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Case Report

An Interesting Case of Amyloidosis of the Lymph Node: A Case Report and Review of the Literature

Nelum D G Gamage [1], Maysoon Farroha*, Sabrena Sulaiman[1]

1. *Department of Cellular Pathology, Royal Oldham Hospital, United Kingdom.*

*__Correspondence to:__ Maysoon Farroha. Department of Cellular Pathology, Royal Oldham Hospital, Rochdale Rd, Oldham, OL1 2JH, United Kingdom.

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Abstract

Amyloidosis encompasses a group of diseases characterized by abnormal extracellular deposition of amyloid proteins, which may cause significant organ dysfunction. Light chain (AL) amyloidosis is the most common systemic form of the disease. Here, we present the case of a 74-year-old woman with a rare presentation of lymph node amyloidosis, initially misjudgment as metastatic cancer due to her history of breast carcinoma. The final diagnosis was established through endobronchial ultrasound-guided biopsy (EBUS) and confirmed with histological analysis. She responded well to a combination of immunotherapy and chemotherapy.

Keywords: *Amyloidosis; Lymph node; AL Amyloidosis; Congo red staining; Lymphadenopathy.*

Introduction

Systemic AL amyloidosis rarely manifests as lymphadenopathy, making it an uncommon presentation of this potentially fatal condition (1,2,3). Diagnosis requires a high degree of suspicion, particularly in patients with vague systemic symptoms and a history of malignancy. Here, we describe a case in which the diagnosis was established through EBUS-guided biopsy, emphasizing the importance of tissue confirmation with Congo red staining and immunohistochemistry.

Case Report

A 74-year-old woman was referred to an upper gastrointestinal (GI) clinic with complaints of reduced appetite and an unpleasant taste in her mouth. Her medical history included a right mastectomy and axillary lymph node clearance for breast carcinoma.

A computed tomography (CT) scan of the thorax, abdomen, and pelvis revealed widespread lymphadenopathy and a left adrenal lesion. Given her oncologic history, she was referred to the breast multidisciplinary team (MDT) under the suspicion of metastatic disease. An axillary lymph node biopsy initially indicated reactive changes but no definitive pathology.

To further investigate, an endobronchial ultrasound-guided biopsy (EBUS) was performed. Histological analysis demonstrated amyloid deposits, confirmed by Congo red staining with apple green birefringence under polarized light for amyloid. A subsequent lymph node biopsy ruled out Waldenström's macroglobulinemia. The patient was ultimately diagnosed with systemic AL amyloidosis (Mayo Revised Stage 3) and started on Daratumumab-based chemotherapy in combination with cyclophosphamide, bortezomib, and dexamethasone (VCD). She was followed up at Haematology day case ward.

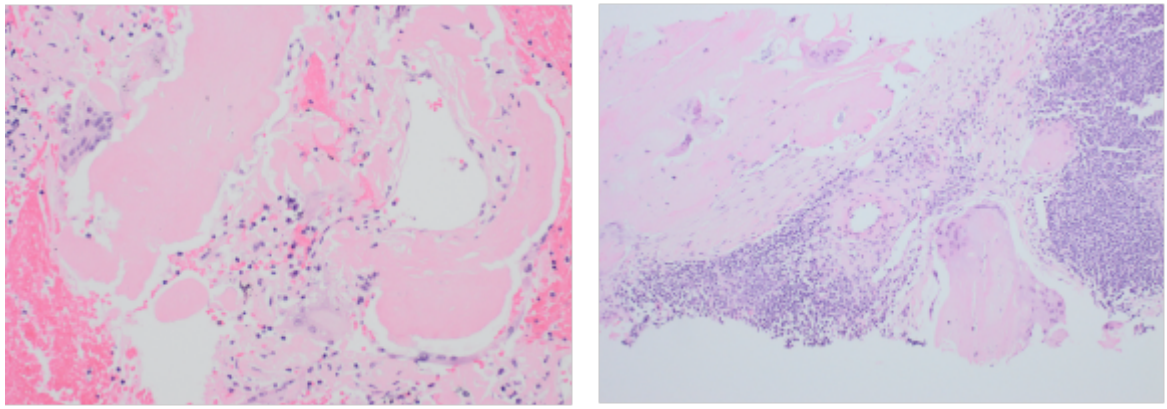

Fig. 1 and 2 H&E sections of EBUS and Lymph node showing extracellular amorphous eosinophilic hyaline deposits with scattered multinucleated giant cells and lymphoid tissue

