VOLUME-7, ISSUE-11, NOVEMBER-2018 • PRINT ISSN No 2277 - 8160

Original Research Paper

General Medicine

AN INTERESTING CASE OF JUVENILE IDIOPATHIC ARTHRITIS.

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ABSTRACT

Juvenile idiopathic arthritis can be difficult to diagnose because there are no specific diagnostic tests and arthritis is often not evident early in the course. We present a case of systemic juvenile idiopathic arthritis involving multiple small and large joints along with flexion contractures. Investigations revealed iron deficiency anemia and a positive rheumatoid factor. Patient was started on DMARDs, corticosteroids and iron supplements.

KEYWORDS : rheumatoid arthritis, juvenile arthritis, anemia, autoimmune disease

INTRODUCTION: A 22 years old female was brought to Op with a primary complaint of puffiness of face and bilateral leg swelling – noticed since past 3 days. History of 1 episode of fever before 4 days, relieved with treatment. History of bleeding per rectum present since 5 days. There was no history of oliguria. No history of chestpain / orthopnoea / PND. No abdomen distention or jaundice or hematemesis. There was a past history of fever with arthritis at the age of 12 years following which she developed joint deformities in all four limbs. Patient has been bed ridden since then. H/o similar episodes were noticed in past in 2012 & 2017 for which she was given blood transfusions. Personal history revealed reduced appetite, normal sleep, normal bowel/bladder habits, no known addictions, no regular medications. Patient also did not attain menarche.

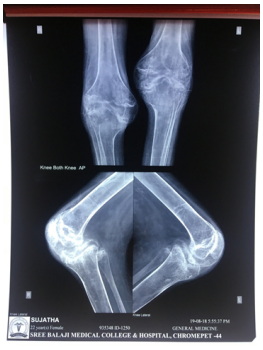
Clinical Examination

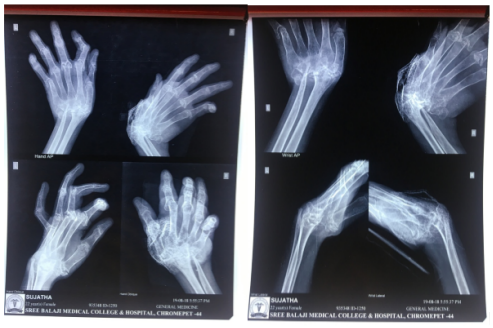
- On general examination, conscious, oriented, afebrile, pallor+, Anasarca+, had short stature, appeared undernourished, bed sores+ in gluteal region.
- There was no icterus, clubbing, cyanosis or lymphadenopathy.
- Small and large joint deformities (involving DIP, PIP, MCP & wrist, knees)with considerable restriction of movement with warmth and tenderness was present, flexion contractures+ in both knees and hips. Wind swept deformity+ , nodules+ in DIP joints.
- Heart rate was 112/min, SpO2 98% with ambient air, BP – 110/60 mmHg – Both arms, sitting position, r/r : 20/min.
- Respiratory examination was normal. Cardiovascular system examination revealed ejection systolic murmur grade-3 in pulmonary area, loud P2+.
- CNS examination did not reveal any dysfunstionL Higher mental functions were intact, motor power was 2/5 on all four limbs with no sensory deficit. The reduced power was attributable to the flexion contractures on large joints which rendered the patient bed ridden.

Investigations

- Hb.: 2.3 gm/dl
- PCV: 10.1%
- MCV: 56.4 fl
- MCH: 12.8 pg
- MCHC: 22.8%
- RBS: 84 mg/dl
- Serum Urea: 24 mg/dl
- Serum Creatinine: 0.3 mg/dl
- Serum Uric Acid: 5.6 mg/dl
- Serum Sodium: 123.1 mEq/L
- Serum Potassium: 4.89 mEq/L
- Chloride: 103.6 mEq/L
- Total bilirubin: 0.2mg/dl
- Direct bilirubin: 0.1 mg/dl
- AST(SGOT): 6 IU/L
- ALT(SGPT): 7 IU/L
- Alkaline phosphatase: 143 IU/L
- Total protein: 4.5 gm/dl
- Albumin: 1.8 gm/dl
- GGTP: 11 IU/L
- RBCs: 1.79 millions/cmm
- Platelet count : 9.86 lakh/cumm
- USG abdomen revealed a hypoplastic uterus measuring ~4.0 x 1.9 x 1.1, anteverted.
- Right and left ovaries measuring ~2.0x0.8 and 2.2x1.2 respectively.
- Normal liver (12.8cm), gall bladder, spleen (9cm), pancreas and KUB.
- Rheumatoid factor : 320 Iu/ml
- Peripheral smear study showed a decreased RBC mass, microcytic hypochromic RBC with moderate anisopoidilocytosis showing elongated cells, pencil shaped cells, fragmented RBCs, and presence of tear drop cells with occasional macrocytes. Increased WBCs with normal morphology and neutrophilic preponderance. Increased platelets with few giant platelets. No blast cells seen.
- Viral markers were negative.

2D ECHO showed a normal LV function and no regional wall motion abnormalities.





Differential diagnosis

- Infections : bacterial/viral
- septic arthritis & osteomyelitis
- Postinfectious and viral arthritis : Parvovirus B19.
- Reactive arthritis – streptococcal & neisserial infections.
- Subacute bacterial endocarditis
- Autoimmune and autoinflammatory:
- IBD
- Kawasaki disease
- SLE
- Mixed connective tissue disease
- Dermatomyositis
- Sarcoidosis

Diagnosis

The presence of a positive Rheumatoid factor and symmetrical involvement of joints was suggestive of juvenile idiopathic arthritis. Complete blood count and peripheral smear study was suggestive of iron deficiency anemia with failure probably due to per rectal bleeding. **S**ystemic juvenile idiopathic arthritis can be difficult to diagnose because there are no specific diagnostic tests, and arthritis is often not evident early in the course of the disease.[1]

Treatment

Patient was treated with multiple PCV transfusions. She was also started on Methotrexate, hydroxychloroquine and prednisolone. Symptoms due to anemia were improved and patient is on regular follow-up.

Clinical manifestations:

Arthralgias are common early in the course of systemic JIA, but arthritis is not always prominent. Unlike the oligoarticular and polyarticular subtypes of JIA, the arthritis of sJIA may begin in the hips and may progress very rapidly, causing severe damage and dysfunction and need for early joint replacement surgery, as well as loss of growth potential in younger patients. **S**ynovial inflammation is the result of a disturbed balance between proinflammatory effector cells such as T-helper-17 cells, and anti-inflammatory regulatory cells such as FOXP3-positive regulatory T cells.[2]

Extra-articular manifestations include fever and macular rash (most common). Hepatomegaly, splenomegaly and lymphadenopathy are also relatively common. Pericarditis and other forms of serositis are also a feature of sJIA. Pulmonary manifestations include pleural effusion, Interstitial lung disease, alveolar proteinosis, pulmonary hypertension are rare.

DISCUSSION

Juvenile idiopathic arthritis is a heterogenenous group of diseases characterized by arthritis of unknown origin with onset before age of 16 years. Pivotal studies in the past 5 years have led to substantial progress in various areas, from disease classification to new treatments. Age at onset may be an important parameter to consider in JIA classification[4.] Different immune mechanisms in distinct subtypes of the disease and can be helpful to redefine disease classification criteria.[2] There is a need to identify early predictors of outcome, to further improve therapy, and to continue long-term follow-up of patients with JIA.[5]

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