

## **Acute Pancreatitis in a Case of Mixed Connective Tissue Disease (MCTD)- A Rare Manifestation of Macrophage Activation Syndrome (MAS)**

**Suparna Guha\***

*Department of Pediatric Medicine, VIMS, India*

**\*Corresponding Author:** Suparna Guha, Department of Pediatric Medicine, VIMS, India.

**Received:** August 17, 2020; **Published:** September 10, 2020

### **Abstract**

The common rheumatological diseases which develop Macrophage activation syndrome (MAS), are Systemic Lupus Erythematosus (SLE), Systemic Onset Juvenile Idiopathic Arthritis (SOJIA) and Kawasaki Disease.

Among the connective tissue diseases, incidence of MCTD is quite rare in children; added to this, MAS in juvenile MCTD is also a very rare occurrence.

Here we report the case of a 12-year-old girl child who had overlapping features of juvenile idiopathic arthritis (JIA) and systemic sclerosis along with a very high titer of Anti U1 RNP antibodies.

She later developed severe pain abdomen along with high fever. Investigations revealed evidence of pancreatitis in the background of a macrophage activation syndrome. She responded to methylprednisolone pulse therapy.

Gastrointestinal complications like esophageal motility disorders, malabsorption have been reported in MCTD. But to the best of our knowledge acute pancreatitis with MAS in juvenile MCTD has not been reported so far.

**Keywords:** MAS; MCTD; Pancreatitis

### **Introduction**

The common rheumatological diseases which develop Macrophage activation syndrome (MAS), are Systemic Lupus Erythematosus (SLE), Systemic Onset Juvenile Idiopathic Arthritis (SOJIA) and Kawasaki Disease.

### **Case Report**

A 12-year female child presented at our outdoor patient department with a history of joint pain for almost a year.

Initially she had morning stiffness and polyarthralgia along with swelling of the hands. She later started developing deformities of the small joints of the hand.

She had been diagnosed as a case of seropositive JIA (since rheumatoid factor was positive 1:120) and was prescribed weekly 15mg subcutaneous methotrexate along with folic acid.

But even after 6 months of therapy she continued to have joint flares and her deformities were progressively increasing.

---

**Citation:** Suparna Guha. "Acute Pancreatitis in a Case of Mixed Connective Tissue Disease (MCTD)- A Rare Manifestation of Macrophage Activation Syndrome (MAS)". *EC Paediatrics* 9.10 (2020): 27-30.

She had recently developed difficulty in swallowing along with a history suggestive of Raynaud's phenomenon.

### **On examination**

She was thin built (wt. 22 kgs, Ht 145 cm). She was pale with pinched-up facies.

Her skin was thin taut shiny and she had sclerodactyly along with a few digital ulcers.

She had synovitis of bilateral knee and ankle joints. It was so severe that she could barely walk.

Investigations revealed a Hb of 6.5 gm/dl; total leucocyte count (TLC), platelets, liver and renal function tests were within normal limits. Inflammatory markers were high, ESR-120 mm and C Reactive Protein (CRP) being 24 mg/dl.

ANA (IIF, Hep2) was positive (1:320, speckled pattern) and ENA revealed a positive anti U1RNP in very high titer.

Her complement (C3 and C4) were within normal limits and urine did not show active sediments or significant proteinuria.

Her CXR and pulmonary function tests and echocardiography were normal.

Since she was having feeding problems a barium swallow was done; but it was normal.

We treated her with a short course of steroid (0.5 mg/kg prednisolone), anti-reflux (pantoprazole) and continued methotrexate. For Reynaud's we advised conservative therapy and started calcium channel blockers.

She was doing well; but after 8 months she was brought to the ER with acute pain abdomen and vomiting and fever.

She was immediately shifted to the intensive care unit with the possibilities of an intestinal obstruction, intestinal perforation or GI vasculitis.

On examination, she was very toxic with severe epigastric tenderness but her vitals were stable.

Investigations revealed a pancytopenia (Hb-6 gm/dl, TLC 4500/cumm; P56L42M2 Platelets 90,000/cumm), with an ESR-45 mm, CRP-120 mg/dl, Ferritin-16,488 ng/ml, triglycerides-326 mg/dl, fibrinogen-124 mg/dl and coagulation profile were deranged.

