Gene expression profiles may distinguish patients that can benefit from imatinib. Also, Notch signalling could be exploited to increase imatinib uptake into fibroblasts, thereby increasing efficacy.

References:

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AB0627
PROGNOSIS AND MORTALITY OF DERMATOMYOSITIS AND POLYMYOSITIS PATIENTS WITH MALIGNANCY

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Background: Previous studies indicate that cancers in DM/PM patients are associated with increased mortality. Hence, identifying predictors of malignancy in PM and DM is crucial. However, few large series studies have reported prognostic and predictive factors of malignancy in patients with PM and DM. Moreover, in recent years, several published studies also allow us to better understand the clinical characteristics of malignancy in PM and in DM.

Objectives: To analyze the mortality and identify the major independent risk factors for death in patients with dermatomyositis/polymyositis (DM/PM) complicated with malignant tumors.

Methods: The clinical data of all patients with DM/PM in Peking University First Hospital from January 2007 to Jan 2019 were retrospectively reviewed. All patients were followed up to confirm whether they had malignant tumors. According to the statistics of the National Bureau of Statistics of China, the standard mortality (SMR) and life lost years (YLL) of patients with DM/PM were combined with malignant tumors. The Kaplan-Meier method was used to analyze the 10-year survival of DM/PM patients with malignant tumors. Cox multivariate regression was used to predict independent risk factors for death in DM/PM patients with malignant tumors.

Results: A total of 334 patients with dermatomyositis and 69 patients with polymyositis were enrolled in the study. The mean age of onset was 50.5 ± 14.8 years and 48.9 ± 16.1 years, with a median follow-up of 40.6 (11.6-77.6) months. Among them, 320 patients were successfully followed up, including 89 patients with death, 46 DM/PM with malignant tumors (38 with dermatomyositis and 8 with polymyositis). The average age of onset of DM/PM patients with malignant tumors was 55.4 ± 15.1 years and 59.5 ± 4.7 years, respectively, of which 17 died. The age-sex adjusted SMR of DM/PM patients without malignant tumors was 9.0 (95% CI 6.8-11.2). The age-sex adjusted SMR of DM/PM with malignant tumors was 12.3 (95% CI 9.0-14.7). The life loss of male patients with dermatomyositis complicated with malignant tumors was 30.1 years, and that of females was 38.6 years; the life loss of male patients with polymyositis was 27.6 years, and that of females was 22.1 years. A 10-year survival analysis showed that DM/PM patients with malignant tumor had significantly worse prognosis than patients without malignancy (p=0.001 Log-rank). The 1-, 5-, and 10-year survival rates of DM/PM patients who did not have malignant tumors were 87.9%, 81.9%, and 78.4%, respectively. DM/PM Patients with malignant tumors 1, 5, and 10 years The survival rates were 73.3%, 56.0%, and 45.7%, respectively. The independent risk factors for death in DM/PM patients with malignant tumors were advanced age (HR=1.11 95% CI 1.02-1.20, p=0.014) and infection (HR=1.707 95% CI 1.66-1.75, p=0.017).

Conclusion: Malignant tumor is a common in patients with DM/PM, and the mortality of DM/PM patients with malignant tumors is high. The independent predictors of mortality for PM/DM patients with malignant tumors were age at disease onset and infection.

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AB0628
EMERGING AUTOANTIBODIES PANEL (MYOSITIS ASSOCIATED AND MYOSITIS SPECIFIC ANTIBODIES) IN INFLAMMATORY MYOPATHIES: THE FREQUENCIES OF AND RELATIONSHIP WITH CLINICAL FEATURES

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Background: The idiopathic inflammatory myopathies (IIM) are characterized by muscle weakness, skin disease and various internal organ involvement and they can overlap with other autoimmune diseases. Recent autoantibody panels improve understanding and management of inflammatory myopathies. Myositis specific autoantibodies including Anti-TIF-1, Anti-NXP2, Anti-MDA5, Anti-SAE1, anti-HMGCGR and anti-cn1A are regarded as key biomarkers aiding the diagnosis of patients. On the other hand, myositis-associated autoantibodies (MAAs) are also found in other autoimmune rheumatic diseases.

Objectives: To investigate the clinical meaning and impact of new myositis autoantibodies panel in real life data.

Methods: A total of 110 subjects (77 female, 33 male) admitted to Hacettepe University Hospitals with the signs and symptoms of IIM were screened by a line immunoblot assay (EUROLINE: Autoimmune Inflammatory Myopathies16 Ag) between 2017 and 2020. Only moderate or strong reactivity results were reported as positive. Demographic, clinical, laboratory, therapeutic data and imaging features were obtained by the retrospective review of medical records. IIM patients were diagnosed by Bohan and Peter’s criteria and classified according to the EULAR/ACR classification criteria for adult and juvenile IIM and their major subgroups.

Results: IIM was diagnosed in 61 patients (42.6% DM/JDM and 57.4% PM) and patients with overlap were in decreasing order, Scleroderma (n=8), RA (n=5), Sjogren (n=4), SLE (n=3), autoimmune hepatitis (n=2).

Myositis-specific autoantibodies (MSAs) were found in 60%, myositis-associated autoantibodies (MAAs) in 41% of inflammatory myositis patients. The most common MSAs were anti-Jo-1 (16.4%) and anti-MDA-5 (13.1%) and the most common MAA was Ro-52 (32.7%) (Table 1). MAAs were more common in patients with polymyositis (54% vs 23% p=0.013). Twenty-one (34.4%) patients (61.0% females) had intestinal lung disease (ILD). Anti-Jo-1 (38.1%) and anti-Ro-52 (52.4%) was the most common MSA and MAA in patients with IIM and ILD.

MSAs were also determined in 10 of the 49 patients who were not diagnosed with IIM. Five patients with Anti-SRP, Anti-Mi-2 alpha, Anti-Jo-1 Anti-PL7, Anti-MDA-5 autoantibodies diagnosed as connective tissue disorder, one of them also had bladder cancer. Three patients with Anti-NXP2, Anti-PM-Scl 75, Anti-Ro-52, Anti-Mi-2 beta, Anti-PL7, Anti-PL12 autoantibodies had ILD. One patient (Anti-Mi-2 alpha, Anti-NXP2 and Anti-Ku autoantibodies positive) had viral myositis and one patient (Anti-Mi-2 alpha, Anti-Ro-52 autoantibodies positive) had inflammatory myoneuropathy.