Original Research Paper

Volume - 12 | Issue - 03 | March - 2022 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Neurology

CLINICAL FEATURES OF SRP MYOSITIS: A SINGLE CENTRE STUDY

Dr. Kamlesh Jagiasi HOD, Neurology.

Dr. Arjun G Shah* *Corresponding Author

ABSTRACT With the advent of sophisticated diagnostic techniques, and the elucidation of disease specific antibodies, it it critical to be able to identify characteristic neurological syndromes at the earliest, since earlier treatment can lead to better outcomes. SRP- Myositis is being increasingly diagnosed. It is a variant of IMNM which is characterised by severe muscle weakness, with biopsy showing a paucity of inflammatory cells. Predominant neck weakness and diminished tendon reflexes in a patient with a subacute to chronic myositis points towards this diagnosis.

KEYWORDS : SRP- Myositis, IMNM, Rituximab

INTRODUCTION

The Idiopathic Inflammatory Myopathies include various entities such as Dermatomyositis, Polymyositis, Immune Mediated Necrotising Myositis and Inclusion Body Myositis. With changing paradigms in the inflammatory myositis, that led to the use of specific anti-bodies to diagnose various myositis, it was seen that approximately 5% of patients diagnosed with Idiopathic Inflammatory Myopathies had antibodies to the signal related peptide (anti SRP-antibody). Muscle biopsies in these patients revealed muscle fibre necrosis and regeneration, prominent endomysial fibrosis, but little or no inflammation. [1], [2].

These patients tend to have severe muscle weakness that is often difficult. To control despite adequate immunosuppression. [3], [4]

Methods:

This study was a prospective observational study, that was carried out in the neurology department of a tertiary care hospital in Mumbai from October 2019. To January 2022. We diagnosed 5 cases of SRP Myositis, based on clinical presentation, Muscle Biospy, Electrodiagnostic findings, Serum CPK levels and presence of anti-SRP antibodies.

Anti-SRP antibodies were evaluated by sending for a Myositis Panel, that included 16 antigens, and was done using the Immunoblot Technique with a sample screening dilution of 1:101.

The patients further underwent cardiac work up (ECG and 2D-Echo), PET Scan, and where appropriate, other ancillary tests.

RESULTS

The mean age of our patients was 35 years with a female : male ratio of 3:2. 4 of 5 patients had presented to us with a subacute to chronic presentation, beyond 6 months. All presented with a limb-girdle pattern of weakness with 3 of five having predominantly lower weakness. The involvement of upper limb weakness, in all cases, was within 6 months of lower limb involvement.

Interestingly, all our patients had prominent neck flexor weakness at presentation. One patient had cranial involvement in the form of facial and bulbar weakness.

All the patients had diminished ankle jerks.

EDx showed myopathic features with the presence of fibrillations, which could help differentiate them from muscular dystrophies.

One patient presented to us in acute respiratory failure, however she was subsequently diagnosed to have COVID-19 illness, and the cardiorespiratory symptoms could be representative of the same. We carried out a detailed cardiac involvement in the patients including ECG and 2D Echo. None of our patients had any cardiac issues during evaluation. None. Had any extra-muscular weakness either.

None of our patients had any extra-muscular features or underlying malignancy.

We treated the patients with methyl-prednisolone in a dose of 1gm over 5 days (Pulse Therapy) followed by oral steroid. Subsequently we initiated therapy with Rituximab. Rituximab was given based on Body Surface Area, in two divided doses 2 weeks apart, with a plan to follow up for maintenance dosing 6 monthly.

At 3months of follow up, none of the patients reported worsening of symptoms, however, neither did any of them show any improvement on Modified Rankin Score.

	Patient-S	Patient-B	Patient-I	Patient-N	Patient H
Age at Onset	26 years	12 years	31 years	46 years	55 years
Sex	Female	Female	Male	Male	Female
Time since onset	8 months	6 months	6 months	10 months	3 months
Upper Limb Power	Yes	Yes	Yes	Yes	Yes
Lower Limb Power	Yes (UL=LL)	Yes (LL>UL)	Yes (LL>UL)	Yes (LL>UL)	Yes (LL=UL)
Bulbar Involvement	Yes	No	No	No	
Neck Weakness	Yes	Yes	Yes	Yes	
Cardiac Involvement	No	No	No	No	
Respiratory Involvement	No	No	No	No	Yes*
Assymetry	No	No	Yes	Yes	No
Extra-muscular Involvement	None	No	No	No	Yes*
Associated Disorders	None	None	None	None	COVID-19 Deep Vein Thrombosis
CPK Levels	530		2568	580	1800
Biopsy Findings	Muscle fibres of varying sizes. Internalisation of nuclei. No inflammation in perimyseal or endomyseal region		Muscle fibres of varying sizes, polygonal in shape with striations and eccentric nuclei. Sparse focal Lymphocytic infiltration	Focal Perifascicular atrophy with scany mononuclear inflammatory infitrate	-

Acute Therapy	Methyl-Prednisolone	Methyl-Prednisolone	Methyl-prednisolone Plasma Exchange	Methyl-Prednisolone	Methyl-Prednisolone IViG
Secondary Immunosuppression	Rituximab	Rituximab	Rituximab Methotrexate	Rituximab	-
Modified Rankin Score at Admission	3	3	3	2	4
Weakness at 3 months after Rituximab	3	3	3	2	Deceased
EMG	Fibrillations with Myopathic Features	Fibrillations with Myopathic Featured	Fibrillations with Myopathic Potentials	Fibrillations with Myopathic Potentials	Fibrillations with Myopathic Potentials
Other labs			ANA Anti Ro +		
Reflexes	Diminished Ankle Jerks	Diminished Ankle Jerks	Diminished Ankle Jerks	Diminished Ankle Jerks	Diminished Knee and Ankle Jerks
Calf	Hypertrophy	Hypertrophy		Hypertrophy	Normal
Clinical Features, Lab Investigations and Therapy					

DISCUSSION

SRP Myopathy can present in a number of ways, ranging from acute, subacute to even chronic. [5] Although rapid clinical deterioration is characteristic of the illness, almost 19% of these patients can present with a more insidious from of the disease. These patients tend to be younger and have more severe disease, perhaps owing to the delay in diagnosis and initiation of therapy. [1], [6].

