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JUVENILE POLYARTERITIS NODOSA: AN EARLY PRESENTATION

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ABSTRACT Polyarteritis nodosa is a primary systemic necrotizing vasculitis predominantly of medium sized arteries leading to microaneurysms, rupture and haemorrhages, manifesting with multisystem involvement. Childhood presentation is rare and very few cases are reported in India(1,2,3).Approximately 45-100% of patient with PAN exhibit cutaneous signs(4). We describe a case of childhood PAN with early onset and on presentation appeared to be having a similar presentation as Systemic onset JIA . It was diagnosed to be a case of Cutaneous PAN ,had an aggressive course but responded well to conventional treatment.

KEYWORDS : livedo reticularis, digital gangrene ,arthritis, vasculitis, JIA

INTRODUCTION

Vasculitis is defined as the presence of inflammation in a blood vessel that may occur as a primary process or secondary to an underlying disease. Primary vasculitides are rare in children. They are classified by the size of vessels involved and the type of inflammatory response. Polyarteritis nodosa is a rare systemic vasculitis characterized by necrotizing arteritis of predominantly medium sized vessels leading to microaneurysms,rupture and haemorrhages, manifesting with multisystem involvement(3,5). It was first described by Kussmal and Maier in 1866(16). The term PAN was coined by Ferrari in 1903 after he recognized this as transmural inflammation(4,16). It has an estimated incidence of 2-9.0/million in adults(6) There are very few reported cases of childhood PAN(17)

But more common presentation seen in 7-11 years(17) . males are more commonly affected(3,7). There are three forms- infantile, cutaneous, systemic. Infantile type has similar picture as severe form of Kawasaki disease. Cutaneous PAN is characterized by skin involvement with no visceral involvement although musculoskeletal system involvement is common,Peripheral nervous system may be involved rarely(15) Fever, rash, musculoskeletal symptoms are common in children. Any evidence of visceral involvement either clinically (central nervous system,pulmonary, cardiac, gastrointestinal or renal), radiographically (abnormal angiography) or by histology (visceral biopsy) were said to be systemic PAN(15)

The characteristic histologic changes of PAN are fibrinoid necrosis of the walls of medium or small arteries with a marked inflammatory response within or surrounding the vessel wall(17). We present a case of childhood cutaneous PAN with early onset and on presentation appeared to be having a similar presentation as Systemic onset JIA . It had an aggressive course but responded well to conventional treatment with steroids and immunosuppressant.

CASE REPORT

A 4 year old male child presented with high grade fever,continuous type ,multiple swellings all over the body, intermittent pain abdomen , swelling of joints of both upper and lower limbs (particularly large joints) with pain,restriction of joint movement, bluish-black pigmented patches over skin(more over lower limbs) for 2 months. There was no history of altered sensorium, seizures, vomiting, loose stools, jaundice,respiratory distress,contact with TB,bleeding from any orifices, any discoloration of urine, burning micturition or decreased urination. Past medical history was unremarkable. Child was borne out of a non-consanguinous marriage , was neuro developmentally normal and immunized as per NIS belonged to a lower socioeconomic strata . family history was not singnificant. On examination, patient was found to be pale, pitting pedal edema was present bilaterally with swelling of ankle and knee joints on both sides and bilateral cervical and right axillary lymphadenopathy was present.

Blood counts showed neutrophilic leucocytosis(50,910/cumm, neutrophils-76%) ,thrombocytosis (9,00,000/cumm)and microcytic hypochromic anemia(Hb-8.4) , renal function tests(S.urea-28mg/dl, S.creatinine-0.8mg/dl, S.sodium-135 meq/L,S.potassium-4.3meq/L) biochemical tests was normal.ESR was raised (132mm),CRP was positive. Chest XRay showed right paracardiac shadow. XRay of pelvis was normal. ANA and Anti-CCP was negative. Blood culture showed growth of Staphylococcus aureus. ASO titre was raised. RA factor, MPICT was negative. Sickling was negative. Bone Marrow study revealed marrow infection. Doppler USG Abdomen was normal. Patient was treated as a case of Systemic onset JIA with multiple pyemic abscess . The treatment given was Tab Naproxen and broad spectrum antibiotics- Inj ceftriaxone, Inj Piperacillin-tazobactam, Inj vancomycin for 14days. There was persistence of symptoms so oral Methotrexate was started. Fever subsided , joint swelling slightly reduced and on day 14 the patient was discharged on oral naproxen and methotrexate.

