

## Grand Rounds in Rheumatology

# Miller Fisher syndrome in adult onset Still's disease: case report and review of the literature of other neurological manifestations

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### Abstract

Adult-onset Still's disease (AOSD) is a multi-system inflammatory disorder characterized by high spiking fevers, evanescent salmon-coloured rash, arthralgias or arthritis, hepatosplenomegaly, lymphadenopathy and sore throat. There is no specific test or combination of tests that can establish the diagnosis of AOSD and patients may present with other systemic involvement including neurological manifestations in 7–12% of cases. We present a complex case of a patient with AOSD who developed the Miller-Fisher variant of Guillain-Barré syndrome. This immunological disorder of the nervous system has not been described in association with AOSD before. We also review the literature on other neurological manifestations in AOSD. AOSD mimics different disease processes and its multi-system manifestations may complicate the picture further.

**KEY WORDS:** Miller Fisher syndrome, Still's disease, Guillain-Barré syndrome, Neurological manifestations, Adult Still's disease, gangliosides.

### Introduction

Adult onset Still's disease (AOSD) is a multi-system, inflammatory disorder characterized by high spiking fevers, evanescent salmon-coloured rash, arthralgias or arthritis, hepatosplenomegaly, lymphadenopathy and sore throat (Table 1). Patients often have a marked leucocytosis while other markers of rheumatological disorders, such as rheumatoid factor and antinuclear antibody, are negative. Other laboratory manifestations observed in AOSD include elevations in liver enzymes, abnormalities in haematological function ranging from anaemia to disseminated intravascular coagulation and markedly elevated levels of ferritin in the active stage of disease [1–4]. There is no specific test or combination of tests that can establish the diagnosis of AOSD. However, in the presence of a compatible clinical scenario, serum ferritin higher than 3200 ng/ml is highly suggestive of AOSD [5].

Almost all organ system involvement has been noted in patients with AOSD. These include pulmonary manifestations ranging from pleuritis to pneumonitis, cardiac abnormalities such as pericarditis, pericardial

tamponade and myocarditis, and renal manifestations presenting as proteinuria during the febrile phase as well as microscopic haematuria and nephrotic syndrome [2]. We describe a complex case of AOSD who developed the Miller-Fisher variant of Guillain-Barré syndrome and review the literature on neurological manifestations of AOSD.

A 19 yr-old Hispanic female with an unremarkable medical history, who was 4 months pregnant, presented to the obstetrics service in late 1995 with arthralgias and a maculopapular rash during her second pregnancy. She complained of sore throat and pain and swelling in the wrists, metacarpophalangeal joints and proximal interphalangeal joints of both hands. Her elbows, shoulders, knees, and toes were involved to a lesser degree. The rash was diffuse and mildly pruritic.

On examination, she was febrile to 38°C with normal blood pressure and pulse. An erythematous maculopapular rash was evident over the forearms, thighs, abdomen and back. There was diffuse muscle tenderness but no synovitis. The remainder of her examination was unremarkable apart from the pregnancy. Her erythrocyte sedimentation rate was 103 mm/h; antinuclear antibody, rheumatoid factor and serologies for human immunodeficiency virus, rubella and Epstein-Barr virus were all negative. The titre of parvovirus B19 IgM was

positive at 1:56. She was diagnosed with acute parvovirus B19 arthritis and was treated with prednisone 10 mg daily with dramatic improvement in her aches and pains as well as rash.

Over the next 3 months, however, she continued to have intermittent arthralgias, rash and fevers. She also complained of fatigue and began losing weight during her third trimester. This as well as evidence of fetal hydrops, necessitated delivery by Caesarian section at 7 months. The baby was diagnosed with congenital cytomegalovirus infection and required prolonged admission to the intensive care unit.

