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Original Research Paper

Paediatrics

# A DIAGNOSTIC TWIST IN A PAINFUL JOINT

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**ABSTRACT** Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides are rare heterogeneous systemic diseases that usually occur in adulthood. They comprise granulomatosis with polyangiitis (GPA, Wegener's), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA, Churg-Strauss syndrome). We report a case, of a child presenting with features of juvenile idiopathic arthritis associated with chronic lung disease and ANCA positivity.

**KEYWORDS :** Antineutrophil cytoplasmatic antibodies( ANCA), microscopic polyangiitis (MPA)

## INTRODUCTION

Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides are rare heterogeneous systemic diseases that usually occur in adulthood. ANCA & Myeloperoxidase positive crescentic glomerulonephritis with vasculitides with haemorrhagic pneumatoceles is not reported yet. Here we present a case of Microscopic polyangiitic vasculitisis[1].

## CASE REPORT

A 5 year old asymptomatic girl presented with complaints of gradual onset, high grade and continuous fever occurring on and off over a period of one month. She had intermittent episodes of painful hip, knee, wrist and elbow swelling, erythema and painful restriction of movements lasting for weeks. She had six vasculitic nodules some with erythema some with ulceration as shown in the picture, they disappeared in 4-6 weeks.

On examination, child had high fever with BP 96/66 mm Hg and pulse 110 beats per minute. She was weighing 13 kg and was 100 cm in height (both less than third centile).

A provisional diagnosis of Juvenile Idiopathic Arthritis with systemic onset [SOJIA] was made and therapy was initiated with Tab Naproxen in a dose of 10mg/kg/day for six weeks and Methotrexate 12.5 mg per day and a blood transfusion was given in view of Hb 4.1 g% with MCV 56. Symptoms resolved in a week and she was discharged on oral iron supplementation. Nutritional anemia is very common in JIA due to hepcidin deficiency so it's easy to miss a unusual cause of anaemia with JIA [2].

After a month, she presented as anemia [Hb 3] in Congestive Cardiac Failure and was treated with diuretics, transfusion and oxygen. Meropenem and vancomycin started in view of counts 30000.

Anaemia did not improve despite transfusion and iron supplementation. Serum creatine was 1.1 and gradually rose to 2 then to 3, suggestive of pre renal failure. Both ESR and CRP were raised to 140mm and 104 units respectively. Renal involvement in a case of JIA is rare so always think for some other diagnosis if renal findings are present [3].

HRCT Thorax was suggestive of multiple foci of consolidation in bilateral lung parenchyma, with small hemorrhagic pneumatoceles in the posterior segment of right lower lobe and right sided pleural effusion suggestive of infective etiology for which antituberculous medications were started.

RA factor, ANA, DsDNA, ICT ,DCT,HIV, HBsAg and HCV were negative.
Kidney Biopsy showed pauci immune crescentric glomerulonephritis showing prominent changes of chronicity.

Immune vasculitis was considered as a probable diagnosis at this point and ANCA levels were tested which tested positive for p- ANCA and MPO suggestive of microscopic polyangiitic vasculitis.

Hence the diagnosis of p-ANCA vasculitides was confirmed. .The child however, gradually progressed to renal failure with rising BUN and creatinine, for Hypertension Nifedipine(0.2 mg/kg/dose) given. Around the twentieth day the child was also started on 2 mg/kg/day of Intavenous Cyclopho sphamide with oral Prednisolone in the dose of 2 mg/kg/day for 1 month. After 30 days of admission however the child progressed very rapidly to renal failure and succumbed to terminal pulmonary hemorrhage.

**TABLE1:LAB INVESTIGATIONS**

| | 1st oct 2017 | 17th oct 2017 | 30th Nov 2017 | 1st dec 2017 | 12th dec 2017 | 24th dec 2017 | 2nd jan 18 |
|---|---|---|---|---|---|---|---|
| **Hb(gm%)** | 4.1 | 6.8 | 3.1 | 6.3 | 9.5 | 7.4 | 8.1 |
| **TLC** | 11.8 | 12.3 | 30.8 | 22.7 | 24 | 15 | 25.8 |
| **Platelet** | 98 | 400 | 476 | 501 | 407 | 330 | 365 |
| **Na/K mg/dl** | 139/4.8 | | 143/5 | 140/3 | 137/4 | 135/6.5 | 137/7.1 |
| **BUN/Cr** | 30/6 | | 26/5 | 28/1.1 | 15/0.95 | 54/2.0 | 64/2.3 |
| **T. prot/ Albumin** | | | 7.7/2.6 | | 6.7/3.5 | 7.1/7.5 | |
| **ESR** | 110 | | | 130 | | | |
| **CRP** | | | | | | | |
| **C3 , C4** | | | | 134/3.9 | | | |

## DISCUSSION

Microscopic polyangiitic vasculitisis is an autoimmune disease characterised by systemic pauci-immune necrotising, small vessel vasculitis without clinical or pathological evidence of necrotizing granulomatous inflammation.

The presence of p-ANCA and MPO antibody are very helpful in making the diagnosis of MPA.

JIA is always a clinical diagnosis and investigations of JIA are only needed for prognosis of the case.

Many rheumaological conditions present as JIA and may evolve into extensive multisystem disease in the long run.

Anemia was consistently noted since first visit in this case which probably got neglected as nutritional anemia. Only when the renal dysfunction manifested; it was an eyeopener for a suspicion and evaluation as a vasculitic disease.

The goal of treatment is to prevent further progression and damage from the disease.

Most children do well with treatment but relapses can occur. Our patient had a major relapse with high grade fever, joint pain, swelling and respiratory distress and almost end stage renal failure. In addition to monitoring for disease relapses, these children also need to be monitored for side effects or complications from the treatment.

## CONCLUSION

It is very easily possible to miss this condition and consider it as juvenile idiopathic arthritis instead of a microscopic polyangiitis vasculitis. Whereas JIA is not life threatening, P-ANCA vasculitis can cause significant and rapid complications in a short span of time .whenever any unusual findings are associated with JIA like anemia, vasculitic or renal dysfunction; a broader diagnosis like MPA should be kept in mind.

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