CASE REPORT

Chylothorax in a patient with HIV-related Kaposi's sarcoma

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SUMMARY

We present a case of a 33-year-old man with a background of HIV and Kaposi's sarcoma (KS), who presented with a right sided chylothorax. He was managed with percutaneous chest drainage and talc pleurodesis, in addition to his chemotherapy and antiretroviral therapy for KS and HIV, respectively. Good clinical control of the chylothorax remained 4 months post drainage. This case report summarises the approach to investigating and managing pleural effusion, and in particular chylothorax, in HIV patients.

BACKGROUND

Kaposi's sarcoma (KS) is caused by human herpes virus-8, often in HIV or immunosuppressed individuals. With advances in HIV treatment, the rate of HIV-associated KS has reduced. Pleural effusions in HIV patients may occur for a variety of reasons; however KS-related pleural effusion, particularly chylothorax is less common, with few published case series.

CASE PRESENTATION

We present a case of a 33-year-old Senegalese man with a background history of HIV and KS diagnosed in May 2017. At the time of his diagnosis, he presented acutely with weight loss, night sweats, bilateral leg swelling and violaceous lesions over his legs. The HIV test was positive and skin biopsy confirmed KS. CT chest, abdomen and pelvis demonstrated widespread parenchymal perilymphatic nodularity with consolidation, perifissural beading, a small right pleural effusion and mediastinal and hilar lymphadenopathy. There was also mild splenomegaly, abdominal lymphadenopathy and lytic lesions in the axial skeleton (figure 1). Bronchoscopy was performed to exclude atypical infection and there were no typical pulmonary Kaposi lesions on endobronchial inspection. The patient was commenced on highly active antiretroviral treatment (HAART—Dolutegravir and Truvada) and liposomal doxorubicin chemotherapy for KS. At the time of diagnosis his HIV viral load was <40 copies/mL with severe CD4 lymphopenia $0.20 \times 10^9/L$ making him at risk of opportunistic infection. Interferon gamma release assay, QuantiFERON TB Gold, was negative.

In March 2018, the patient presented with dyspnoea and right sided chest discomfort. He reported poor compliance with chemotherapy and

variable compliance with HAART. Examination revealed stony dull percussion in the right hemithorax with reduced breath sounds. There was bilateral leg swelling with persistent violaceous lesions over the thigh.

INVESTIGATIONS

Chest radiograph revealed a lenticular pleural effusion occupying greater than one-third of the hemithorax (figure 2). A diagnostic pleural aspiration was opalescent with lactate dehydrogenase 156 iu/L, protein of 66 g/L and triglycerides of 11.7 mmol/L consistent with a chylothorax. Pleural fluid cytology was negative with proteinaceous debris. There was no growth of bacteria or tuberculosis on pleural fluid culture. Repeat CT chest, abdomen and pelvis, when compared with his initial CT in 2017 demonstrated improvement in the parenchymal changes with resolution of previous lymphadenopathy but residual right sided effusion (figure 3). Serum albumin was 36 g/L, C reactive protein was 18 mg/L with a white cell count of $5.8 \times 10^9/L$. CD4 count was $0.33 \times 10^9/L$ with HIV-1 viral load of <40 copies/mL. HHV-8 was detected on PCR with 1,120 HHV8 DNA copies/mL.

DIFFERENTIAL DIAGNOSIS

The differential for an opalescent effusion in the context of HIV, immunosuppression and a low CD4 count would include bacterial infection, tuberculosis or malignancy. As his CT scan had shown resolution of his lymphadenopathy, Castleman disease (which may also be associated with HHV8) and lymphoma were less likely to be the cause of his effusion.

TREATMENT

A thoracic ultrasound guided 12Fr Seldinger chest drain was inserted and placed on negative suction of -2.5 kPa, and drained milky pink coloured fluid. When the fluid output was less than 150 mL over 24 hours we performed pleurodesis using talc slurry (4g Steritalc (Novatech) +50 mL 0.9% normal saline) via the chest drain (figure 4).

He was reviewed by a dietician and commenced on a medium chain triglyceride (MCT) diet. MCTs are directly absorbed by the intestinal mucosa into the portal system and therefore bypass the lymphatic system thus reducing chyle formation.¹

OUTCOME AND FOLLOW-UP

The patient was followed-up by infectious diseases and medical oncology teams and changed to second

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Figure 1 CT Chest abdomen and pelvis May 2017

line paclitaxel chemotherapy. Follow-up chest radiograph at 1 and 4 months post pleurodesis revealed a small residual basal effusion with otherwise good re-expansion of the lung (*figure 5*). The patient maintained a MCT diet for 4 months and remained under dietician review. HIV-1 viral load remained undetectable and the patient has maintained a good clinical response to treatment.

DISCUSSION

Pleural effusion occurs in up to 60% of patients with KS.² However, chylothorax caused by KS is rarer, with a few case reports published.^{3–12} There are a number of differentials for pleural effusion in HIV patients. Infective causes include bacterial infection, tuberculosis, nocardia, cryptococcus and histoplasmosis.^{13–16} KS-associated HHV-8 Castleman disease is a rare non-cancerous B-cell lymphoproliferative disorder that should also be considered in HIV patients. Non-infectious causes can

Figure 2 Chest Radiograph of the right pleural effusion pre-drain March 2018

Figure 3 CT Chest abdomen and pelvis March 2018

consist of heart failure and hypoalbuminaemia. Malignant causes include non-Hodgkin's lymphoma, primary effusion lymphoma, lung cancer and KS.

