

Case report

Surgical management of chylopericardium and chylothorax in a patient with Behçet's disease

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ABSTRACT

Behçet's syndrome is a chronic multisystem inflammatory disease characterised classically by recurrent oral and genital ulcers with ocular lesions. It can affect blood vessels of all sizes, but involves veins more commonly than arterie. The presence of chylothorax in Behçet's syndrome is rare, with only a few cases cited in the literature. The most likely pathogenesis is SVC thrombosis with obstruction of the orifice of the thoracic duct resulting in leakage of chyle from the pleural lymphatics into the pleural space. The majority of the previously reported cases were managed medically without surgical intervention. We believe that this report describes the first use of surgery to ligate the thoracic duct and create a pericardial window in a Behçet's syndrome with chylothorax and chylopericardium.

Case presentation

A 20-year-old man was admitted to hospital with a 3 month history of feeling generally unwell with neck swelling, facial redness, difficulty of swallowing, fevers, mouth ulcers as well as widespread ulcers over the genitourinary regions. A diagnosis of Behçet's syndrome had been made three months prior to admission by a consultant dermatologist, and he was started on cyclosporine 75 mg BD. He also had been taking oral prednisolone 30 mg OD, co-chicine 0.5 mg TDS and topical agents to treat the oral and genital ulcers. His past medical history and family history were unremarkable, other than admission 2 months prior with epididymo-orchitis. Blood tests on admission revealed raised inflammatory markers, with a white cell count of $22.9 \times 10^9/L$ and C-reactive protein of 211mg/L. The patient developed hypoxia and

the subsequent chest radiograph demonstrated left lower lobe consolidation along with some pleural effusion. A CT and MRI scan of the thorax, abdomen and pelvis confirmed the presence of large left pleural and pericardial effusions (Figs. 1-3). A Venogram was performed as though Doppler is able to demonstrate flow, it is limited in delineating the anatomy and location of obstruction. Venogram imaging revealed thrombosis of the left brachiocephalic vein and superior vena cava, with extensive mediastinal and pericardial collaterals (Fig. 4).

In an attempt to drain the left pleural effusion a 12 F Seldinger Portex chest drain was inserted and over 6 litres of chylous fluid was drained over the next 4 days. The patient subsequently developed sinus tachycardia (240 beats per minute). He was then transferred to a tertiary centre with expertise in Behçet's syndrome. Soon after his arrival, the patient developed clinical signs of tamponade and was confirmed by transthoracic echocardiogram. An emergency pericardiocentesis was performed and 1.1L of chyle was drained with immediate relief of tamponade and symptoms. In view of the life threatening chylopericardium and continued high chyle output from the left pleural drain, the patient was referred for definitive surgical management.

The patient underwent a right thoracotomy and the thoracic duct was identified in the posterior mediastinum between the descending thoracic aorta and the azygos vein. The duct was ligated and divided. A pericardial window was constructed to allow pericardial fluid to drain into the right pleural cavity. On the fourth postoperative day the pericardial and pleural drains were removed and transthoracic echocardiogram and chest radiograph confirmed

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Competing interests: none declared.

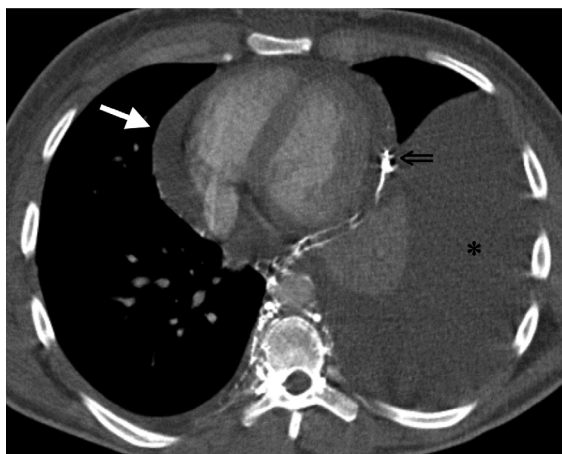


Fig. 1. Contrast enhanced CT scan showing chylous pericardial effusion (white arrow), large left pleural effusion (*) and high density from previous lymphangiogram contrast material in mediastinal lymphatics (⇐), collapsed left lung (black arrow).

