Systemic-Onset Juvenile Idiopathic Arthritis with unusual Cutaneous Manifestation and peripheral eosinophilia: Case Report

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AbstractIntroduction: Systemic-onset juvenile idiopathic arthritis (SoJIA) is unique subtype of juvenile idiopathic arthritis (JIA) with very special clinical manifestations, complications and management options. The simultaneous presentation of tinea capitis has not been reported in the context of Systemic-Onset Juvenile Idiopathic Arthritis before. *The case:* in march 2021 a 5-years old Sudanese male presented to Ahmed Gasim Hospital with fever and bilateral ankle arthritis in a background of extensive scalp lesions which were scaly, itchy and associated with hair loss. On examination: his weight was 15 kg (on the 5thcentile). There was cervical lymphadenopathy, hepatomegaly and swelling and tenderness in both ankle joints with restriction of movements. Complete blood counts revealed leucocytosis, thrombocytosis, mild eosinophilia and microcytic hypochromic anaemia. Anti-dsDNA antibody was 45 IU/ml (positive), ANA profile was 0,8 Ratio (Equivocal), CRP was 34.4 mg/l, HDL was very high, these results support the diagnosis of SoJIA in a background of a kerion. Patient received: antibiotic, systemic antifungal, Corticosteroids, Hydroxychloroquine, Calcium and Vitamin D to which he achieved good results.**Consent:**Witten informed consent was obtained from patient's parent to publish this report in accordance with the journal's patient consent policy.**Keywords**: Systemic-onset juvenile idiopathic arthritis; Systemic juvenile idiopathic arthritis; Tinea capitis; eosinophilia; case report.**Introduction**Juvenile idiopathic arthritis is a group of arthritis that occur before 16 years and last more than 6 weeks after exclusion of other aetiologies which is classified according to the International league of Associations for Rheumatology (ILAR) into 3 subtypes according to the clinical manifestations, complications and therapeutic options.(1–3) Systemic-onset juvenile idiopathic arthritis (SoJIA) is a very special subtype of juvenile idiopathic arthritis (JIA) that is characterized by fever which has a characteristic one or two spikes (>39°C) per day.(4) Also more than 80% of patients of this disease has a transient salmon-coloured macular or maculopapular rash that that accompanies the fever. They may have also myalgias and tenosynovitis and arthritis which may be oligoarticular to polyarticular.(3) The widely affected joints include wrists, knee, and ankles; but any joint can be affected, even the temporomandibular joints(5), cervical spine(6), hips(7), and the small joints of the hand and feet. SoJIA may present as painless lymphadenopathy (25%) hepatomegaly, splenomegaly or pericarditis which may be complicated by cardiac tamponade (3,4) There is no specific laboratory features that distinguishes SoJIA from other conditions, but the pattern of laboratory abnormalities may support the diagnosis, for examples; microcytic hypochromic anaemia, neutrophilic leucocytosis, thrombocytosis, high ESR, high CRP, high serum ferritin, low serum

