SAT0507

CLINICAL CHARACTERISTICS AND TREATMENT RESPONSE IN JUVENILE GOUT PATIENTS: A SINGLE CENTER EXPERIENCE

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Background: The incidence of juvenile gout is increasing in China. The clinical manifestations of juvenile gout and treatment strategies to reduce uric acid levels in children are not well described due to the limited number of cases in the past. **Objectives:** We aim to describe the clinical characteristic of children with gout and study the treatment response to febuxostat.

Methods: These studies were approved by the Institutional Review Board of Guangdong Second provincial General Hospital. We performed a retrospective analysis on 98 juvenile gout patients (age ≤ 18 years) evaluated in our hospital from Jan 2016 to Dec 2019. We analyzed clinical parameters, laboratory data and treatment response.

Results: The average age of disease onset in children with gout was 15.2 ± 2.0 years and the youngest patient was 9 years old. The majority of patients were male (94/98) and mean serum uric acid (sUA) level were 705.8 ± 145.7 $\mu\text{mol/L}$ (reference range <420 $\mu\text{mol/L}$). More than half of the cohort had normal body mass index (mean 24.7 ± 4.7 kg/m^2 ; range 14.9 to 36.1 kg/m^2). Renal function was generally normal in these children (serum creatinine 96.9 ± 17.8 $\mu\text{mol/L}$). In terms of joint manifestations, juvenile gout preferentially affected finger joints (29%), ankles (28%) and metatarsal joints (MTP; 20%). The most frequent sites of initial gout attack were ankles (45%), MTP (39%) and fingers (6%). In addition, tophi can occur in pediatric patients and typically develop in the finger joints (54%). Tophi was observed in about 25% of juvenile gout patients, typically within the first two years of disease onset (mean duration 1.7 ± 0.9 years). We have found tophi in children as young as 10 years of age.

For treatment for chronic hyperuricemia, 32 patients (32.7%) were started on febuxostat and 5 patients (5.1%) received allopurinol. A decrease in sUA was observed in both groups after the first month of treatment (febuxostat: baseline 690.4 ± 99.7 $\mu\text{mol/L}$ to 482.7 ± 140.8 $\mu\text{mol/L}$ vs. allopurinol: baseline 728.8 ± 112.8 $\mu\text{mol/L}$ to 565.0 ± 116.7 $\mu\text{mol/L}$, $P=0.477$). Serum uric acid of 6 patients in the febuxostat group (none in the allopurinol group) dropped below 360 $\mu\text{mol/L}$. There were no statistical differences in Cr, AST and ALT between the groups. During follow-up after 3 months, further decline in sUA level were observed in patients treated with febuxostat (409.5 ± 83.4 , compared with baseline $P<0.001$).

Conclusion: Juvenile gout has a different pattern of joint involvement and is less associated with elevated BMI compared to gout in adults. We show that febuxostat is effective in reducing uric acid levels in juvenile gout. These findings will help clinicians better understand the clinical manifestations and treatment response in juvenile gout.

Figure 1 Compared treatment response with allopurinol and febuxostat

References:

- [1] Kishimoto K, Kobayashi R, Hori D, et al. Febuxostat as a Prophylaxis for Tumor Lysis Syndrome in Children with Hematological Malignancies. *Anticancer Res.* 2017 Oct;37(10):5845-5849.
- [2] Lu, C.C., et al. Clinical characteristics of and relationship between metabolic components and renal function among patients with early-onset juvenile tophaceous gout. *J Rheumatol.* 2014. 41(9): p. 1878-83.

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Other orphan diseases

SAT0508

DIAGNOSIS OF BEHÇET'S DISEASE: COMPARISON OF TWO SETS OF CLASSIFICATION CRITERIA. APPLICATION IN 111 PATIENTS OF A WELL-DEFINED POPULATION

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Background: Behçet's disease (BD) is a systemic, chronic, relapsing vasculitis with no pathognomonic diagnostic test. The most widely used classification criteria are those of the International Study Group (ISG) for BD (1). These criteria were repeatedly found to have low sensitivity. Therefore, the International Criteria for Behçet's Disease (ICBD) were published in 2014 (2).

