrows) and a left pleural effusion (red arrow).

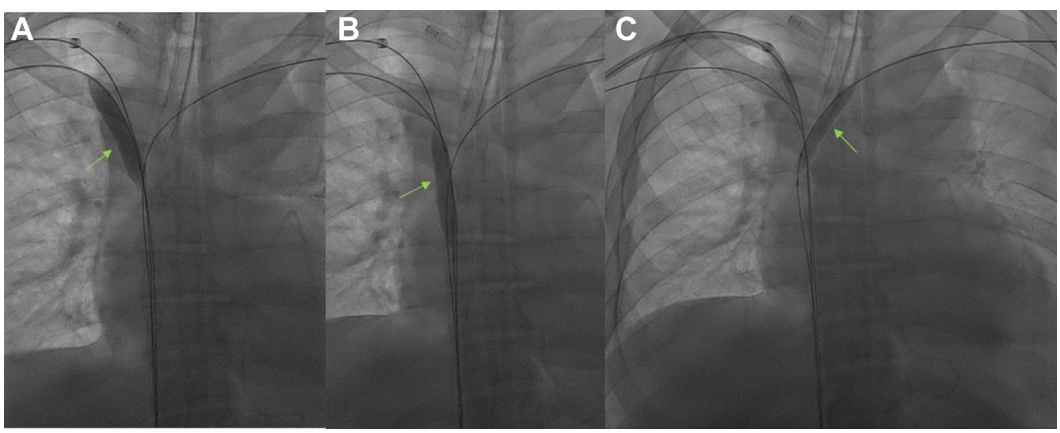
FIGURE 2 Pericardiocentesis Findings



(A) Echocardiogram showed a lateral pericardial effusion (arrow), which revealed (B) milky, white, turbid chylous fluid when drained.

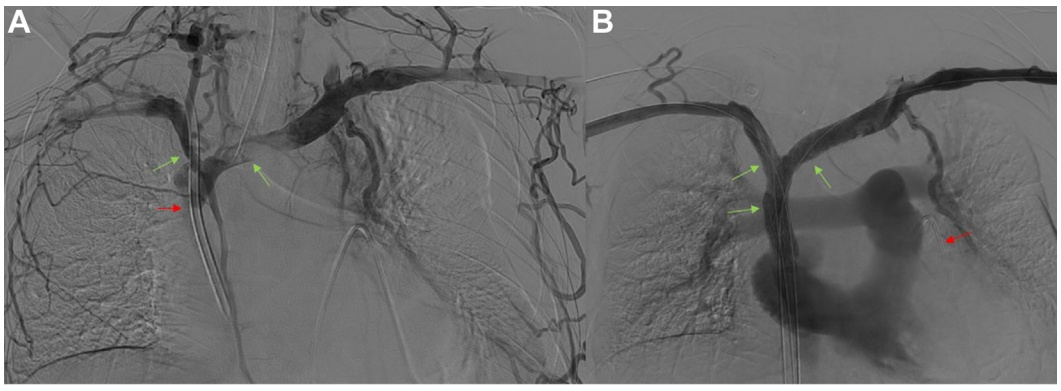
chylopericardium present with dyspnea, and 33% present with tamponade, as our patient did.[1] The following pericardial fluid findings confer a diagnosis of chylopericardium: milky white appearance, triglycerides >500 mg/dL, lymphocytic predominance, negative bacterial culture results, and a cholesterol-to-triglyceride ratio <1.0; in a diagnostic algorithm, each criterion counts for 1 point, and a score ≥2 points is required for diagnosis (100% specificity and sensitivity).[1] Our patient met all but the last of these criteria, thus confirming his chylopericardium diagnosis. Other diagnostic modalities include lymphangiography, lymphangioscintigraphy, and ingestion of Sudan III dye or iodine-131-labeled triolein.[1,2] The optimal treatment for chylopericardium is unknown. Conservative therapy can be attempted with pericardiocentesis and a pericardiostomy tube, medium-chain triglyceride diet initiation, or total parenteral nutrition, but these measures often fail. If chylopericardium develops again, surgical thoracic

FIGURE 3 Balloon Dilation During Central Venous Reconstruction



Balloon dilation (arrows) was performed before stent deployment in the (A) right brachiocephalic vein, (B) superior vena cava, and (C) left brachiocephalic vein.

FIGURE 4 Preintervention and Postintervention Venography



(A) Preintervention venography revealed bilateral brachiocephalic vein stenoses (green arrows) and superior vena cava occlusion (red arrow). (B) Postintervention venography revealed patent stents in the bilateral brachiocephalic veins and superior vena cava (green arrows), thus resulting in restoration of in-line venous flow from the subclavian veins to the right atrium. A pericardial drain (red arrow) is also seen.

duct ligation with pericardial window creation is recommended and is effective at preventing recurrence.[1]

Treatment of secondary chylopericardium centers on addressing the underlying condition.[1] Our patient's case is unique because his chylopericardium was associated with central venous occlusion/stenoses related to multiple previous dialysis catheters that resulted in SVC syndrome. This was likely triggered by increased venous flow from a maturing AVF ipsilateral to his CVC, which generated elevated thoracic duct pressure and chyle reflux into the pericardial/pleural lymphatic vessels. Successful SVC/brachiocephalic venous stenting led to resolution of his chylopericardium.

Although SVC stenosis from hemodialysis catheters is not uncommon, CVC-induced SVC syndrome leading to chylopericardium/chylothorax is exceedingly rare. To the best of our knowledge, only 4 previous cases have been reported (only 2 of which were in patients like ours whose CVCs were used for hemodialysis).[3-7] A similar case related to pacemaker leads has also been reported.[8] Benign causes account for 30% of SVC syndrome cases, and CVC-induced SVC obstruction is related to neointimal hyperplasia after repetitive trauma from high-flow states.[3,9] Although no formal guidelines exist regarding optimal treatment of SVC syndrome from a benign cause, expert opinion recommends endovascular stenting as first-line therapy; this has a high technical success rate, with symptomatic relief in >90% of patients and low restenosis or SVC syndrome recurrence rates.[9] Surgical bypass is reserved for cases where endovascular stenting fails. Because of the central venous system's high degree of elastic recoil, balloon angioplasty alone is generally ineffective.[9]

Limited data exist on long-term outcomes of patients with chylopericardium from CVC-induced SVC syndrome, but in 2 previously reported cases (1 patient treated with balloon venoplasty and another with SVC stenting), follow-up at 6 and 42 months post-intervention, respectively, revealed stent patency in the second case and no recurrent chylopericardium/chylothorax or symptoms.[3,4]

FOLLOW-UP

At a follow-up appointment 3 weeks post-stenting, our patient noted sustained resolution of his face/neck edema. Chest CT with contrast enhancement 6 months post-stenting is ordered to assess stent patency.

CONCLUSIONS

Chylopericardium is a rare complication of long-term CVCs, and it can manifest with tamponade. If it is found in a patient with ESRD who has a hemodialysis catheter (especially if an ipsilateral maturing AVF is present), SVC syndrome should be suspected, and SVC and/or brachiocephalic vein stenosis should be sought. If this condition is identified, endovascular stenting is a potentially curative therapy. Prompt recognition and management of this condition can lead to improvement in outcomes for some of our most chronically ill patients.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS chylopericardium, chylothorax, pericardial effusion, stenosis, stents, superior vena cava syndrome, tamponade