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CHURG STRAUSS SYNDROME WITH NEUROPATHY: A CASE REPORT

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ABSTRACT

A 24 years old man with known history of asthma came with acute presentation of ascending weakness for four days following flu like symptoms and worsening of asthma. His complete blood counts showed elevated eosinophil count. Nerve conduction showed neuropathy. He was diagnosed as Churg Strauss syndrome due to presence of asthma, eosinophilia, neuropathy and pulmonary infiltrates. Churg Strauss is a systemic eosinophilic vasculitis involving multiple organ systems that may present with neuropathy, as in this case.

Key Words: Churg Strauss Syndrome, Vasculitic neuropathy, Mononeuritis multiplex, Systemic vasculitides, Eosinophilia

INTRODUCTION:

Churg Strauss syndrome is an eosinophilic vasculitis of small to medium sized blood vessels with autoimmune etiology.[1] It involves multiple organ systems in body and is characterized by presence of asthma, peripheral eosinophilia, neuropathy, and sinus mucosal disease[2]. It can be associated with MPO type ANCA antibodies; biopsy usually shows polymorphonuclear infiltrates of blood vessels with necrosis.[3] It responds well to steroids, additional immune suppression may be needed in some cases.

CASE PRESENTATION

A 24 years old male with past history of asthma presented with four days history of asthma worsening and symptoms of upper respiratory tract infection that were followed by difficulty walking. Patient's weakness was rapid onset, progressive and ascending in nature. Patient was wheel chair bound at presentation with power in upper limbs 4/5 in proximal upper limb and 3/5 in distal upper limb and in proximal lower limbs 3/5 in thigh and 3/5 in distal lower limb in leg muscles and 2/5 in foot and ankle muscle groups on medical research counsel grading. His deep tendon reflexes were absent with no sensory loss, no bulbar involvement and intact control of sphincters. The patient did not report any pain sensations. Auscultation of chest showed scattered wheeze.

The patient was admitted with initial diagnosis of GB syndrome on clinical basis. Patient routine labs showed a CBC with eosinophil count of 22 percent. His nerve conduction studies showed a pattern suggestive of mononeuritis multiplex. Given the history of asthma coupled with eosinophilia and mono neuritis pattern on electrodiagnosis (Figure 1, 2, 3), a diagnosis of Churg Strauss syndrome was suspected and patient was started on one gram daily intra venous methylprednisolone. Workup was started to confirm the diagnosis of Churg Strauss syndrome. A peripheral film of patient's blood sample confirmed the eosinophil counts. His serum IgE levels were over 1000 (normal <150). Other lab tests showed an ESR of 65 mm in first hour, qualitative CRP was positive. His hepatitis serology and HIV were reported negative and his autoantibodies including ANA and C-ANCA and P-ANCA were all reported negative. Chest x-ray confirmed the presence of transient pulmonary infiltrates (Figure 4) that are characteristic of this disease.

MCS

Site	Latency	Duration	Amp.	Area	Segment	Dist.	Interval	NCV
Median		**Left**						
WRIST	3.7ms	6.0ms	14.4mV	41.9mVms	*WRIST		3.7ms	
ELBOW	8.4ms	5.6ms	12.7mV	39.0mVms	*WRIST - ELBOW	230mm	4.7ms	49.1m
					ELBOW			
Median		**Right**						
WRIST	4.9ms	5.3ms	5.8mV	19.0mVms	*WRIST		4.9ms	
ELBOW	8.6ms	6.2ms	4.5mV	16.3mVms	*WRIST - ELBOW	220mm	3.7ms	59.5m
					ELBOW			
Ulnar		**Left**						
WRIST	3.6ms	5.7ms	5.6mV	12.2mVms	*WRIST		3.6ms	
B- ELBOW	8.0ms	5.8ms	5.2mV	11.6mVms	*WRIST -B- ELBOW	230mm	4.4ms	52.5m
					B-ELBOW			
Ulnar		**Right**						
WRIST	3.8ms	5.8ms	7.1mV	18.0mVms	*WRIST		3.8ms	
B- ELBOW	8.1ms	6.0ms	7.4mV	19.0mVms	*WRIST AXILLA	180mm	4.3ms	41.5m
					AXILLA			
Peroneal		**Left**						
AWE	5.1ms	6.3ms	780.0uV	2.5mVms	*WRIST		5.1ms	
B- KNEE	10.5ms	8.4ms	1.0mV	4.1mVms	WRIST-ELBOW	300mm	5.4ms	55.8m
					ELBOW			