On follow up after 1 month, there was aggravation of symptoms with onset of gangrene of ring finger of right hand and big toe of right foot. Blood counts Liver function tests, Renal function tests was normal. Eye examination was normal. CT abdominal Angiography and aortography was normal.2D echocardiography was normal. The patient was suspected to be a case of juvenile polyarteritis nodosa and started on tab prednisolone at 1 mg/kg/day and tab azathioprine at 3mg/kg/day . The patient recovered and was discharged on steroid and azathioprine therapy.

Figures-Gangrene Of Fingers With Purpuric Patches On Lower Limbs

DISCUSSION

Polyarteritis nodosa is a rare vasculitis of childhood that Causes necrotising inflammation of small and medium sized arteries in multiple systems with predilection for visceral arteries(1,3).

EULAR/PRINTO/PRES definition of childhood-PAN includes Histopathology or angiographic abnormalities (mandatory) plus one of the following five criteria –skin involvement, myalgia or muscle tenderness, hypertension, peripheral neuropathy, renal involvement(19). It can occur at any age ,two peaks have been seen from previous studies one at 5-7 years other at 11-13 years(3,7) Notably, the disease varies in its presentation from a relatively benign cutaneous form, which may resolve without treatment, to a severe systemic form.(3)

Fever is the most common constitutional symptom in childhood PAN Articular involvement in terms of arthralgia and or arthritis had been reported among 43-80% of childhood PAN ,previous literatures did not show any correlation between ASLO titer and articular manifestation in childhood PAN. (3) Myalgia had been reported variably in 33-100% children (4,17).Skin involvement includes tender subcutaneous nodules,purpuric skin rashes, livedo reticularis(purplish reticular pattern irregularly distributed around subcutaneous fat lobules which becomes more prominent on cooling),skin ulcerations, infarctions-digital pulp necrosis,nailbed infarctions,splinter haemorrhages, digital phalanx or peripheral tissue(ear,tip of nose) necrosis and gangrene.

Earlier studies had variable skin manifestations involving 45-100% of children diagnosed with childhood PAN. But, detailed manifestation of the skin involvement was rarely being published except very few studies. Though previous studies had no mention about the incidence of autoamputation, there are case series reporting autoamputation in their patients . the study by Shivappa et al reported an incidence of amputation in 27% of their patients(4). we had observed autoamputation involving the distal phalanges of fingers and and toes in our patient. In contrast to anti neutrophil cytoplasm antibodies associated vasculitis, lung and glomerular involvement are distinctly uncommon in PAN. Renal involvement is characterized by proteinuria, haematuria, decreased GFR. Peripheral neuropathy may cause glove and stocking neuropathy or mononeuritis multiplex.

Definitive diagnosis is by histopathologic evidence of necrotizing inflammation of medium and small sized arteries. Most children respond to corticosteroids. Penicillin to be used in case of raised ASO titres(8,9,10).Other drugs with good results are low dose methotrexate, cyclophosphamide, IV immunoglobulin and biologics(9,10) Very few cases of cutaneous PAN evolve into systemic PAN(17,18).

This patient had involvement of skin in the form of livedo reticularis,purpura and ecchymotic patches ,ulceration of extremities and musculoskeletal system involvement in the form of polyarthritis .

Initially this patient was evaluated as a case of Suspected Haemophilia, the results were normal. There was no history of trauma. Other conditions with similar presentation would be Systemic lupus erythematosus, leukemia,Acute rheumatic fever.These were excluded by history and biochemical examinations.

Then it was suspected to be a case of Systemic onset JIA although the rash was not typically evanescent and there was no hepa tosplenomegaly. But the patient did not respond to the therapy with NSAID. The patient deteriorated and developed gangrene of extremities which led to autoamputation of the gangrenous part.Other causes of gangrene in children are infective endocarditis, hypercoagulable states, Scleroderma, drugs .Children with Sickle cell disease have been reported to develop gangrene(13). No pertinent history and investigations relating to these illnesses were found in our patient.

So this patient was diagnosed to be a case of childhood PAN cutaneous type. Cutaneous PAN presenting with recurrent gangrene associated with Streptococcal infection has been described(14). Prednisolone and methotrexate was started and the patient showed significant improvement. On follow up after 1 month the patient was doing well, afebrile , vitals being stable, no new onset ulceration,rash or arthritis was seen. The same treatment was continued with advice to follow up every month with a routine blood count test.

Despite being a rare disease , PAN should be included as a differential diagnosis in children with rash, ecchymosis, arthritis, unexplained fever to enable early diagnosis and proper management which can lead to reduced morbidity and mortality.

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