An unusual cause of pleural effusion

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Systemic amyloidosis is uncommon and can affect different organs in different patients. Pleural effusion and pericardial effusion due to systemic amyloidosis are rare. To date, only a few reports have been reported in the literature.

We report an interesting case of systemic amyloidosis causing heavy proteinuria, pleural effusion and pericardial effusion.

Case report

A previously well 72-year-old lady presented with a 10-day history of worsening dyspnoea, chest tightness and palpitation.

She was afebrile on presentation. Heart rate was 130 beats per minute and the ECG showed atrial fibrillation. Her respiratory rate was 30 breaths per minute. Heart sounds were dual with no added sounds.

Chest examination revealed dull percussion notes up to mid zones with absent breath sounds consistent with bilateral pleural effusions. She was clinically in right heart failure with raised jugular venous pressure and significant peripheral oedema.

A chest X-ray showed newly developed large bilateral pleural effusions and significantly increased cardiothoracic ratio comparing to her previous chest X-rays.

Serum albumin was low at 21 g/L.

Echocardiogram showed large pericardial effusion, maximum dimension 3.6 cm anterolateral to left ventricle. There was evidence of diastolic compromise of right ventricle. Her heart rate and symptom of dyspnoea improved following pericardiocentesis.

The pleural fluid was drained but quickly re-accumulated. The fluid was transudate according to Light’s criteria: Pleural LDH 126 U/L (serum LDH 306 U/L), protein 24 g/L (serum protein 63 g/L). Urine sample showed heavy proteinuria (1.5 g/L [0–0.15 g/L]).

Renal biopsy showed deposition of fibrillary material measuring 9 to 13 nanometers in thickness that were eosinophilic in nature and stained positive with Congo red consistent with amyloidosis (Figure 1).

Supplementary serology showed IgG lambda band (5.6 g/L) and high lambda light chains levels. The diagnosis was AL amyloidosis and she was promptly started on chemotherapy (bortezomib [Valcade], cyclophosphamide and dexamethasone).

She responded well to the treatment with no further hospital admission. Her serum protein electrophoresis normalised. No recurrence of pericardial effusion since commencement of chemotherapy. She had had no recurrence of pleural effusion in the first 8 months following chemotherapy. She developed small pleural effusion subsequently but responded to oral frusemide.
Discussion

Amyloidosis is a systemic illness with extracellular deposition of protein fibrils of low molecular weight, insoluble and β pleated. Two commonest forms are AL (light chain) and AA (reactive) or secondary amyloidosis.

AL amyloidosis is the most common and severe form. It is a plasma cell dyscrasia with a detectable monoclonal immunoglobulin in the serum or monoclonal light chains in the urine. This could be due to any haematological disorder including monoclonal gammopathy of unknown significance, Waldenstrom’s macroglobulinemia and multiple myeloma.

The most commonly affected organs are kidneys, lungs and heart. It commonly presents with heavy proteinuria, nephrotic syndrome, heart failure, pleura effusions, oedema and organomegaly.

Hypoalbuminaemia and nephrotic range proteinuria are traditionally believed to be contributing to formation of pleural effusion. A retrospective study of 636 AL patients, published by Berk’s Editorial in 2003, found nephrotic range proteinuria and hypoalbuminaemia in combination with restrictive AL cardiomyopathy does not induce pleural effusions.

It is believed that pleural amyloid infiltration plays a central role in persistent pleural effusions. The amyloid impairs pleural lymphatic drainage system and probably promotes pleural fluid secretion.

Biopsy is paramount for diagnosis and can be obtained from affected organs, such as kidney and commonly abdominal fat pad aspirate for the ease of access and the high sensitivity and specificity.

Electron microscopy and post-staining with Congo red show green birefringence under polarised light. Further characterisation of biopsies is vital to distinguish...
between AL and AA amyloidosis using immunohistochemical staining (looking for Kappa or lambda suggesting AL variant).

The management of amyloidosis is mainly divided into haematopoietic cell transplantation or chemotherapy. A chemotherapy approach for a patient unfit for transplant, using mephalan and dexamethasone, is generally recommended although in recent years changes towards other regimens have been implemented including the use of bortezomib, cyclophosphamide, lenalidomide and thalidomide.5,6

Cyclophosphamide-bortezomib-dexamethasone (CyBorD) produces a rapid and complete haematological response in most AL amyloidosis patients but the use of this regimen warrants further trials.7

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