Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2015; 3(3):249-251 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjmcr.2015.v03i03.021

Systemic Onset Juvenile Idiopathic Arthritis: Don't be fool by Positive Serology

Naresh Tayade*[1], Hement Murkey[2], Manjusha Deotale[3]
[1]Consultant, Life Care Neonatal and Pediatric critical care center Amravati, Maharashtra, India
[2]Director & Head, Ayushkam Bal Rugannalaya Amravati, Maharashtra, India
[3]Community Medicine, Assistant professor at Dr PDMMC, Amravati, Maharashtra, India

***Corresponding Author:**
Name: Dr. Naresh Tayade
Email: nareshtayade@yahoo.co.in

Abstract: A 5 years female child was admitted with prolonged fever and rash of 3 week. On history systemic onset juvenile idiopathic arthritis (SOJIA) was one of the possibilities but positive widal titer shifted our diagnosis to typhoid fever. After some day she developed circulatory failure with respiratory distress with IgM positive dengue titer from which she survived with supportive care. During this episode we reviewed whole case again and able to reach the correct diagnosis of SOJIA with possibility of macrophage activation syndrome (MAS).
Keywords: Fever with rash, Fever of unknown origin, Systemic onset juvenile arthritis, Macrophage activation syndrome

INTRODUCTION

Fever of unknown origin (FUO) is due to various causes, of which infectious is common one but other causes like malignancy, connective tissue diseases, etc should also be considered. Occurrence of rash along with fever and characterization of rash really help us to narrow down the possibilities and reach the exact diagnosis. Sometimes too much dependence on lab investigations rather than clinical judgment may lead to wrong diagnosis and management.

CASE REPORT

AB 5 year old female 2nd child born of nonconsanguinious marriage presented with high grade fever of 3 weeks duration (Fig. 1) with maculopapular fine rash on trunk and extremities whenever she got fever and disappear as fever subside. She also had hepatospleenomegaly for which she was investigated and treated in other hospital as indoor patient for 10 days. She received antimalarial and azithomycin along with intravenous fluid. She came to us as there was no improvement of fever. On admission she had high grade fever (103^0 F) with maculopapular rash, tachycardia, and hepatomegaly (4 cm below costal margin) with soft to firm in consistency, spleen just palpable. No desquamation of skin, lympahadenopathy, red/ white tongue was noted. Blood pressure was normal for her age & weight. Her weight was 13 kg with height of 105 cm both at 3rd percentile as per IAP growth chart. On detailed enquiry, parents gave history of arthralgia in wrist and small joints of hand and both knee but without any obvious joint swelling or restriction of joint movement. There was no history of redness of eyes, swelling at neck, dysuria, desquamation of skin, bleeding from any site. Investigations done outside showed anemia, with leukocytosis and neutophilia, platelet was increased in next CBC (Table 1), widal and malarial antigen was negative. USG Abdomen showed hepatospleenomegaly. No past history of previous admission. She was immunized as per national immunization schedule till 1.5 year with normal developmental history. Diagnostic possibility of urinary tract infection, typhoid, systemic onset juvenile arthritis or other connective tissue diseases, brucellosis, leukemia, disseminated tuberculosis, bacterial endocarditis was kept in mind. Investigations like CBC, ESR, Urine routine and widal were done. CBC showed anemia, with neutrophilia (Table 1), widal came positive with titer 1:160 for both O & H Antigen. Urine showed 4-5 pus cells and malaria antigen test was negative. She was started on IV Ceftriaxone but fever continued so IV Oflaxacin was added later. As fever continued after 3 days of antibiotics, bone marrow examination and culture, tuberculin test, and CSF was done which came normal. On 5th day evening she developed convulsions, petechiae and purpura along with respiratory distress and circulatory collapse. Immediate resuscitation was done with intravenous fluid boluses, ventilatory and ionotrophic support with dopamine and dobutamine. Hemogram showed decrease in all cell line (Table 1) so given FFP, Platelet and PCV transfusion. USG Abdomen and Thorax was done which showed ascites with bilateral plural effusion (PLEF). Dengue titer was sent which came positive for IgM. But unusal presentation of patient and previous suspicion of connective tissue disorder, case was

revised, discussed with pediatric rheumatologist and Systemic onset Juvenile arthritis (SOJIA) with macrophage activation syndrome (MAS) was the most likely possibility. Other investigation showed serum albumin 2.40gm, serum creatinine 0.38mg/dl and ESR was 4 during this episode. X-ray showed bilateral PLEF with cardiomegaly. Patient gradually became better, weaned off from ventilator and ionotrophic support. Echocardiography and color doppler was done which were suggestive of dilated cardiomyopathy. So started on digoxin, ramapril and carnitor and continued on prednisolone. NSAID, Methotrexate & steroid was started. Follow up after 10 days and 1 month showed normal count of patient and improved cardiac function.

