19. A CASE OF LARGE GRANULAR LYMPHOCYTOSIS (LGL) WITH NEUTROPAENIC SEPSIS ASSOCIATED WITH POSITIVE ANA, ANTI-RO, ANTI-RIBOSOMAL P AND ANTI-NEUTROPHIL ANTIBODIES RESPONDING TO METHOTREXATE

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Introduction: A very challenging case where the diagnosis became evident after extensive rheumatological, haematological, and general medical (sepsis and malignancy) work up.

Case description: 84 year old gentleman with 1 month history of generally unwell, lethargy, reduced appetite but no previous or current history of CTD symptoms or signs. Basic septic and neoplasia screening came back negative including CT TAP and blood culture. ANA was positive with 1:640 speckled pattern, anti-Ro and Ribosomal P antibodies positive, raised IgG and CRP. His WBC 1.3, Neutrophils 0.45 and Hb was 98. Blood film demonstrated Large Granular Lymphocytosis (LGL). After literature search, LGL is found to be associated with haematological malignancies, RA and Sjogren's syndrome. We checked his Anti-Neutrophilic antibodies which came back positive and haematology did bone marrow biopsy which confirmed peripheral consumption and no evidence of haematological pathology all confirming a connective tissue disease as the cause of LGL on blood film. We initially commenced him on high dose Prednisolone which was ineffective, but Methotrexate improved the neutrophil count to normal.

Discussion: Large granular lymphocyte (LGL) comprise 5 percent of the population of peripheral blood mononuclear cells, are larger than most circulating lymphocytes, and have characteristic azurophilic granules containing acid hydrolases (picture 1). They may be either T cells (T-LGL), the more common type, or natural killer cells (NK-LGL). Most common cause of LGL is haematological malignancies. Up to one-third of patients with T-LGL also have rheumatoid arthritis (RA). LGL can also occur in association with Sjögren's syndrome in the absence of RA and with other autoimmune disorders, including inflammatory bowel disease, systemic lupus, and autoimmune thyroid disease.

Key learning points: This is a case where the presence of ANA, anti-Ro, anti-Ribosomal P and anti-Netrophil antibodies presented with clinically significant neutropaenia rather than typical symptoms of a connective tissue disease. A holistic approach is required to come to the diagnosis of such challenging patients.