Objectives: To compare the ISG with ICBD diagnostic criteria for BD.

Methods: The study included all consecutive 111 patients diagnosed with definitive or possible BD by expert rheumatologists. They were diagnosed at a well-defined population in Northern Spain between 1980 and 2019. The ISG and ICBD diagnostic criteria for BD were applied to and compared among all patients.

Results: We studied 111 patients (62 Women/49 Men), mean age 36.8 ± 13.2 years. BD was diagnosed in 65 (58.5%) by ISGBD criteria and in 86 (77.5%) by ICBD criteria. No significant differences were observed between both criteria ($p < 0.001$). The overall concordance was fair (Kappa 0.3; $p < 0.001$). Sensitivity was 58.6% for ICBD criteria and 80.2% for ISG. **(TABLE-1)**

Conclusion: ICBD criteria exhibit higher sensitivity than ISG criteria. Thus, the application of these new criteria can achieve a more correct and earlier diagnosis of BD.

References:

- [1] Criteria for diagnosis of Behçet's disease, International Study Group for Behçet's Disease, The Lancet, Volume 335, Issue 8697, 1078 – 1080
- [2] The International Criteria for Behçet's Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria, *J Eur Acad Dermatol Venereol.* 2014; 28:338-47

TABLE 1

	Expert diagnosis (N=111)	ISG criteria (N=65)	ICBD criteria (N=86)
Age, mean (SD)	36.7 (13.2)	36 (12.8)	36.7 (13)
Gender, men/women, N (%)	49/62 (44.1/55.8)	29/36 (44.6/55.4)	38/48 (44.2/55.8)
Oral aphthosis	110 (99)	65 (100)	85 (100)
-Recurrent (3 times/year)	91 (87.2)	61 (93.8)	74 (86)
Genital aphthosis	59 (53.1)	42 (64.6)	56 (65.1)
Skin manifestations	76 (68.4)	56 (86.15)	71 (70.9)
-Pseudofolliculitis/ Erythema nodosum	51 (67.1)/ 27 (35.5)	38 (58.5)/ 21 (32.3)	42 (68.8)/ 22 (36.1)
Ocular lesions	39 (35.1)	32 (49.2)	39 (45.3)
-Anterior/ Posterior/ Panuveitis	17 (43.6); 12 (30.8)/ 0	16 (50)/ 8(25)/ 7 (21.9)	17 (45.6); 0; 12 (30.8)
-Retinal vasculitis	4 (10.3)	1 (3.1)	4 (10.6)
Joint manifestations	76 (68.5)	43 (66.1)	58 (67.4)
-Arthralgias / Arthritis	69 (92.8)/ 45 (60)	39 (90.7)/ 24 (55.8)	52 (89.6)/ 33 (56.9)
Neurological manifestations	20 (18)	11 (16.9)	16 (18.6)
-Peripheral / Central	11 (55)/ 14 (70)	7 (63.6)/ 7 (63.6)	12 (75)/ 10 (62.5)
Vascular manifestations	9 (8.6)	7 (10.8)	10 (11.6)
-Arterial/ Vein thrombosis/ Phlebitis	0/ 5 (55)/ 1 (11.1)	0/ 4 (57.1)/ 1 (14.3)	1 (12.5)/ 5 (62.5)/ 0
Gastrointestinal features	4 (4.5)	4 (6.1)	4 (4.6)
Pathergy test positive (available data; %)	6 (28; 21.4)	4 (19; 21)	4 (25, 16)
HLA B51 positive (available data; %)	38 (86; 44.2)	19 (47; 40.4)	28 (63; 44.4)

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SAT0509

MYCOPHENOLATE MOFETIL VERSUS AZATHIOPURINE FOR THE MAINTENANCE TREATMENT OF CONNECTIVE-TISSUE RELATED INTERSTITIAL LUNG DISEASE FOLLOWING CYCLOPHOSPHAMIDE TREATMENT

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