Post-partum, she continued to complain of febrile episodes, arthralgias and diffuse myalgias severe enough to interfere with her ability to care for her two small children. She denied oral ulcers, alopecia, shortness of breath, pleuritic chest pain, abdominal pain, Raynaud's phenomenon, photosensitivity, numbness, paraesthesias and focal weakness. The macular rash was apparent during febrile episodes and again involved primarily the proximal limbs and trunk. A new finding was posterior cervical and axillary lymphadenopathy. Laboratory investigations revealed haemoglobin of 7.9 g/dl, which was attributed to her recent Caesarian section as well as poor nutritional status. The white blood cell count was in the range of 13 000 mm<sup>3</sup>. Repeat parvovirus B19 IgM and IgG were negative.

Six months after her initial presentation, she was admitted for further work up of persistent arthralgias, intermittent fevers, progressive weight loss and profound anaemia, which had proven refractory to iron supplementation. This was the first of many admissions and the beginning of a very thorough and expensive evaluation. Over the course of the next 18 months, multiple investigations were undertaken, as outlined in Tables 2–4. In August 1996 she underwent a laparotomy with liver biopsy, sampling of multiple abdominal lymph nodes and repeat bone marrow biopsy. Splenectomy was performed for 'lymphoma staging' and possible idiopathic thrombocytopenic purpura as her platelets dropped as low as 20 000 mm<sup>3</sup>. Serum and urine protein electrophoresis was non-specific and did not suggest a monoclonal gammopathy. The liver biopsy and histopathology of lymph nodes and spleen did not show evidence of lymphoma. She developed a leucocytosis in the range of 20 000–49 000 cells/mm<sup>3</sup> and her serum ferritin levels were noted to be markedly elevated, to between 2400 and 9850 ng/ml (normal value 16–400 ng/ml).

A diagnosis of AOSD was made (Table 1). In addition to diagnostic criteria, supportive features included the high serum ferritin concentration and wrist radiographs showing non-erosive bony fusion of the carpal bones (Fig. 1).

She was treated initially with non-steroidal anti-inflammatory drugs and then with prednisone at doses of 10–20 mg/day, though doses in the range of 1 mg/kg/day were often required for flares. Methotrexate was added in November 1996. However, despite doses up to 20 mg per week, she remained symptomatic.

TABLE 1. Criteria for the diagnosis of adult onset Still's disease (Yamaguchi *et al.* 1992 [28]). Five or more criteria (including two major) required for diagnosis

Major criteria	Minor criteria
Fever > 39°C	Sore throat
Arthralgias	Lymphadenopathy or splenomegaly
Still's rash	Hepatic dysfunction
Neutrophilic leucocytosis	Negative RF and ANA

ANA, antinuclear antibody; RF, rheumatoid factor.

TABLE 2. Positive/abnormal tests

Variable	Value	Normal range
Erythrocyte sedimentation rate (mm/h)	120	(0–20)
C-reactive protein (mg/dl)	21.6	0–0.8
Haemoglobin (g/dl)	7.9	12–15
White blood cell count (× 10 <sup>6</sup> /mm <sup>3</sup> )	25.6	3.4–10
Aspartate transaminase (U/L)	88	11–32
Alanine transaminase (U/L)	65	5–30
Lactate dehydrogenase (U/L)	317	110–205
Ferritin (ng/ml)	9846	12–250

TABLE 3. Negative/normal tests

Antinuclear antibodies
Anti-double-stranded DNA antibodies
Anti-Smith
Anti-Ro
Anti-La
Anti-RNP
C3
C4
Rheumatoid factor
ANCA
Serum protein electrophoresis
Creatine kinase
Angiotensin converting enzyme concentration
Anti-cardiolipin antibody
Familial Mediterranean fever genetic test
Lyme antibody
VDRL test
Purified protein derivative
Cytomegalovirus (buffy coat, urine)
Human immunodeficiency virus antibody
Cultures: blood, urine, cerebrospinal fluid, bone marrow, broncho-alveolar lavage fluid

TABLE 4. Pathology reports

Bone marrow	Marked granulocytic hyperplasia; no dysplasia
Liver	Patchy periportal inflammation; moderate steatosis
Cervical lymph node	Non-specific reactive histology
Lacrimal gland	Normal histology with no lymphoid infiltrate or evidence of epithelial malignancy
Spleen	Follicular hyperplasia; negative for lymphoma
Small intestine	Mild villous atrophy; Congo Red stain negative for amyloid

Methotrexate was stopped in October 1997 because of treatment failure.