Peroneal		**Right**						
ANKLE	5.1ms	7.5ms	860.0uV	42.9mVms	*ANKLE		5.1ms	
B-KNEE	11.8ms	7.3ms	1.0mV	2.8mVms	*ANKLE-B-KNEE	140mm	6.8ms	20.6m
					ELBOW			
Tibial		**Left**						
ANKLE	4.8ms	5.2ms	5.4mV	14.9mVms	*ANKLE		3.5ms	
POPLITIAL	14.2ms	8.0ms	4.4mV	24.8mVms	ANKLE-POPLITIAL	400mm	10.7ms	37.5m
					POPLITIAL			
Tibial		**Right**						
ANKLE	3.5ms	5.2ms	5.4mV	14.9mVms	*ANKLE		3.5ms	
POPLITIAL	14.2ms	8.0ms	4.4mV	24.8mVms	ANKLE-POPLITIAL	400mm	10.7ms	37.5m
					B-ELBOW			

SCS

Site	Lat. 1	Lat. 2	Amp.	Area	Segment	Dist.	Intvl.	NCV
Median	Right							
WRIST	3.6ms	4.9ms	9.0uV	122.8uVms	WRIST	145mm	3.6ms	40.7ms
					WRIST			

F-wave

Nerve	Tibial	Side	Right	
Stim./Site	ANKLE	Rec. Site	AHB	Distance
M-Latency		M-Amplitude		F-Occurrence
	Min	Max	Mean	
F-Latency				
F-Amplitude				
FWCV				

