She is the first born of a 51-year-old father and 40-year-old mother, non-consanguineous parents, at full term, and has a history of early hypotonia, psychomotor developmental delay and failure to thrive. Physical exam revealed arthritis of both ankles, the right knee and the MCP joint of the left thumb. We noted loose joints, high-arched palate and facial dysmorphism (hypertelorism, large protruding ears, flat nose and short philtrum). Body weight and height were at -3SD. Intellectual disability was mild.

Laboratory tests showed slightly increased ESR (35mm at first hour) with positive ANA (>1/1000) and negative RF and anti-CCP. Karyotype is 46XX with deletion of the short arm of chromosome 18. Both parents had normal Karyotypes. MRI of the brain showed cortical atrophy and periventricular leukomalacia.

Ophthalmic screening detected bilateral anterior uveitis.

The patient was treated by AINS and methotrexate along with topical ophthalmic steroids. 6 months after treatment, the child had complete regression of uveitis and arthritis.

Results: This case of de-novo 18p deletion is associated to oligoarticular JIA with uveitis. According to our knowledge, there have only been 2 published cases of juvenile inflammatory arthritis with 18p deletion although 5 other cases were reported in association to other chromosome 18 abnormalities (deletion of long arm and ring). Few reports of adult-onset arthritis with monosomy 18p are available.

The exact explanation of this association is still unclear. As gene loss is a major factor in the pathogenesis of inflammation, this presentation might be related to deletion of PTPN2, a gene that has been associated to rheumatoid arthritis. PTPN2 encodes Protein Tyrosine Phosphatase Non-receptor type 2 and is located in the short arm of chromosome 18. PTPN2-deficiency stimulates T follicular helper cell and B cell responses thus promoting autoimmune reaction. Of the 67 genes in 18p, only around 12 (of which PTPN-2) are responsible of a phenotype when hemizygous. Further genetic studies are needed to better elucidate the role of deleted genes in the pathogenesis of inflammation in monosomy 18p.

Conclusion: In addition to skeletal deformities such as kyphosis and feet deformities, children with 18p deletion should be screened for inflammatory arthritis.

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AB0957

ATYPICAL PRESENTATION AND EARLY RECURRENCE OF KAWASAKI DISEASE IN A FEMALE INFANT: CASE REPORT

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Background: Kawasaki Disease (KD) is the second most common vasculitis in the pediatric population and the leading cause for pediatric acquired heart disease in developed countries. It is commonly diagnosed in the Mexican pediatric population, epidemiology in this country is not established, since cases are not usually reported to the healthcare system. The clinical features are quite variable, but diagnosis and prompt treatment will decrease morbidity and mortality. Coronary artery aneurysms are the most common complication, which represent the leading cause of acute coronary syndrome before 40 years of age. Recurrence of KD is estimated to be around 3% in Japanese patients and 1% in the United States, nevertheless, this data in Latin-American children is unknown. It usually affects patients before reaching 3 years old and within 2 years of

the initial attack, presenting with an abrupt onset and higher complication rates, requiring aggressive workup, treatment and follow-up.

Objectives: To review an atypical presentation of KD and early recurrence in a 7-month-old female.

Methods: We present a 7-month-old female diagnosed with KD at 48 days old. She presented to the emergency unit with irritability, high and persistent fever and acholia. During workup, she was found with cholestasis and gallbladder hydrops. Negative CSF, blood and urine cultures were documented during hospitalization, and fulfilled the KD criteria. The cardiac ultrasound revealed coronary abnormalities: a RCA of 2.8mm (Z-Score +4.79), LCA of 3.3mm (Z-Score +5.69) and LAD of 2.4mm (Z-Score +4.25), which fit into classification 3 and 4, as small and medium aneurysms, according to AHA 2017. Immunoglobulin (2g/kg) and aspirin (80mg/kg) were administered and she was discharged 36 hours after the IVIg infusion, afebrile and with ambulatory follow-up, 30 days later the coronary abnormalities showed RCA of 2.0mm (Z-Score +2.3), LCA of 3.0mm (Z-Score +6), LAD 2.3mm (Z-Score +2.9). Six months later, she presented fever for 6 days, irritability and polymorphous rash. On physical exam BCGitis, non-suppurative conjunctivitis and pallor in hands and feet, elevated CRP and ESR, leukocytosis, thrombocytosis and sterile leukocyturia. Echocardiography reported RCA of 2.4mm (Z-Score +4.8), LCA of 3.1 (Z-Score +4.2) and LAD of 2.3mm (Z-Score +3.2), diagnosing KD recurrence, admitting the patient for IVIg and aspirin administration.

Results: The patient was treated with IVIg and aspirin. Follow-up by Cardiology determined improvement of Z-Scores. Recurrence occurred with worsening of the cardiac abnormalities. Cardiac prognosis is importantly affected due to the atypical age, vascular abnormalities and repeated vasculitic process. Rheumatologic consult should be considered since disease like Takayasu Arteritis, Polyarteritis Nosa and ADA2 deficiency need to be ruled out.

Conclusion: KD needs prompt diagnosis and treatment due to the potential consequences when delayed. Clinical suspicion is important due to the possible atypical presentation. As with this patient, age, gender and presentation are not exclusive. Despite adequate treatment, recurrence and worsening of the cardiac abnormalities occurred. Both KD events before 1 year old and with atypical presentations. Rheumatologic and cardiac follow-up need to be stringent through lifetime, to determine pharmacologic treatment, as well as physical activity and reproductive counseling.

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AB0958

PEDIATRIC BEHCET'S DISEASE WITH SINUS VENOUS THROMBOSIS: THREE CENTER EXPERIENCE FROM TURKEY

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Background: Behçet's disease (BD) is an autoinflammatory disorder which may present with multi-systemic involvements including the vascular, cutaneous, articular, gastrointestinal, and/or central nervous systems (CNS). CNS involvement appears in two main groups: parenchymal or non-parenchymal. Neurological symptoms in children and adolescents can be confused with many other disorders and may be the initial symptom of BD.

Objectives: To report our experiences of the juvenile Behçet's Disease patients with cerebral venous sinus thrombosis (CVST) and to review previous studies reporting the clinical characteristics and outcomes of Juvenile Behçet's Disease with CVST.

Methods: The patients who met Pediatric Behçet's Disease (PEDBD) classification criteria for juvenile Behçet's Disease from 3 referral centers in Turkey were reviewed retrospectively. Disease activity was assessed by BD current activity form (BDCAF). A systematic review of literature of all published data was conducted.