Over a 2-yr period, she needed more than 20 admissions to the medical ward for flares of AOSD. These were manifested as high fevers, evanescent rash, arthralgias, abdominal pain, an impressive leucocytosis (average 20 000–30 000 cells/mm<sup>3</sup>) and high serum ferritin levels. A typical admission lasted 2–3 days and consisted of multiple investigations, primarily aimed at ruling out an infectious aetiology in a splenectomized patient. After another flare in May 1998, however, she experienced an 8-month period during which her disease was quiescent. In January 1999, her disease flared again, requiring admission almost monthly for management of flares.

In May 1999, she was admitted with generalized weakness, postural dizziness, painful oral ulcers, facial paraesthesias and an acute onset of watery diarrhoea. Initial examination confirmed severe orthostatic hypotension (standing blood pressure 50/25 mmHg). She was afebrile and there was no rash. Ragged-edged and shallow based ulcers were noted on the tongue, buccal mucosa and gingiva. Neurological examination was remarkable for diffuse and symmetric weakness (graded 4–5 throughout) and absence of both deep tendon

and corneal reflexes. During the next 2 days, she developed diplopia (without objective extraocular muscle dysmotility), dysphagia and facial diplegia. Magnetic resonance imaging of the brain showed no significant abnormality. Examination of cerebrospinal fluid revealed markedly elevated protein (421 mg/dl) but a normal opening pressure, cell count and glucose. Gram stain and bacterial and fungal cultures were negative. Stool cultures were negative. Biopsy of the oral lesions was consistent with aphthous ulcer and the Tzanck smear for herpes infection was negative. Nerve conduction studies demonstrated a substantial lag time in combined potentials, suggestive of a demyelinating process (Fig. 2).

These findings were consistent with the Miller Fisher variant of GBS. She was treated with pooled intravenous immunoglobulin 2 mg/kg/day for 3 days. Her cranial nerve palsies resolved within 1 week and she was discharged to a nursing home for rehabilitation.

After discharge, the areflexia has persisted and her strength has improved only marginally. Her course has been complicated by malnutrition and weight loss, necessitating gastrostomy tube feeding. She is ambulatory, but has been unable to work or independently care for herself and her two children.



FIG. 1. X-ray demonstrating non-erosive fusion of carpal bones in a pericarpitate pattern, a distinctive feature of AOSD.

## Discussion

Still's disease, first described by George Still in 1896, is the systemic form of juvenile rheumatoid arthritis [6]. Bywaters in 1971 described fourteen adults with an illness resembling Still's disease and coined the term AOSD [7].

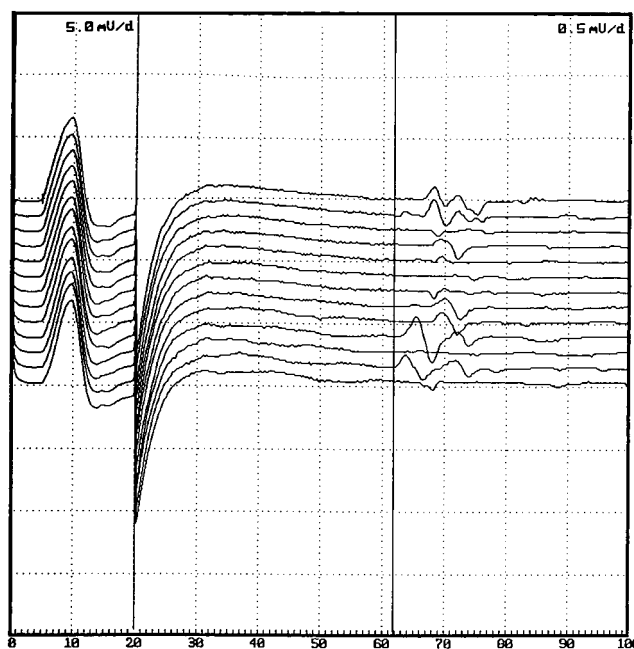


FIG. 2. F response abnormality: right tibial F response, 13 traces. Minimal F response is prolonged at 61.8 ms (normal < 50 ms).

