# EXACERBATION OF EROSIVE LICHEN PLANUS OR PARANEOPLASTIC TUMOUR?

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## INTRODUCTION

Paraneoplastic pemphigus is a fatal autoimmune blistering disorder which can be challenging to diagnose due to its rarity and polymorphic appearance. The majority of cases are due to an underlying B-cell lymphoproliferative malignancy such as Castleman disease. This case highlights the need to exclude paraneoplastic pemphigus in patients with oral lichen planus that is refractory to treatment, or if there is a rapid and significant deterioration in the clinical picture. Referral to an oral medicine department for further specialist investigation is warranted, and management of such cases requires a multi-disciplinary approach.

## CASE REPORT

A female in her twenties presented to the Emergency Department with severe widespread painful oral ulceration and atrophic lesions. Other complaints included: oral swelling; oral pain; bleeding gums; sore throat; sleep disturbance; reduced oral intake and a 2 week history of a productive, chesty cough with shortness of breath. She denied nausea, vomiting, fever, night sweats or weight loss. She was previously diagnosed with erosive lichen planus which was refractory to treatment and under regular review. She was otherwise fit and well, a non-smoker and rarely consumed alcohol.

On presentation, she was tachycardiac at 115 heartbeats/minute, but remaining vital signs were normal. Haematological assessment revealed raised C-reactive protein at 17mg/L. A chest radiograph revealed a well circumscribed lesion in the right base of lung. Computed tomography thorax with contrast and magnetic resonance imaging (MRI) of the thorax demonstrated a well defined 5.5 cm soft tissue mass in the right lower lobe of the lung.

An upper left lip perilesional biopsy revealed histological features typical of lichenoid inflammatory infiltration with basal cell damage, and intraepithelial clefting above the level of the basement membrane. Direct immunofluorescence revealed intercellular deposits of C3 along with IgG within the epidermis. She underwent a bilobectomy of the middle and inferior lobe of the right lung which demonstrated features representative of unicentric hyaline-vascular type of Castleman's Disease.

The final diagnosis was paraneoplastic pemphigus with underlying unicentric Castleman disease.

## CLINICAL AND RADIOGRAPHIC IMAGES

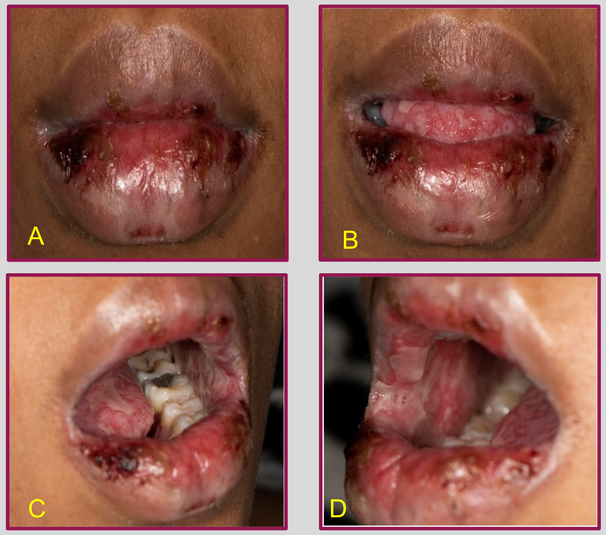


Figure 1a-d: Frontal (1a, b) and lateral views (c, d) of oral lesions.

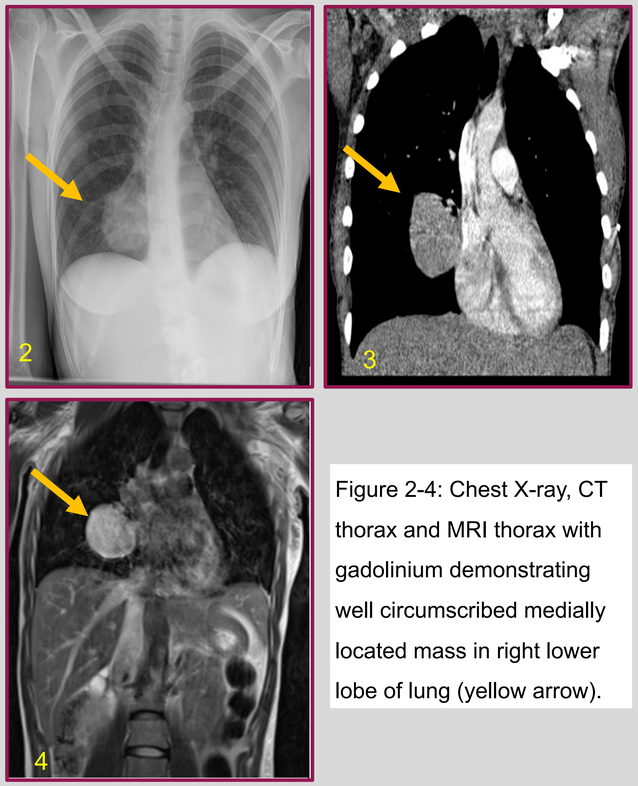


Figure 2-4: Chest X-ray, CT thorax and MRI thorax with gadolinium demonstrating well circumscribed medially located mass in right lower lobe of lung (yellow arrow).

## REFERENCES

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## DISCUSSION

Paraneoplastic pemphigus (PNP) is a rare and often lethal form of pemphigus, defined by the presence of muco-cutaneous lesions in an individual with an underlying neoplasm[1]. It was initially described in 1990 by Anhalt et al in five patients with lymphoproliferative disorders[2]. Autoantibodies implicated in the pathogenesis of PNP include: desmoglein 1 and 3; alpha-2-macroglublin-like-1 and the plakin family of proteins (envoplakin, periplakin, BP230, desmoplakins I and II and plectin). The majority of cases are due to an underlying B-cell lymphoproliferative disorders such as Castleman disease (CD), or malignancies such as non-Hodgkin lymphomas and chronic myeloid leukaemia[1]. The prevalence of PNP in CD patients is 18% [3]. Subsequently, management of PNP focuses on detection and prompt treatment of the underlying tumour. Most deaths are due to progression of the malignancy, infections and bronchiolitis obliterans which results in respiratory failure[1].

CD, also known as giant lymph node hyperplasia and angiofollicular lymphoid hyperplasia, was first described in the literature by Castleman et al in 1956 [4]. It encompasses a rare group of heterogeneous conditions characterised by proliferation of lymphoid tissue in affected lymph nodes. Although CD itself is not a malignancy, patients are at increased risk of developing certain malignancies such as lymphomas and follicular dendritic cell sarcoma. CD is typically categorised into two subtypes: unicentric or multicentric[3]. There are three histological variants of CD: hyaline vascular, plasma cell and a mixed variant. The hyaline vascular variant, commonly associated with unicentric CD, is characterised by proliferating lymphoid follicular tissue composed of cells of varying maturity surrounding a central hyalinised vessel. The plasma variant, commonly seen in multicentric CD, is characterised by sheets of mature plasma cells embedded in interfollicular tissues surrounding a germinal centre. The mixed variant demonstrates a combination of the above histological features, and is commonly associated with unicentric CD or idiopathic multicentric CD[5].

The undetermined aetiology of CD, the rarity of the disease along with lack of evidence based established treatment guidelines makes management of CD challenging. UCD has good prognosis, and surgical resection of enlarged lymph node or lymph node region is the treatment of choice and mostly curative[3].