Her LFT showed an SGOT of 381 IU/L and SGPT 90 IU/L. Her pancreatic enzymes were elevated with an amylase of 353U/L and amylase of 3036 U/L.

Infective serology for Cytomegalovirus (CMV), Epstein Barr Virus (EBV) and Parvovirus & Hepatitis virus (A, B, C) were negative. Her blood culture was also sterile.

USG and Straight Xray abdomen were normal.

CT abdomen revealed diffuse enlargement of pancreas with peripancreatic edema.

In the background of the above clinical scenario she was diagnosed to have MAS with acute pancreatitis.

She was managed conservatively and IV methylprednisolone @30 mg/kg was given for 3 days. She gradually improved, her pancreatic enzymes started declining and she became afebrile. She was discharged with tapering doses of oral steroids and methotrexate was continued.

## Discussion

MCTD in the pediatric age group is very rare. The overall incidence of juvenile MCTD may be as low as 0.5% [1]. Although there are studies on adult MCTD [2,3], the juvenile variety has been hardly reported from the Indian sub-continent; there are some studies from the West [4,5].

There are very few case reports of MAS in MCTD in adults [7]. There was a report of MAS with refractory Raynaud's phenomenon developing in a 13-year-old girl with MCTD [6]. Our patient developed MAS with acute pancreatitis. Bone marrow could not be done as parents did not give consent to an invasive procedure.

Gastrointestinal involvement mainly occurs in the form of esophageal dysmotility, reflux esophagitis or malabsorption syndrome [8,9]. More common causes of pain abdomen in MCTD are colonic perforation, pseudo obstruction etc. In a large series of gastrointestinal complications occurring in MCTD cases, acute pancreatitis was present in only 1 patient [8].

The main objective of reporting this case was to highlight the fact that acute pancreatitis may be a rare but possible cause of acute abdomen in MCTD. In the background of a connective tissue disease with immune suppressive therapy, the most common cause is infective, though autoimmune is also a possibility.

Infective causes (since the child was on immune suppressive therapy) of acute pancreatitis were ruled out. So, the next possibility was an autoimmune cause and she responded dramatically to IV steroids.

This autoimmune pancreatitis might have been a probable complication of MAS.

Again, the trigger for MAS could not be identified as most of the infective work up was negative.

So, it is very difficult to suggest whether MAS was complicated by pancreatitis or pancreatitis triggered MAS.

## Conclusion

Acute pancreatitis is a rare cause of pain abdomen in MCTD and along with routine investigations, a pancreatic enzyme study should also be done.

## Bibliography

1. Berard RA and Laxer M. "Pediatric Mixed Connective Tissue Disease". *Current Rheumatology Reports* 18.5 (2016): 28.
2. Lawrence A., et al. "MCTD: Is it rare in India?" *Clinical Rheumatology* 26.2 (2007): 205-207.
3. John JK., et al. "Clinical and Immunological Profile of Mixed Connective Tissue Disease and a comparison of four diagnostic criteria". *International Journal of Rheumatology* (2020): 1-6.
4. Deepak S and Waarier KC. "Mixed Connective Tissue Disease in Children- case series". *Rheumatology and Orthopedic Medicine* 2.2 (2017): 1-7.
5. Siri Opsahl Hetlevik., et al. "Long-term outcome in juvenile -onset mixed connective tissue disease: a Norwegian study". *Annals of the Rheumatic Diseases* 76.1 (2017): 159-165.

6. Amar HK, *et al.* "An unusual association between hemophagocytic lymphohistiocytosis, mixed connective tissue disease and autoimmune hemolytic anemia". *Medicine* 96 (2017): 28.
7. Parvaneh V J and Shiari R. "Juvenile onset mixed connective tissue disease associated with macrophage activation syndrome". *The Egyptian Rheumatologist* 41.4 (2019): 323-325.
8. Marshall J B, *et al.* "Gastrointestinal manifestations of mixed connective tissue disease". *Gastroenterology* 98 (1990): 1232-1238.
9. Daniel L, *et al.* "Gastrointestinal systemic sclerosis in serologic mixed connective tissue disease". *Arthritis and Rheumatism* 21.7 (1978): 811-819.

**Volume 9 Issue 10 October 2020**

**All rights reserved by Suparna Guha.**