TABLE 5. Neurological involvement of AOSD: review of the English literature

Reference	Patient and disease characteristics			Neurological findings	Conclusion
	No. of cases	Sex/age	AOSD characteristics		
Reginato <i>et al.</i> 1987 [2]	3 (of authors' own patients)	20/F	Fevers, arthritis, sore throat, rash	(1) Acute disorientation, hallucinations during high fever (preceding liver failure)	First two patients had improvement in neurological symptoms with resolution of liver failure while on high-dose steroids Third patient had residual Bell's palsy after steroid therapy Overall, only one patient had a true manifestation of a neurological disorder with an acute flare of Still's disease. The other two were complicated by liver failure Authors' review on neurological manifestations of these six patients. Three non-English references, 1 reference unavailable. Bell's palsy patient had palsy since childhood; status epilepticus patient with erosion of sella turcica died with status epilepticus and cardiovascular collapse (see below). Overall not clear who had true neurological manifestations
		28/F	Fevers, arthritis, sore throat, rash	(2) Asterixis, mental status confusion, increased intracranial pressure during liver failure	
		25/M	Fevers, arthritis, sore throat, rash	(3) Lower motor neurone facial nerve palsy during acute flare	
Garrote <i>et al.</i> 1993 [14]	6 (in review)	Not available	Unknown	Transient pyramidal tract signs (French), third nerve palsy (French), Bell's palsy [18], status epilepticus [27], brain stem haemorrhage (German), symmetric sensory neuropathy (English)	Neurological symptoms resolved prior to non-steroidal therapy. The authors hypothesize that the neurological manifestations may be due to vascular involvement of the CNS and may be focal and transient; however, it is not clear that it is associated with AOSD as it resolved without therapy Evidence of meningoencephalitis: resolved without therapy raising the issue of whether it was related to Still's disease or a presentation of an initial viral illness The sensorimotor peripheral neuropathy resolved with steroid therapy No discussion in paper of timing of diagnosis, i.e. acute flare of Still's or not, association with other complications, etc. No details of timing of diagnosis, resolution of symptoms or timing with therapy; however, patients with disorientation and confusion also had AOSD related hepatic failure
		16/M	Sore throat, myalgia, arthralgia, headache and vomiting, rash, fever and hepatosplenomegaly present on admission	Transient diplopia, oscillopsia, horizontal nystagmus on left gaze, paraesthesias, dysaesthesias on left side of face, unstable gait, weakness upper right extremity and clonus right ankle CT with hypodensity in left pons; MRI with T2 hyperintensity on left middle cerebellar and cerebral peduncles	
Denault <i>et al.</i> 1990 [15]	1	20/M	Headache, fever, cough, myalgias and weight loss with development of arthritis later in the course	Neck stiffness, confusion, decreased consciousness and incontinence. CSF pleocytosis noted. CT normal. Mentation slowly improved without therapy. Two months later patient developed sensorimotor peripheral neuropathy documented by NCV and sural nerve biopsy	
Ohta <i>et al.</i> 1990 [4]	11	Age/Sex not specified	Unknown in specific cases. All patients had definite Still's fever, joint symptoms, rash, lymphadenopathy	5 with peripheral neuropathy; 4 with aseptic meningoencephalitis; 1 with delirium, convulsion, rigidity (1 person had both peripheral and CNS involvement)	
Pouchot <i>et al.</i> 1991 [1]	5	Age/Sex not specified	Unknown in specific cases; Patients had diagnosis of AOSD based on Medsger and Christy criteria	2 patients with disorientation and confusion who developed hepatic failure; 1 patient with peripheral facial nerve palsy; 2 patients with transient deafness (not due to ASA); 1 patient with vertical diplopia with ptosis due to 3rd nerve palsy	