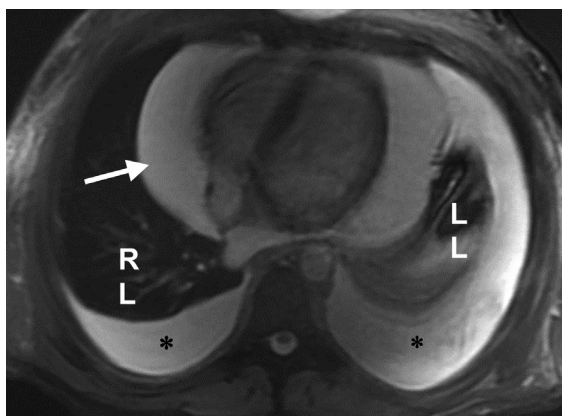


Fig. 2. Transverse T2-weighted MR scan showing large chylous pericardial effusion (white arrow), bilateral chylous pleural effusions (*), portion of collapsed left lung (LL), normal right lung (RL).

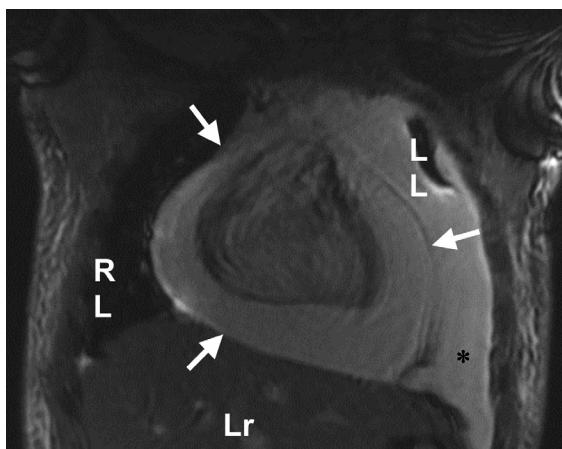


Fig. 3. Coronal T2-weighted MR scan showing the large chylous pericardial effusion (white arrow), large left chylous pleural effusion (*), portion of collapsed left lung (LL), normal right lung (RL), normal liver (Lr) beneath the diaphragm.

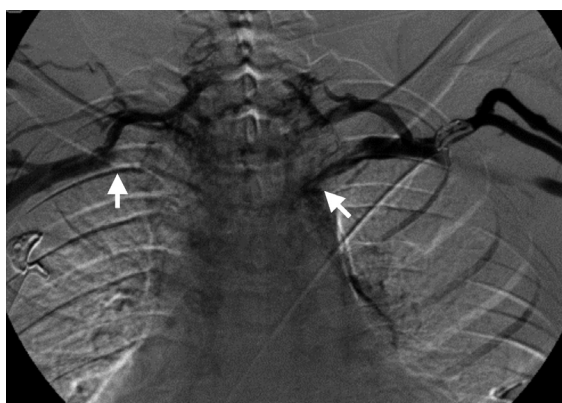


Fig. 4. venogram, with intra-venous contrast injected bilaterally into each arm simultaneously, showing bilateral occlusion of both brachiocephalic veins (arrows) and the superior vena cava.

the complete resolution of pericardial and pleural effusions respectively. He was anticoagulated with warfarin for the SVC obstruction and started on azathioprine 50 mg TDS, to be followed up by the rheumatology team in 3 months as an outpatient. The patient went onto make a good recovery and was discharged home in a stable condition.

Discussion

Behçet's syndrome is a chronic multi-system inflammatory disease characterised classically by recurrent oral and genital ulcers with ocular lesions (1). It can affect blood vessels of all sizes, but involves veins more commonly than arteries (2). The presence of chylothorax in Behçet's syndrome is rare, with only a few cases cited in the literature (3-11). The most likely pathogenesis is SVC thrombosis with obstruction of the orifice of the thoracic duct resulting in leakage of chyle from the pleural lymphatics into the pleural space.

A majority of the previously reported cases were managed medically without surgical intervention (3-7, 11). Maayan *et al.* successfully used injection of autologous blood into the chest tube to treat persistent chylothorax (10). Oz *et al.* described the use video-assisted surgery for creating a pericardial window for the treatment of cardiac tamponade caused by chylous pericardial effusion (8). Moon *et al.* performed thoracic duct ligation via a thoracotomy in a young patient presenting with chylopericardium and chylothorax as the initial presentation (9).

In the management of this case, after multidisciplinary team discussion between respiratory and rheumatology physicians and cardiothoracic surgeons, it was decided that due to the large volume of pericardial effusion, performing a different approach such as talc pleurodesis was not considered. We believe that this report describes the first use of surgery to ligate the thoracic duct and create a pericardial window in a Behçet's syndrome with chylothorax and chylopericardium. We believe that this is a safe and effective definitive technique in managing this potentially life threatening complication.

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