BEHÇET’S DISEASE IN CHILDHOOD. A CASE REPORT

Diego Ruben Salinas Encinas1, and Ingrid Vianey Vidal Huarchi1. 1Caja Nacional De Salud, Santa Cruz De La Sierra, Bolivia.

Objectives: To describe a case of recurrent aphthous stomatitis, genital ulceration, and uveitis, compatible with Behçet’s disease (BD) and be aware of its complications

BD, a systemic vasculitis of unknown etiology, is characterized by recurrent attacks of acute inflammation. The frequency and duration of these outbreaks are unpredictable. Diagnosis and disease monitoring require clinical awareness. Although the usual onset of disease is between the second and fourth decade, there has been an increased awareness of BD during childhood. BD is very prevalent in Mediterranean countries and the incidence is rare in the West. Therefore, we present a female patient with BD.

Methods: 12-year-old female patient with no previous history. Referred 15 months of the presence of a papulopustular lesions with erythematous halo on the scalp, thorax and back which recurred at various intervals with periods of worsening and improvement. Had received multiple topical and oral antibiotics; painful oral ulcers disseminated throughout the mucosa; genital painful ulcers with scar formation and vaginal discharge, and foreign body sensation; conjunctival hyperemia without purulent discharge and foreign body sensation. In addition, fever, weight loss, arthralgias, knee arthritus and malaise were present.

Results: This patient had received various treatment schemes, with no resolution. She was referred to pediatric rheumatology, requesting complementary studies WBC: 7,800/mm3, Hemoglobin: 12,4g/dL, hematocrit 37%, Platelets: 315,000/mm3 CRP Negative, urine test: not pathologic, glu-

Conclusions: BD is very prevalent in Mediterranean countries and the incidence is rare in the West. Therefore, we present a female patient with BD.

ACUTE LUPUS PNEUMONITIS AND DIFFUSE ALVEOLAR HEMORRHAGE AS A PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOUS PRESENTATION. A CASE REPORT

Diego Ruben Salinas Encinas1. 1Caja Nacional De Salud, Santa Cruz De La Sierra, Bolivia.

Objectives: To describe serious respiratory symptoms as a clinical initial feature of pediatric systemic lupus and to address itsproper diagnosis and treatment. Systemic lupus erythematosus (SLE) is an autoimmune disorder characterized by the production of autoantibodies.

Methods: 9-year-old female. Refers a 7-day history of fever, cough, pleurisy, dyspnea with hypoxia, and sometimes hemoptysis, acute diffuse lung infiltration on chest radiograph, abrupt drop of hemoglobin level. The mortality rate of DAH in SLE patients is about 23 to 90%. How- ever, these complications are seldom diagnosed early, because of their abrupt onset and rapid progression.

Results: Twenty patients were included, mostly female (n=15; 75%); mean age: 16 years (SD: 2.9). The main reason for referral was RP (n=11; 55%). Digital
ulcers, heliotrope, and Gottron signs were observed in two (10%) of the in-cluded patients, respectively. Antinuclear antibodies were positive in 11 of 17 children (64.7%). In NVC, six patients (30%) had a normal pattern (NP), nine (45%) non-specific autoantibody pattern (NSAP), three (15%) systemic sclerosis pattern (SSP), and two (10%) exhibited a systemic sclerosis-like pattern (SSLP).

Most of the RP patients (n=8; 72.8%) had a pattern different than normal, predominantly NSA (n=4; 36.4%). Regarding capillary density (CD), pa-tients with SSP had a lower CD (median: 5) than patients with SSLP (me-dian: 7.5) and with NSA (median: 9). Megacapillaries were observed in three patients: two with SSP and one with NSA. Microhemorrhages were identified in nine patients: predominantly NSA (n=6; 67.7%). Avascular zones were present only in SSP.

Conclusions: The main reason for ordering NVC in pediatric patients was RP, finding a high frequency of capillaroscopic abnormalities. The SS capillaroscopic pattern changes described in adults also were observed in children. The usefulness of NVC in the approach to RP in the pediatric popu-lation is highlighted.