TABLE 5. (Continued)

Reference	Patient and disease characteristics			Neurological findings	Conclusion
	No. of cases	Sex/age	AOSD characteristics		
Ohta <i>et al.</i> 1987 [3]	Review of 228 cases of AOSD from the literature; 11 patients with neurological symptoms		Unknown in specific cases	3 patients with meningoencephalopathy 1 patient with brainstem haemorrhage 1 patient with status epilepticus 1 patient with pyramidal syndrome 2 patients with CNS abnormalities 2 patients with cranial nerve paresis 1 patient with orbital pseudotumour	French; Ohta's own 2 patients See [16] See [27] French paper See Baker <i>et al.</i> [17] French paper; see [18] See [25]
Wouters <i>et al.</i> 1985 [16]	1	33/M	High fever, evanescent rash, polyarthritits	Patient developed diplopia and ptosis of left eyelid after having a total hip replacement; CT with small haemorrhage in brainstem. Patient presumably with active flare given significant polyarthritits	Paper not specific on activity of disease but presumably active with polyarthritits. Prednisone therapy relieved polyarthritits. No mention of neurological complications/resolution
Baker <i>et al.</i> 1979 [17]	2	Age/sex not specified	Spiking fever, evanescent rash, polyarthritits, leucocytosis	Abstract mentions CNS manifestations in two patients. Unclear if two patients with acute liver failure who developed delirium and encephalopathy with increased ICP were those two patients	If 2 patients described with liver failure and mentation difficulties are the 2 with CNS manifestations, this would not qualify as a direct result of AOSD
Kaplinsky <i>et al.</i> 1980 [18]	1	34/F	Polyarthralgia, fever, rash	Facial nerve paresis 'that has been present since childhood'	No relationship between AOSD and neurological complications
Marie <i>et al.</i> 1999 [12]	1	17/M	Headache, left 7th cranial nerve palsy, sore throat, high fever, arthralgias, myalgia, lymphadenopathy and meningial syndrome	7th cranial nerve palsy Meningeal symptoms with CSF showing lymphocytosis CT scan normal	Treatment with high dose aspirin resulted in complete resolution of symptoms. There appears to be a true relationship between the AOSD and a neurological condition
Markusse <i>et al.</i> 1988 [20]	1	26/M	9 year history of fevers, evanescent rash, arthralgias, lymphadenopathy, leucocytosis and liver enzyme abnormalities	Gradual development of hearing loss and tinnitus on admission with fevers, arthritits, rash, and weight loss. CT normal, CSF low cell count; indomethacin treatment stopped—hearing loss remained. Improved with prednisone; all other symptoms improved	All studies pointed to cochlear involvement, not related to indomethacin, all occurring during acute AOSD flare. This case supports an association between neurological conditions and AOSD
Scully <i>et al.</i> 1989 [21]	1	59/F	Patient initially had a facial palsy with resolution. 1 week later patient developed sore throat, weakness and fevers	Facial palsy preceded symptoms of AOSD and resolved within 1 week	Unclear if AOSD and neurological symptom were related. NEJM discussant describes three other patients with lymphocytic meningoencephalitis, transient bilateral third cranial nerve palsy, and unilateral peripheral facial nerve palsy of 3 months duration. Timing of diagnosis, resolution of symptoms, etc. is not further described
Cush <i>et al.</i> 1985 [22]	1	21/M	Fever, chills, sweats, sore throat, myalgia and anorexia, leucocytosis	Patient developed ptosis of the right eye with diplopia and orbital pain on lateral gaze. CT normal. Patient developed ptosis in the left eye along with fever and confluent rash	All symptoms resolved with anti-inflammatory therapy. Patient was believed to have developed an acute inflammatory orbital pseudo-tumour

