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| **A retrospective review of the management of MDA5 associated ILD** |
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| **Introduction/Aim:**  Anti-melanoma differentiation-associated gene 5 (MDA5) antibodies are associated with rapidly progressive interstitial lung disease (RP-ILD). Our aim was to evaluate the management of MDA5 associated ILD at our centre.  **Methods:**  All patients at Fiona Stanley Hospital who tested positive for MDA5 antibodies from January 2017 to July 2023 were identified. Records were reviewed for demographics, clinical presentation, presence of ILD, pulmonary function tests, radiologic pattern and survival. Patients with ILD were categorised as having MDA5 associated ILD (dermatomyositis, myositis or RP-ILD) or alternative cause for ILD. MDA5 antibodies were assessed as weak, moderate or strong.  **Results:** Of 33 patients with MDA5 antibodies, 8 had MDA5 antibody associated ILD. Of these patients, 5 had RP-ILD, all of whom died. Treatments are summarised below.   |  |  |  |  |  |  | | --- | --- | --- | --- | --- | --- | | Phenotype | First line | Second line | Third line | Fourth line | Outcome and comments | | CADM with RP-ILD | Methylprednisolone Mycophenolate IVIg  Rituximab | CYC | CsA |  | Died 15 days after treatment initiated.  Results returned after treatment commenced. | | CADM with RP-ILD | Methylprednisolone Mycophenolate CsA IVIg | Tofacitinib | IVIg Rituximab | PLEX | Died 48 days after treatment initiated.  CYC avoided for fertility. | | CADM, exacerbation of ILD | Prednisolone Mycophenolate IVIg Hydroxychloroquine | Methylprednisolone Tacrolimus Rituximab |  |  | Improved but did not recover to baseline.  Clinically stable 12 months later. | | RP-ILD | Methylprednisolone CYC |  |  |  | Died 12 days after treatment initiated. Results returned after death. | | RP-ILD | Methylprednisolone |  |  |  | Died 14 days after treatment initiated. VV-ECMO used. Results returned after death. | | RP-ILD | Prednisolone |  |  |  | Died 24 days after treatment initiated. Co-morbidities and adherence impacted treatment. | | Myositis with progressive ILD | Prednisolone Mycophenolate | Rituximab |  |  | Slowly progressive ILD over 5 years. Multiple myositis antibodies. | | Myositis with mild ILD | Prednisolone IVIg Rituximab |  |  |  | Stable, mild ILD at 12 months of follow-up. | | CYC=cyclophosphamide, CsA=cyclosporin, IVIg= intravenous immunoglobulin, PLEX= plasma exchange, VV-ECMO=Veno-venous extracorporeal membrane oxygenation. | | | | | |   **Conclusion:**  Management of MDA5 antibody associated ILD at our centre is variable. This is in part due to the spectrum of presentations, time to definitive diagnosis, access to and toxicities of treatments. Mortality rate in our centre is high, even when extensive lines of treatment are used.  **Grant Support:**  None. |