Figure 1: Motor and sensory Nerve Conduction studies

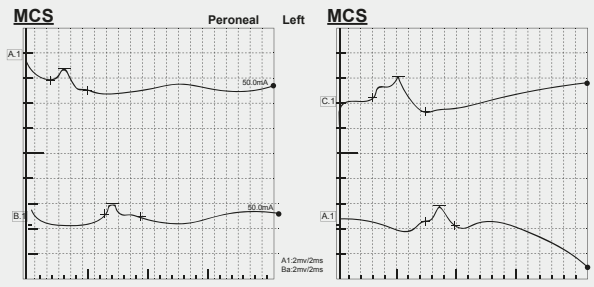

Figure 2: Motor Axonal neuropathy in Bilateral Peroneal Nerves

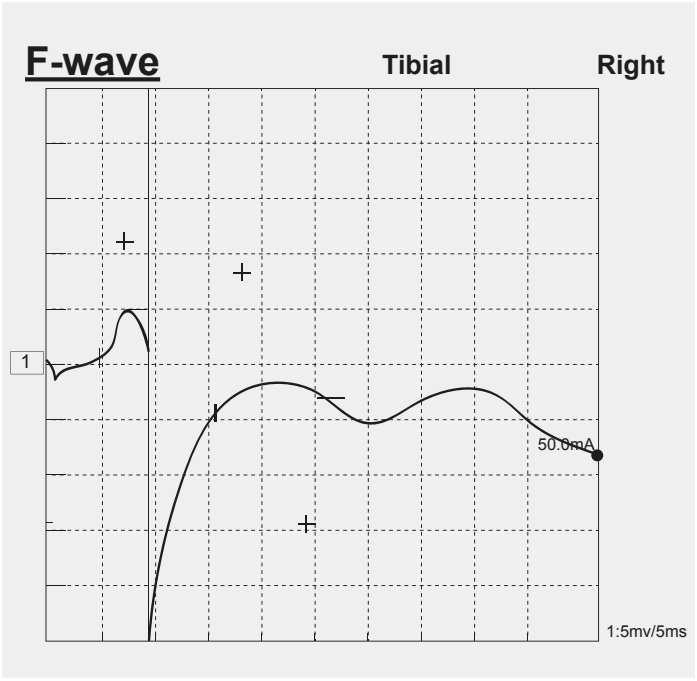

Figure 3: Absent F waves in right Tibial Nerve

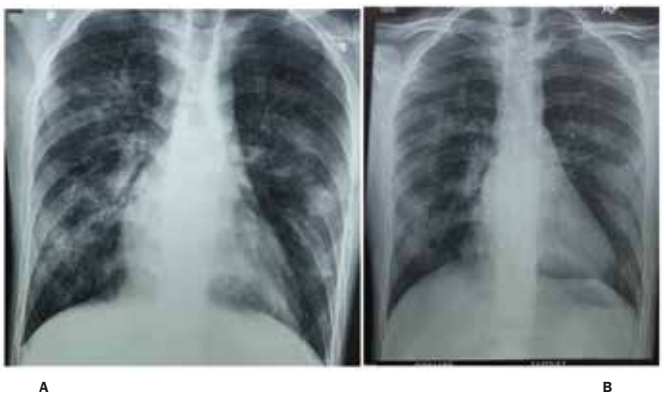

Figure 4: Transient Pulmonary Infiltrates seen in Chest Xray one day apart: A(Before) B(After)

HRCT of patient lungs showed septal thickening with possibility of pulmonary eosinophilia. A muscle biopsy taken from left rectus femoris confirmed necrotic vasculitis with polymorphonuclear (eosinophilic) infiltrate. Patient's echocardiogram showed evidence of cardiomyopathy with reduced left ventricular function and hypokinetic septum, with an ejection fraction of 35%. Patient fulfilled five out of six (four /six needed) of criteria required for diagnosis of Churg Strauss syndrome set by American college of rheumatology.

Patient was started on 1 gram daily methyl prednisolone for five days. Methyl prednisolone resulted in moderate improvement in patient symptoms which were followed by five sessions of alternate day plasmapheresis. Patient recovered significantly following plasmapheresis and was able to walk without support, his respiratory symptoms also improved. After rheumatologist consult patient was also given cyclophosphamide which resulted in remission of any residual disease and patient was asymptomatic on follow up.

DISCUSSION

Churg Strauss syndrome, also known as eosinophilic granulomatosis with polyangiitis, is a disorder of likely auto-immune etiology causing eosinophilic vasculitis of small vessels in multiple organs of body. Diagnosis is made on base of American college of rheumatology criteria which requires four out of six features being present in patient.[4] The features include asthma, paranasal sinus disease, neuropathy, evidence of eosinophilic vasculitis, pulmonary infiltrates, and eosinophilia more than 10%. Our patient satisfied five of these criteria. Other clinical features include skin rashes such as maculopapular erythematous rashes and cardiomyopathy.[3] ANCA antibodies can also be positive in up-to 60% of patients which can be associated with more fulminant disease and more risk of cardiomyopathy.[5] Acute neuropathy presenting clinically with GB syndrome like presentation is a possible presenting feature of Churg Strauss syndrome as was the case in our patient.[6]

Our patient had five out of six clinical features for diagnosis and also had evidence of cardiomyopathy. ANCA was not positive in our patient. Treatment is with IV steroids which can be supplemented with other forms of immune therapy as required. Our patient received five cycles of plasmapheresis, and cyclophosphamide.

CONCLUSION

A patient presenting with acute neuropathy and past history of asthma, a diagnosis of Churg Strauss syndrome should be kept in mind, and a high eosinophil count on routine blood counts warrants a further work up and treatment for Churg Strauss syndrome.

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Zaid Waqar; data collection, data analysis, manuscript writing, manuscript review