Kurabayashi <i>et al.</i> 1996 [23]	1	75/F	High fevers, salmon-pink eruption, lymphadenopathy, splenomegaly, high ferritin, negative ANA	Left sided hemiparesis and disturbance of consciousness CT with cerebral haemorrhage in region of right thalamus to putamen	Given patient's age it is difficult to know if haemorrhage was secondary to age or to AOSD
Mok <i>et al.</i> 1998 [24]	0		Sixteen Chinese patients identified as having AOSD by Medsger and Christy criteria		No neurological abnormalities were seen in a Chinese patient panel
Cush <i>et al.</i> 1987 [25]	2	Age/sex not specified	Total of 21 patients identified with AOSD—fever, rash, arthralgia, etc.	2 patients described having peripheral neuropathy and ulnar neuropathy	No further comment on onset or resolution with therapy
Goldman <i>et al.</i> 1980 [27]	13: 1 noted with neurological symptoms	8/M 5/F	All with spiking fever, polyarthralgias, leucocytoses and negative RF	1 woman described to have erosion of the sella tursica who died with status epilepticus followed by cardiovascular collapse (no autopsy)	No clear association with AOSD and status epilepticus

Nervous system involvement has been described in approximately 7–12% of patients with AOSD [1, 4]; however, neither the GBS nor the Miller Fisher syndrome (MFS) has been described before in a patient with AOSD. We present the first case report of a patient with AOSD who developed the Miller Fisher variant of GBS. Our patient had the characteristic findings of MFS confirmed by electromyographic studies.

GBS is characterized by progressive symmetrical limb weakness, areflexia, absent or mild sensory signs and variable autonomic dysfunctions. C. Miller Fisher described a syndrome consisting of ophthalmoplegia, ataxia and areflexia [8]. He regarded this syndrome as a variant of GBS; however, the relationship between MFS and GBS is controversial. Many patients with GBS and MFS report an antecedent, acute infectious illness or gastroenteritis that often resolves at the onset of neurological symptoms, such as our patient demonstrated. *Campylobacter jejuni* has been recognized as the most frequent antecedent pathogen for GBS and has also been reported in the Miller Fisher variant syndrome [9, 10]. Immune responses against *C. jejuni* have been postulated to be involved in both GBS and MFS by induction of a specific anti-ganglioside antibody which cross-reacts with neural tissues [11]. In GBS the specificity of anti-ganglioside antibodies is very variable and the presence of anti-GM1 ganglioside IgG antibodies may be associated with a more severe form of GBS. High titres of anti-GQ1b antibodies were found in MFS patients, as opposed to control subjects [12, 13]. Although we did not detect the anti-ganglioside antibody we believe that our patients' antecedent diarrhoeal illness was probably related to the subsequent development of the MFS. It is not possible to speculate whether this patient's MFS was causally related to her AOSD as the immunological basis of AOSD still remains unclear. It is interesting to postulate that the two disorders may share an infectious trigger, thus allowing the presentation of one with another.

A review of literature on neurological manifestations of AOSD revealed multiple problems with the case reports (Table 5). It is notable that in many case reports the neurological manifestations have no relationship to AOSD. Neurological syndromes, such as encephalopathy, delirium and decreased mentation, are often described in the literature as neurological complications of AOSD when they could be the manifestations of acute liver failure secondary to AOSD. Some neurological manifestations are reported during the acute phase of AOSD but resolve prior to appropriate therapy with anti-inflammatory medication, making it less likely to be related to the original disease. In most references it is difficult to distinguish a neurological complication associated with AOSD from a separate complication entirely. Overall, the review of the literature shows that there is a trend for some neurological complications, such as cranial nerve paresis, peripheral neuropathy and meningoencephalitis, to be associated with AOSD or a flare.

The diagnosis of AOSD is often difficult as it mimics systemic infections, systemic vasculitis or even neoplastic conditions such as lymphoma. As our case demonstrates, neurological manifestations can complicate the picture further.

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