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AOSD Presenting with Myositis: Two Rare Case Reports

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ABSTRACT
Myalgia can be a frequent association with AOSD but myositis is rare. Herein we describe two cases of AOSD presenting with clinical features of myositis which was confirmed with electromyography and laboratory investigations. In both cases treatment was initiated with high dose steroids and methotrexate and improvement was noticed clinically which was also reflected in laboratory parameters.

INTRODUCTION

Adult-onset Still's disease is a rare systemic inflammatory disorder characterized by spiking fevers, salmon coloured rash, arthralgia, myalgia and leucocytosis. It was first described in 1971 by Bywaters in 14 adult patients who were followed up for a mean period of 20 years[1]. In 1992, Yamaguchi et al proposed a classification criterion for AOSD after analyzing 90 Japanese patients which included fever, arthralgia, typical rash and leucocytosis as major and sore throat, lymphadenopathy and/or splenomegaly, liver dysfunction and the absence of rheumatoid factor and anti nuclear antibody as minor criteria, with 5 criteria including 2 majors required for diagnosis[2]. Although myalgia has been reported in at least 84% of the cases, inflammatory myositis, is extremely rare[3].

Case 1: A 32-year-old female patient presented with high grade fever for the past 10 days along with myalgia and arthralgia. On examination she had a salmon coloured pruritic rash on her legs, there was generalized muscle tenderness due to which the power of the muscles could not be appropriately graded. From her history and examination findings, myositis was initially considered. Based on her clinical features and investigations (Table 1, case 1) and according to Yamaguchi criteria, she was diagnosed with AOSD with inflammatory myositis based on EMG. After initiating steroids, NSAIDs and methotrexate, the patient became afebrile and her myalgia improved. Gradually her muscle power improved from 3/5-4+/5 within a span of 2 weeks.

Case 2: A 42-year-old male presented with fever for 25 days along with sore throat, arthralgia and myalgia. On examination he had rashes on his front of chest, back and arms. His vitals were normal except for tachycardia having pulse rate of 120/min and temperature of 101F. There was proximal muscle weakness of both upper and lower limbs as manifested by 4/5 power around both shoulder joint and both hip and knee joints. He was diagnosed as AOSD as per Yamaguchi criteria with myopathy as per finding of EMG. After starting steroid in dosage of 1mg/kg body weight his fever, arthralgia and myalgia subsided, power improved to5/5, TLC came down to 11520 and CRP came down to 25.

RESULTS AND DISCUSSION

In AOSD, myalgia has been found in 68.3-84% of cases[3,4]. Due to associated joint involvement and myalgia, grading of muscle power is usually faulty and as such muscle weakness may be attributed to myalgia rather than myositis[5]. Our patient was found to have symmetrical weakness of proximal limb muscles, elevated skeletal muscle enzymes in serum and EMG patterns suggestive of inflammatory myositis without the characteristic rash of dermatomyositis. Muscle biopsy could not be done and hence the patient can be considered as a probable case of polymyositis[6]. Our literature review revealed handful of cases with overlapping AOSD and myositis, highlighting the rarity of the association. Of the 34 cases reported in literature (Table 2), at least 33 cases (97%) had myalgia and 20 (58%) had proximal muscle weakness. 9 cases showed presence of some changes on EMG and of 21 patients who underwent muscle biopsy, 12 showed no changes (57%) and 9 cases (43%) showed some degree of pathological changes, either in the form of mononuclear infiltrates or atrophic fibres with necrosis. MRI evidence supported the findings in one case reported by Umeda[7] in which there was presence of inflammatory changes in the muscles of the thigh. In a rare case, as reported by Yanai[8], the subject developed NSAIDs induced myositis, as evidenced by elevated enzyme markers on initiation of NSAIDs, which reduced after corticosteroids therapy. Most of these cases responded well to steroids and NSAIDs and 2 cases, on addition of methotrexate showed good response. In one of our cases, the patient initially didn't respond to prednisolone (60mg at 1mg/kg) dose and NSAIDs. Therefore, dose of steroids was increased to 80mg and methotrexate was added. The patient became afebrile within 3 days.

CONCLUSIONS

Our cases highlight the rarity of association between myositis and AOSD and the need to consider myositis as a rare presentation of AOSD. Presence of myositis, if not drug induced, may respond poorly to only steroids and may need addition of DMARDs. Due to the need for prolonged therapy with steroids and risk of side effects, the use of DMARDs and steroid sparing agents may be considered while planning management.

Table 1: Cases

Investigation	Case 1	Case 2	Normal Value
Hemoglobin (g/dL)	8.3	11.5	11-17
Tlc (per mm3)	21370/mm3	24470	4000-11000
Tpc (lacs/mm3)	2.04	2.9	1.5-4.5
Esr (mm in 1st hr)	105	41	<30
CRP (mg/L)	65.8	135	<6
LDH (U/L)	1608	400	313-618
Ferritin (ng/mL)	>2000	>2000	13-400
CPK (U/L)	32	21	39-238
SGOT (IU/L)	124	142	<35
SGPT (IU/L)	87	64	<45
ALP (IU/L)	249	267	<369
Procalcitonin (ng/mL)	0.175	0.24	<0.5
Thyroid function test	normal	Normal	
ANA	negative	negative	
p-ANCA	negative	NA	
c-ANCA	negative	NA	
Bone marrow aspiration	Accelerated myelopoiesis	Normal	
Ultrasound(abdomen and pelvis)	Mild hepatomegaly	Hepatosplenomegaly	
Hrct(thorax)	normal	Fibrotic band in b/l lowerlobe	
Echocardiography	normal	Hypokinesia of LV(myocarditis)	
Blood culture	sterile	Sterile	
Urine culture	sterile	Sterile	
Electromyography	Myopathic pattern with insertional activity	Myopathic pattern insertional activity	

Table 2: Highlight of Diseases

SL no Ref	Age	Sex	Diagnosis of AOSD	markers	EMG	BIOPSY	MRI	Treatment
1Umeda[7]	71y	F	Fever, arthralgia, skin eruption Throat pain lymph node swelling, hepatosplenomegaly,myalgia	raised		-CD68 positive cell infiltration (inflammatory myopathy with abundant macrophages thigh and fascia	massive inflammatory changes in	Steroid
2Moreno-Alavarez[5]	41Y	F	Fever, rashes, arthralgia, myalgia	raised	myogenic involvement	Type2 fibre atrophy sparse focal endometrial and perivascular inflammatory and macrophage invasion		Steroid, NSAIDS
cell infiltrates, necrotic fibre								
3Yanai[8]	43y	M	Fever, myalgia, arthralgia, salmon rashes, muscle weakness	raised		Active myogenic changes		Steroid, NSAIDS
4Samuels[9]	42y	M	Fever, myalgia, arhralgia, proximal muscle weakness, muscle tenderness	elevated	Myogenic changes			Methotrexate, Steroids
5Scwarzberg[10]	42	M	Fever,myalgia,muscle tenderness	elevated	Myogenic changes	Perivascular accumulation of round cells		steroid
6Bujak[13]	-	-	-	normal	-			NSAIDs,
7Pouchot[3]	17 cases	-	Fever, muscle atrophy, proximal muscle weakness	Elevated in 17 cases	3-normal 1- non inflammatory myopathy	12- normal 3-mononuclear cell infiltrates 2- atrophic fibres, single fibre necrosis		Steroids, NSAIDs, DMARDs
8Defuentes[12]	43	- M	Fever, rash, myalgia, proximal muscle weakness	elevated	Myogenic changes	Necrotic myocytes, mononuclear infiltrates		Steroids, aspirin, IVIg

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