Table 1: Hemogram report of patient

Date	9/5/11	16/5/11	20/5/11	27/5/11	5/6/11	17/6/11
Hb (g/dl)	9.9	8.8	8.9	9.7	12.6	11.9
PCV (%)	20.2	26.5	26	28.4	36.4	33.3
TLC (cells/ microlit)	20100	15500	14700	4100	2700	13400
DLC	P71L19M10	P61L30	P81L14E2M3	P70L27	P55L40E1M4	P75L18E2M5
RBC	3.72	3.27	3.48	3.83	4.83	4.47
MCV	73.45	81.1	74.51	74-13	75.36	74.5
MCH			25.57	25.33	26.09	26.62
MCHC			34.23	34.15	34.62	35.74
PLT	573000	801000	391000	42000	31000	408000

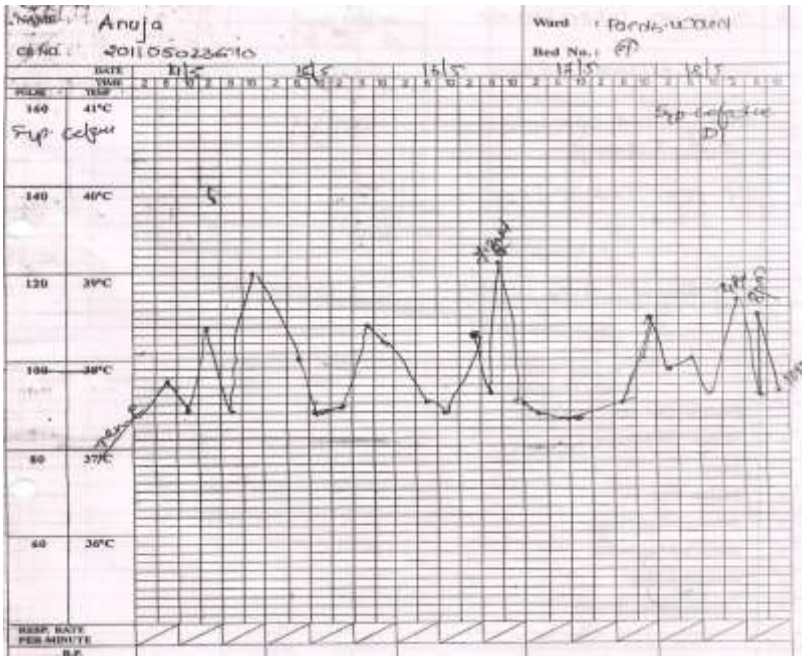

Fig. 1: Fever charting of this patient

Any patient with rash and fever of prolonged duration not responding to antibiotics should be evaluated for rheumatic disease. We can get positive serological test for infectious diseases due to hypergammaglobunimia. Complication like macrophage activation syndrome may result in morbidity and mortality. Patient presented only with fever and rash without any joint swelling. She was both widal and dengue positive. She had complication like MAS and dilated cardiomyopathy. But after conservative management she came out of this complication.

DISCUSSION

The term fever of unknown origin is applied to fever documented by health care provider and for which the cause could not be identified after 3 wk of evaluation as an outpatient or after 1 wk of evaluation in hospital [1]. Connective tissue disorder is always a possibility in such child diagnosis of which may be difficult as all manifestations may not present at same time. Systemic onset juvenile idiopathic arthritis (SOJIA) is special form of Juvenile idiopathic arthritis which is without any age, gender or HLA association [2]. At onset extra-articular manifestations including rash, fever, lymphadenopathy, hepatosplenomegaly and serositis predominates. Children with SOJIA typically have 2 weeks of high-spiking fever classically with 2 peaks daily (Double quotidian) during episode of fever,