Chylothorax is an accumulation of chyle in the pleural space which is often due to disruption of the thoracic duct along its path from the cisterna chyli to where it empties into the venous circulation. The differentials for chylothorax would be traumatic (post surgery) or non-traumatic which commonly includes lymphoma, with rarer causes such as metastatic cancer, sarcoid, Castleman disease and KS.¹⁷

The diagnosis of chylothorax is based on thoracentesis and pleural fluid analysis. Fluid often has a milky, sanguineous or serous appearance. Triglyceride levels over 110mg/dL are usually diagnostic of chylothorax. The role of medical thoracoscopy in diagnosing Kaposi-associated pleural effusion is debatable as KS often affects the visceral pleura and therefore parietal pleura biopsies have a low yield. The main diagnostic aim in performing thoracoscopy would be to exclude other causes especially lymphoma, as HIV patients are at increased risk particularly with low CD4 counts.¹⁸

Figure 4 Chest drain bottle with fluid

Figure 5 Chest Radiograph June 2018

The mechanism behind the development of chylothorax in KS is thought to be due to thoracic duct infiltration or blockage.¹⁷ Our patient's CT scan did not show obvious thoracic duct injury or chyle leakage though CT is not the definitive imaging method to determine this. Lymphography is considered to be the gold standard for investigating suspected chyle leak or thoracic duct injury.¹⁹ Lymphography was considered but it was felt that this investigation would not change the overall clinical management for our patient. We recognise that the lack of histology may be considered a limitation in the diagnostic work-up, however there were no parenchymal lesions or significant mediastinal adenopathy on his recent CT scan to indicate other underlying causes, therefore thoracoscopy was not performed. Given the lack of adenopathy, we suspected the cause of his chylothorax was due to KS causing thoracic duct infiltration secondary to non-compliance with chemotherapy.

Review of the literature shows poor outcomes in patients with KS-associated pleural effusions that are managed solely with repeated thoracocentesis or chest drain without pleurodesis, as HIV treatment and chemotherapy rarely control effusion alone.^{5 9 10 20} Natarajan and Miller describes a case of bilateral chylothorax secondary to relapsed visceral KS which was refractory to repeated aspiration, drainage and talc pleurodesis however with time responded to paclitaxel.²¹ Alexander *et al*, reported a case of KS-associated chylothorax managed initially with percutaneous drainage and talc pleurodesis. The patient re-presented 5 months later with worsening symptoms and bilateral chylous effusions which were resistant to serial drains. They then underwent bilateral decortication with chest tube placement but later died during admission.²² Rubio *et al*, reported a patient with bilateral chylothorax managed with thoracoscopy and 5 g talc poudrage with a survival of 8 months.²³ While Bouladakis *et al*, reported a case of bilateral chylothorax managed with bilateral video-assisted thoracoscopic surgery (VATS), talc pleurodesis and thoracic duct ligation with stable disease at follow-up at 3 years.²⁴ The therapeutic role of thoracoscopy versus percutaneous chest drainage with pleurodesis is unclear given the limited case series.

In addition to pleural drainage, chemotherapy and HAART are important for disease control. Supportive measures include dietary modification with a MCT diet or fat-free diet,

supplementation of essential fatty acids and fat soluble vitamins and protein supplementation if high drain output. Long-chain triglycerides (LCT) are converted into monoglycerides and free fatty acids, which are transported as chylomicrons to the intestinal lymphatic ducts. In contrast MCTs are absorbed directly across the intestinal mucosa into the portal system and are therefore not transported via the lymphatic system. Therefore, omission of LCT from the diet reduces chyle production.²⁵ Compliance with MCT diets is often poor given the common adverse side effects of MCT oil including nausea, vomiting, abdominal pain and poor palatability. There are no randomised controlled trials regarding nutritional management of chylothorax and the duration of nutritional therapy varies in the literature from 1 to 24 weeks.¹ Particularly in patients with HIV, loss of proteins and lymphatic fluid increase the risk of malnutrition and infection, therefore adherence to nutritional therapy is paramount.

In conclusion, we describe a case of chylothorax in HIV related KS managed with percutaneous chest drain and talc pleurodesis, with good outcome at 4 months.

Learning points

- This case is important to medicine as it highlights that thorough investigation of pleural effusions in patients with HIV infection is key as there is a wide diagnostic differential including atypical infection, tuberculosis and malignancy and therefore reaching the correct diagnosis is paramount.
- Although chylothorax and Kaposi's sarcoma (KS) is not common, it is potentially fatal if not recognised and managed appropriately.
- The treatment of chylothorax involves treating the underlying cause, managing the pleural effusion and using supportive measures with a medium chain triglyceride diet.
- The role of thoracoscopy versus percutaneous drainage in the management of KS-associated chylothorax remains unclear.

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