albumin, mildly elevated AST, high D-dimer and negative autoantibodies.(3) The treatment of JIA as whole focuses on suppressing inflammation, preserving functions, and preventing deformity and blindness.(8) the currently available drugs include nonsteroidal anti-inflammatory drugs (NSAID), Systemic corticosteroids and Disease-modifying anti-rheumatic drugs (DMARDs).(3,9,10) Eosinophil cells were associated with allergic bronchopulmonary aspergillosis and asthma exacerbation due to fungal antigens.(11) Moreover, a case report of kerion associated eosinophilia was described in which a dermatophytid reaction was thought to be the cause which responded to corticosteroid and antifungal.(12) 2 Case reports of 2 patients with systemic lupus erythromatosis in which tinea capitis was disseminated, one of them was using steroid the other was not.(13,14) and this is the first report related to juvenile idiopathic arthritis.**Case reportHistory** In March 2021; a 5-years old Sudanese male from Algazira, center of Sudan presented with bilateral ankle swelling, bilateral knee and hip pain and fever which started 1 month prior to presentation. His mother was also concerned about scales and hair loss all over the scalp which started 2 months prior to presentation. His symptoms started insidiously with the fever mainly at night and relieved by antipyretics. He had anorexia and weight loss but neither abdominal pain, vomiting, diarrhea, cough, upper respiratory tract symptoms, headache nor history of trauma. He had a past history of right knee swelling which was resolved spontaneously. He had no family history of autoimmune disease or malignancy and not exposed to any medications.**Examination**On examination the patient was ill but not pale, jaundiced or cyanosed. His weight was 15 Kg (along the 5^{th} centile) (fig. 1). The scalp had white-yellow scales with some swelling, hair loss and dried pus over some areas of the scalp (Fig. 2). There were Bilateral cervical lymphadenopathy which were non-tender, discrete and maximum diameter was 1×1 cm. Both ankle joints were Swollen (fig. 3),and tender to palpation and there was restriction of movements. Other joint examinations were normal. Abdominal examination revealed palpable liver which was 4 cm below the costal margin and palpable paraaortic lymph nodes.**Investigations**Complete blood count and peripheral blood picture revealed leucocytosis (WBCS=13.4*10^3 which is high), neutrophil% = 49%, lymphocyte% = 38%, monocyte% = 5% and eosinophil% = 8% (mild eosinophilia) Thrombocytosis (614*10^3) and mild hypochromia (Hb=10.3 g/dl, MCV=71.8 fl, MCH=22.3 pg, MCHC=31.1 g/dl) were also present. Anti-dsDNA antibody was 45 IU/ml (positive). ANA profile was 0.8 Ratio (Equivocal). CRP was 34.4 mg/l (high) and ESR = 49 mm/hr. LDH was very high. Patient was treated with: Methylprednisolone 30mg/kg/day for 3 days. Followed by oral prednisone 1mg/kg/day. Hydroxychloroquine tabs 5mg/kg/day. Griseofulvin syrup 7.3mg/kg/day. Antibiotic Calcium and vitamin D. After 1 month of treatment follow-up the patient was improved and we refer him to the ophthalmologist for slit lamp examination.**Discussion** In this case the full detailed history and proper clinical examination in addition to laboratory findings are in favour of the diagnosis of systemic-onset juvenile idiopathic arthritis. This case may show that, musculoskeletal manifestations may be present earlier than any other symptoms. Also, we noticed that in the lipid profile the HDL was very high but serum ferritin and ESR was normal, a complicating macrophage activation syndrome was putted in mind and then excluded.(15,16) Does tinea capitis occur more extensively in this patient due to the disease itself we don't know fully but in a previous case report to a patient with systemic lupus erythromatosis in which disseminated infection occurred simultaneously at the time of the diagnosis before even the use of corticosteroid.(13) in that case the causative agent was Microsporum gypseum but unfortunately in our case the diagnosis was made clinically only and microbiological consultation was not ordered due to financial problems but what make our case unique is its association with systemic-onset juvenile idiopathic arthritis and not Systemic lupus erythromatosis even before the start of immunosuppressive therapy.(13,14) Mild peripheral eosinophilia explanation in our case was challenging; whether it is related to a dermatophytid reaction with no obvious morbilliform or lichenoid lesions or due to other causes that are not typical with the present history such as drug tubulointerstitial nephritis or any other occult helminthic infection. But according to the other laboratory and history points the later causes were excluded and we left with the occult dermatophytid reaction which was consistent with the same observation from another case report of kerion due to T. tonsurans with 21% eosinophil in the complete which was reduced to 6% one month later after oral griseofulvin and corticosteroid were used.(12) but unlike that case in which the patient was 45 year old female with a clear medical background unlike our case. But this may point to the fact that complete blood count is not routinely ordered in tinea capitis and eosinophilia may be underreported if we associate this to the fact that eosinophil recognizes ß-glucan of the fungal cell wall and react to it by releasing

its granules and this area is an area of investigation in the future.(11)**Conclusion:**Physicians should be alert to the presentation of systemic-onset JIA in our country in order to make prompt diagnosis and treatment decisions as early as possible. Careful follow-up of prolonged febrile patients with arthritis of unknown origin is important to reaching the diagnosis early and initiating treatment.**Conflict of interest:**NO conflict of interest.**Recommendations:**

1. Further studies about the eosinophil count and its role in tinea capitis and systemic-onset juvenile idiopathic arthritis.
2. Further studies about the immune response against fungi in the setting of juvenile idiopathic arthritis.
3. Multidisciplinary team consultation (rheumatologist, ophthalmologist, orthopaedics and paediatrician) in case of SoJIA.
4. Educate the patients about the disease and its complications, which are important to monitor the disease and long-term morbidity and mortality.

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