chills are common and child appears ill but when the fever breaks the child appears well [2]. The classical rash is rarely pruritic, evanescent (usually coming and going with fever spikes) and consist of discrete, circumscribed, salmon pink macules 2-10 mm in size. Lesions are more common on the trunk and proximal extremities. Stress, warm bath may exacerbate rash and koebner phenomenon is present. Its most diagnostic feature is its transient nature with a group of lesion usually lasting < 1 hour. Ten percent of patients may present with extra-articular manifestations only and may not develop arthritis for many months. Our patient classically fit in this description showing high grade fever with chills and rash, which coming and going with fever and not responding to antibiotics. She had history of arthralgia but no arthritis was evident. Laboratory finding in active SOJIA include anemia, leukocytosis, thrombocytosis, elevated liver enzymes, elevated acute phase reactants, ANA titer is rarely positive [2]. Our case also showed characteristics lab findings before development of complication possibly macrophage activation syndrome (MAS). The false positive serology of widal & dengue was also described in rheumatoid arthritis [3]. Most of deaths in SOJIA patients are secondary to MAS, infection resulting from immunosuppression, or cardiac complication [4]. MAS is a rare but life threatening complication of SOJIA which have preliminary diagnostic guideline of which our patient fulfilled 2 lab criteria of decreased platelet count and decreased white blood cell count along with all clinical criteria like Central nervous system dysfunction manifested by convulsion and lethargy, presence of purpura and hepatomegaly (> 3 cm below costal margin) [5]. She also had other abnormal laboratory findings like anemia, decreased erythrocyte sedimentation rate, decreased albumin, along with splenomegaly. Dilated cardiomyopathy was also described with MAS which rapidly resolved with steroid and supportive treatment along with renal involvement [6]. Our patient had dilated cardiomyopathy which also resolved rapidly but without any renal involvement. Treatment of MAS required pulse dose methylprednisolon and cyclosporine while refractory patient required dexamethasone and etopside [7-10]. But our patient responded to conservative management along with ventilator support. We presented this case as positive serology for infectious disease may mislead us from our diagnosis similarly any patient with prolonged fever we must rule out other noninfectious causes for it.

CONCLUSION

In any case of fever of unknown origin we should suspect connective tissue disorder as a possibility especially if not responding to antibiotics. Only positive serology for infective disease like typhoid or dengue shouldn't preclude us from possibility of noninfective etiology specially if blood culture is negative, as hypergamaglobulinemia lead to positive serology.

REFERENCES

1. Miller ML, Cassidy JT; Juvenile Rheumatoid Arthritis. In Kliegman RM, BehrmanRE, Jenson HB editors; Nelson Textbook of Pediatrics, 18th edition, Elsevir, Philadelphia, 2007: 1001-1011.
2. Weiss JE, Ilowite NT; Juvenile Idiopathic Arthritis. Pediatr Clin N Am., 2005; 52(2): 413-42
3. Buchy P, Yoksan S, Peeling RW, Hunsperger E; Laboratory tests for the diagnosis of dengue virus infection. In Working paper for the Scientific Working Group on Dengue Research. Geneva, Switzerland, 1-5 October, 2006: 74-84.
4. Wallace CA, Levinson JE; Juvenile rheumatoid arthritis: outcome and treatment for the 1990's. Rheum Dis Clin North Am 1991; 17(4):891-904.
5. Ravelli A, Magni-Manzoni S, Pistorio A, Besana C, Foti T, Ruperto N *et al*.; Preliminary diagnostic guidelines for macrophage activation syndrome complicating juvenile idiopathic arthritis. J Pediatr 2005; 146(5): 598-604.
6. Stephen JL, Kone – Paut I, Galambrun C, Mouy R, Bader- Meunier B, Prieur AM; Reactive hemophagocytic syndrome in children with inflammatory disorder: A retrospective study of 24 children. Rheumatology, 2001; 40: 1285-92
7. Sawhney S, Woo P, Murray KJ; Macrophage activation syndrome: a potentially fatal complication of rheumatic disorders. Arch Dis Child., 2001; 85(5): 421-426.
8. Henter JI, Aricò M, Egeler RM, Elinder G, Favara BE, Filipovich AH *et al*.; HLH-94: a treatment protocol for hemophagocytic lymhohistocytosis. HLH Study Group of the Histocyte Society. Med Pediatr Oncol., 1997; 28(5): 342-347.
9. Henter JI, Samuelsson-Horne A, Aricò M, Egeler RM, Elinder G, Filipovich AH *et al*.; Treatment of hemophagocytic lymphohistiocytosis with HLH-94 immunochemotherapy and bone marrow transplantation.. Blood, 2002; 100(7): 2367-2373.
10. Tristano AG; Macrophage Activation syndrome: A frequent but under –diagnosed complication associated with rheumatic diseases. Med Sci Monit., 2008; 14(3): RA27-36.