CASE REPORTS

Association of chylothorax and direct pleura involvement in a case of Waldenström’s macroglobulinaemia

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Abstract

An 82-year-old male was hospitalised for dyspnoea, hypoxaemia and general fatigue; a predominant left chylothorax was revealed. Previously, he had been diagnosed with Waldenström’s macroglobulinaemia (WM). Chylothorax complications in patients with WM are rare events and only six such cases have so far been reported. The most common malignant causes of chylothorax are through mediastinal adenopathy. Direct infiltration of the pleura by tumour cells is the most likely cause with this patient, and for this reason, we believe that this case is an instructive one. Chemotherapy induced rapid and persistent improvement after 10-month follow-up.

Keywords: chylothorax, pleura involvement, Waldenström’s macroglobulinaemia, lymphoplasmacytic cells, older people

Case report

An 82-year-old man experienced weight loss, asthenia and bilateral pleural effusion lasting 18 month. Few months earlier, a monoclonal IgM kappa (mIgM) gammopathy in blood serum was discovered, with moderate lymphoplasmacytoid infiltration of the bone marrow and a diagnosis of Waldenström’s macroglobulinaemia (WM) was made, no treatment was started. The man was admitted in our service for dyspnoea and hypoxaemia with a large left pleural effusion. The pleural fluid analysis results showed a chylothorax with predominant lymphocytic cells. The thoraco-abdominal CT revealed a lymphadenopathy in the left posterior costomediastinal angle associated with bilateral pleural effusion that was predominant on that side. The thoracic duct was on the right side.

We retained a hypothesis of progressive WM and corticosteroids were started, rapid improvement followed.

Fifteen days later a positron-emission tomography/computed tomography (PET/CT) revealed one moderate hypermetabolic uptake in the left pleura (SUV max 2.1) and another in the left posterior mediastinum (SUV max 2.3) corresponding to a mediastinal lymph node.

Given the clinical improvement, and the absence of significant tumour syndrome, it was decided to not begin chemotherapy.

Three months later readmission was necessary due to respiratory failure with recurrent large left pleural effusion that coincided with a drop of prednisolone to 10 mg/day. The thoracentesis showed a chylothorax with the presence of lymphoplasmacytic cells and mIgM kappa (Figure 1). This suggested pleural localisations of WM. Chemotherapy was then started with rituximab, cyclophosphamide orally and dexamethasone, and respiratory improvement soon followed.

After six cycles of chemotherapy, PET/CT confirmed the improvement with stability of the posterior inferior right pleura uptake (SUV max = 1.9), and regression of the left posterior mediastinal uptake. Additionally, the left pleural effusion decreased significantly, as did the monoclonal IgM. Rituximab was continued as maintenance therapy, which resulted in persistent clinical and biological improvement after 10-month follow-up.
WM is a rare disease (1–2% of haematological neoplasms), with an estimated incidence of 2 and 4.2 cases per million per year in women and in men, respectively [1], with a median onset age of ∼65 years [2]. WM is a B-cell non-Hodgkin lymphoma called lymphoplasmacytoid lymphoma in the WHO classification. It is defined by lymphoplasmacytic cells infiltration in the bone marrow with production of a monoclonal immunoglobulin M in serum. Common manifestations include general symptoms, tumoural syndrome and cytopenia. Pulmonary involvement represents only 3–5% of cases (parenchymal opacities such as reticulonodular infiltration, pulmonary masses or pleural effusion usually unilateral) [3]. Serous or serosanguineous effusions are the most commons [4].

Chylothorax conventionally correspond to a milky fluid in the pleural space. The chyle flows through the thoracic duct which travels upwards along the right side of the anterior thoracic vertebral surfaces and around the fifth thoracic vertebra crosses to the left side [5]. In non-traumatic aetiologies, malignancy is the most common cause, usually due to obstruction of the thoracic duct by mediastinal adenopathy (lymphoma in 70% of cases) [6].

Chylothorax complications in patients with WM are rare events and only six such cases have so far been reported [4, 7–10]. The most common manner in which a malignancy causes chylothorax is through mediastinal adenopathy [6]. However, in our case the adenopathy was below the fifth thoracic vertebra at the level of T11, which suggested another mechanism. Infiltrated lymphoplasmacytic cells can induce dilatation and proliferation of the lymph vessels, which drain the pleura, resulting in a chylothorax. In our case, we suspect that this is the most likely cause due to the presence of lymphoplasmacytic cells and the pleural proteins electrophoresis [5,7].

The direct pleural infiltration in WM associated with a chylothorax is extremely rare. Pleural proteins electrophoresis, cytologic analysis of the pleural fluid and a TEP Scan can help for diagnosis when pleural biopsies cannot be performed.

**Key points**

- The direct pleural infiltration in WM associated with a chylothorax is extremely rare.
- Chemotherapy induced rapid and persistent improvement.
- Direct infiltration of the pleura by tumour cells.

**Conflicts of interest**

None declared.
Voltage-gated potassium channel antibody-associated limbic encephalitis

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Abstract

We are emphasising the importance of considering a rare diagnosis, voltage-gated Potassium channel antibody-associated limbic encephalitis, in an 80-year-old gentleman who presented with memory impairment, seizure and hyponatraemia. He was found to have high titre of voltage-gated potassium channel antibodies in his serum. He was given high-dose steroids and he responded biochemically and clinically with marked improvement in symptomatology.

Keywords: encephalitis, limbic encephalitis, VGKC antibody-associated encephalitis, older people

Case report

An 80-year-old gentleman, with a history of dual chamber pacemaker inserted in 2011 for sinus arrest, presented with falls. He was commenced recently on carbamazepine for generalised tonic clonic seizures.

His general physical and systemic examinations were unremarkable; his abbreviated mental score was 10/10. His blood tests showed sodium of 121. Other baseline blood tests were normal. 24-h electrocardiogram showed functioning pacemaker. CT head showed moderate global atrophy. After extensive investigations, low sodium was thought to be due to antiepileptic.

The neurology team diagnosed him as immune-mediated encephalitis based on increasing falls, short-term memory loss (reported by his wife and evening ward staff), seizures and low sodium.