ARA Abstract – Grand Rounds

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69yo Indian Man presented to a peripheral hospital in Queensland, 2 hours north of Brisbane whilst on holiday visiting his children in Australia. Presented with a two week history of back & leg pain, facial rash and weakness. This is on a background of Renal Cell Carcinoma with subsequent left partial nephrectomy in 2011, benign prostatic hypertrophy with a transurethral resection of the prostate in 2011 and distant history of treated bovine tuberculosis completed in India. Prior to this presentation he was not taking any regular medications.

On presentation initial investigations demonstrated normal renal function and full blood count. CK 6380, CRP 22, ESR 30. ANA >2560 speckled, ENA / dsDNA neg. Antiphospholipid antibodies neg, RF/CCP negative, ANCA neg. C3/4 normal. MRI right upper limb demonstrated pathologic enhancement of muscles surrounding the elbow joint. Extensive subcutaneous oedema. Muscle biopsy of the right forearm consistent with dermatomyositis.

Initial treatment with 80mg PO prednisolone and appropriate gastric/PCP/bone prophylaxis. Upon discharge was mobilising with 4WW requiring some assistance with ADL's provided by his family.

Re-presented to the 2 weeks later to a Brisbane Tertiary Hospital with worsening weakness, now bed bound. Unable to swallow and profoundly oedematous. On examination at this time oral secretions pooling with significant mucositis, Peak Flow 225mL. Profound weakness of upper (UL) and lower limbs (LL). Gross oedema UL/LL/Abdomen. Petechiae noted on tongue and in axillae. Healing heliotrope rash noted. At this time was commenced on IV Methylprednisolone 750mg x 3 and IVIG. At this time myositis ENA panel positive for NXP2 antibodies.

Subsequently developed haemolytic anaemia (attributed to IVIG - nadir Haemoglobin 70) , thrombocytopenia (nadir 38) and sepsis with normal renal function throughout. Admission to intensive care unit requiring ventilation for poor forced vital capacity in addition to inotropic and blood product support. He underwent several laparotomies for perforated bowel and received broad spectrum antibiotics for intra abdominal sepsis.

Day 19 of admission histology results available from laparotomy revealing duodenal perforation secondary to neutrophilic vasculitis. This was associated with focal necrosis of the muscularis propria with neovascularisation in the lamina propria focally indicative of subacute disease. Also fibrin thrombi were not demonstrated instead; immunohistochemistry for platelet antibody (CD42B) was positive. Concurrent peripheral film and BMAT demonstrates normal platelet production and morphology.

So this patient has a novel diagnosis of biopsy proven dermatomyositis associated with NXP2 antibodies with intestinal vasculitis with demonstrated platelet consumption in the small bowel leading to perforation.