On histology they show necrotising Myopathy with a paucity of inflammatory cells. The cornerstone of treatment is immunosuppression. Various agents have been tried. They are poorly responsive to corticosteroids, however, studies indicate that Rituximab might be effective in these patients. Patients might show improved clinical outcomes along with a decrease on CPK and anti-SRP titres. The effect can start within two months and last 12-18 years as per a study by Valiyil et al. [7]

Extra-muscular features such as dermatological and cardiac are rare in SRP myositis. SRP is probably not associated with an underlying malignancy. [8] [9]

CONCLUSIONS

SRP Myopathy is a distinct clinical entity that requires a keen clinical accumen. It might present with a chronic course. It is essential to identify such patterns as they need to be treated early with immunosuppression.

Peculiar features of this disease that we have observed are neck muscle weakness and diminished tendon jerks. At present the best option to offer these patients is Rituximab.

Early initiation of the same may lead to better outcomes. More research is needed to elucidate the precise mechanism of disease and therapy for the same.

REFERENCES:

1. MILLER, T. (2002). MYOPATHY WITH ANTIBODIES TO THE SIGNAL RECOGNITION PARTICLE: CLINICAL AND PATHOLOGICAL FEATURES. *JOURNAL OF NEUROLOGY, NEUROSURGERY & PSYCHIATRY*, *73*(4), 420-428. HTTPS://DOI.ORG/10.1136/JNNP.73.4.420
2. TARGOFF, I., JOHNSON, A., & MILLER, F. (1990). ANTIBODY TO SIGNAL RECOGNITION PARTICLE IN POLYMYOSITIS. *ARTHRITIS & RHEUMATISM*, *33*(9), 1361-1370. HTTPS://DOI.ORG/10.1002/ART.1780330908
3. HENGSTMAN, G., TER LAAK, H., VREE EGBERTS, W., LUNDBERG, I., MOUTSOPOULOS, H., & VENCOVSKY, J. ET AL. (2006). ANTI-SIGNAL RECOGNITION PARTICLE AUTOANTIBODIES: MARKER OF A NECROTISING MYOPATHY. *ANNALS OF THE RHEUMATIC DISEASES*, *65*(12), 1635-1638. HTTPS://DOI.ORG/10.1136/ARD.2006.052191
4. PINAL-FERNANDEZ, I., PARKS, C., WERNER, J., ALBAYDA, J., PAIK, J., & DANOFF, S. ET AL. (2016). LONGITUDINAL COURSE OF DISEASE IN A LARGE COHORT OF MYOSITIS PATIENTS WITH AUTOANTIBODIES RECOGNIZING THE SIGNAL RECOGNITION PARTICLE. *ARTHRITIS CARE & RESEARCH*, *69*(2), 263-270. HTTPS://DOI.ORG/10.1002/ACR.22920
5. DIMITRI, D., ANDRE, C., ROUCOULES, J., HOSSEINI, H., HUMBEL, R., & AUTHIER, F. (2007). MYOPATHY ASSOCIATED WITH ANTI–SIGNAL RECOGNITION PEPTIDE ANTIBODIES: CLINICAL HETEROGENEITY CONTRASTS WITH STEREOTYPED HISTOPATHOLOGY. *MUSCLE & NERVE*, *35*(3), 389-395. HTTPS://DOI.ORG/10.1002/MUS.20693
6. SUZUKI, S., HAYASHI, Y., KUWANA, M., TSUBURAYA, R., SUZUKI, N., & NISHINO, I. (2012). MYOPATHY ASSOCIATED WITH ANTIBODIES TO SIGNAL RECOGNITION PARTICLE. *ARCHIVES OF NEUROLOGY*, *69*(6). HTTPS://DOI.ORG/10.1001/ARCHNEUROL.2011.1728
7. VALIYIL, R., CASCIOLA-ROSEN, L., HONG, G., MAMMEN, A., & CHRISTOPHER-STINE, L. (2010). RITUXIMAB THERAPY FOR MYOPATHY ASSOCIATED WITH ANTI-SIGNAL RECOGNITION PARTICLE ANTIBODIES: A CASE SERIES. *ARTHRITIS CARE & RESEARCH*, *62*(9), 1328-1334. HTTPS://DOI.ORG/10.1002/ACR.20219
8. KAO, A., LACOMIS, D., LUCAS, M., FERTIG, N., & ODDIS, C. (2004). ANTI-SIGNAL RECOGNITION PARTICLE AUTOANTIBODY IN PATIENTS WITH AND PATIENTS WITHOUT IDIOPATHIC INFLAMMATORY MYOPATHY. *ARTHRITIS & RHEUMATISM*, *50*(1), 209-215. HTTPS://DOI.ORG/10.1002/ART.11484
9. Love, L., Leff, R., Fraser, D., Targoff, I., Dalakas, M., Plotz, P., & Miller, F. (1991). A New Approach to the Classification of Idiopathic Inflammatory Myopathy. *Medicine*, *70*(6), 360-374. https://doi.org/10.1097/00005792-199111000-00002define useful homogeneous patient groups. Medicine. 1991;70(6):360–74.