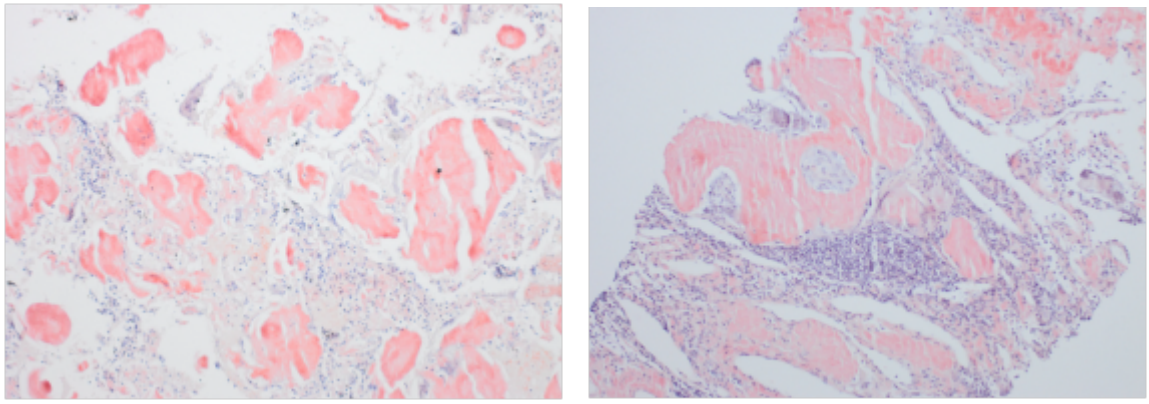

Fig. 3 and 4 Congo red stain of EBUS and Lymph node showing pinkish amyloid deposits.

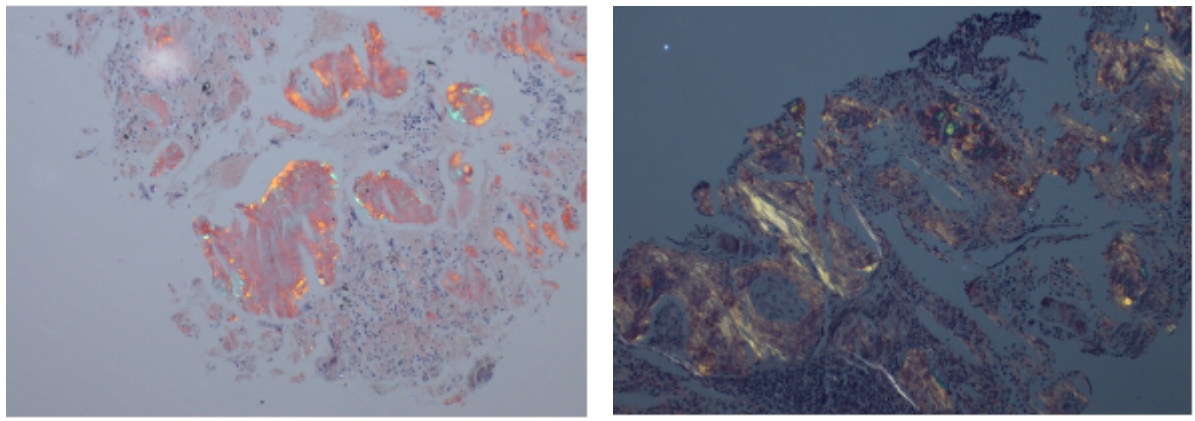

Fig. 5 and 6 Congo red stain of EBUS and Lymph node showing apple green birefringence under polarized light

Discussion

Amyloidosis comprises a diverse group of disorders characterized by extracellular deposition of non-branching amyloid fibrils, which adopt a beta-pleated sheet configuration (3). This configuration is responsible for the pathognomonic apple-green birefringence under polarized light observed with Congo red staining (4, 5). Tissue biopsy remains the gold standard for diagnosis.

Lymphadenopathy is an uncommon manifestation of amyloidosis, most frequently associated with the AL subtype (6). Its presentation often mimics other conditions such as reactive lymphadenopathy or metastatic cancer, necessitating a thorough histopathological workup for accurate diagnosis.

The patient's history of breast carcinoma complicated the clinical picture, initially leading to a presumed diagnosis of metastatic disease. However, the diagnostic use of lymph node biopsy was pivotal in identifying systemic AL amyloidosis, highlighting its utility in cases of unexplained lymphadenopathy.

Conclusion

Lymph node involvement in amyloidosis is rare but clinically significant. This case underscores the importance of considering amyloidosis as a differential diagnosis in patients presenting with unexplained lymphadenopathy, especially when accompanied by other systemic illnesses. Early and accurate diagnosis, achieved through histological examination of tissue biopsy (which is the gold standard) with Congo red staining, is crucial for initiating appropriate therapy and improving patient outcomes.

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