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POLYARTERITIS NODOSA: A COHORT OF COLOMBIAN PEDIATRIC PATIENTS
Ana María Medina1,2, Mateo Calle1, Juan Peinado1,3, Monica Velasquez Mendez1, Carlos Horacio Molina Vahos1,2, Adrina Luz Vargas García1,3, Luis Jhonnah Hernández Zapata1,2,3, Daniel Jaramillo-Arroyave4,5,6,7, and Ruth Enzo Gamica1,2,3
1Grupo de Estudio de Vasculitis Sistémicas, Universidad de Antioquia, Medellin, Colombia, 2Departamento de pediatría, Facultad de Medicina, Universidad de Antioquia, Medellin, Colombia, 3Departamento de Medicina Interna, Universidad de Antioquia, Medellin, Colombia, 4Hospital Universitario San Vicente Fundación, Medellin, Colombia, 5Sección de Reumatología, Facultad de Medicina, Universidad de Antioquia, Medellin, Colombia, 6IPS Universitaria, Servicios de Salud Universidad de Antioquia, Medellin, Colombia, 7Grupo de Epidemiología y Bioestadística del CES, Medellin, Colombia, 8Hospital Pablo Tobón Uribe, Medellin, Colombia.

Objectives: Polyarteritis nodosa (PAN) is a necrotizing vasculitis characterized by inflammatory changes predominantly in medium sized arteries. PAN is a rare vasculitis in childhood and it is considered a difficult-to-diagnose disease, being these patients often diagnosed late. Objective: To characterize pediatric patients who were diagnosed with PAN in two centers from Medellin, Colombia.

Methods: A descriptive study was conducted using medical records data. Patients under 18 years of age, diagnosed with PAN according to the attending pediatric rheumatologist and confirmed by histologic or angiographic findings compatible with disease, between January 2009 and December 2018 at two re-fERENCE centers from Medellin-Colombia were included. Data from medical records regarding demographic, clinical, laboratory features, treatment and total follow-up period were registered. Data were expressed in median and ranges and mean and standard deviation (SD) according to their distribution.

Results: Nine patients were included. The median age at diagnosis was nine years (range 3-15). Girls were 55.6%. The median follow-up period was 12 months (range 1-105). Cutaneus PAN (cPAN) was diagnosed in 66.4%, and Systemic PAN (sPAN) in 33.6%. All patients (100%) presented fever. Weight loss and fatigue were present in 77.8% and 55.6% respectively. Nodules were observed in 100% and mucosal ulcerations in 44.4%. Lingual and digital necrosis were present in two patients respectively. Calf pains, arthritis and ar-thralgia were present in 66.7%, each one. Abdominal pain was present in 33.3%. Meningoencephalitis as a neurologic manifestation of sPAN was present in one patient. Peripheric nervous system involvement was present in two pa-tients. Erythrosedimentation rate (ESR) and C reactive protein (CRP) were high in all: median ESR was 80 mm/h (range 50-110) and CRP: 20.6mg/dl (range 3,6-45,3). All patients required treatment with glucocorticoids, none of them died during follow up.

Conclusions: In this childhood series, cPAN was more frequent than sPAN, as has already been reported in the literature. All patient presented fever and cuta-neous manifestations and although nodules were the most frequent manifesta-tion, mucosal ulcerations could be observed. All patients required treatment with glucocorticoids.

In pediatric patients with fever, high acute phase reactants (ESR and CRP) and cutaneous findings, PAN should be considered in the differential diagnosis. Mucosal ulcerations although not very frequently reported in childhood PAN patients, also could be present. Despite cPAN has been considered a benign dis-ease, these patients may be severely ill requiring glucocorticoid treatment.

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SEVERE POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES), AN INFREQUENT MANIFESTATION IN A PEDIATRIC PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) AND RENAL INVOLVEMENT
Mariela Viviana Villagómez Estrada1,2, Diego Noboa Torres1,2, Líder Escudero Abad1, Silvia Montaguno Tipantasig2, Edison Narváz Guerrero2, Michelle Fusseau Herrera1, and Lizeth Lozano Herrera1.
1Hospital de las Fuerzas Armadas HE-1, Quito, Ecuador, 2Universidad Central del Ecuador, Quito, Ecuador, 3Universidade de la República, Montevideo, Uruguay, 4Hospital Pediatrício Baca Ortiz, Quito, Ecuador.

Objectives: To describe an unusual presentation of posterior reversible enceph-alopathy syndrome (PRES) in a pediatric patient with systemic lupus erythe-matosus (SLE) and renal involvement, at the hospital de especialidades de las fuerzas armadas N°1 (Armed Forces Hospital) in the city of Quito, Ecuador.

Methods: Fourteen-year-old female patient from Quito-Los Rios, in August 2017 diagnosed with systemic lupus erythematosus, with renal in-volvement (nephrotic syndrome), receiving prednisone 30 mg, hydroxy-chloroquine 200 mg, atovastatin 10 mg, enalapril 5 mg, furosemide 40 mg. Three months after the diagnosis she was admitted to the emergency department for 5 episodes of generalized tonic-clonic seizures of approxi-mately 2 minutes in duration, receiving phenytoin initially and then at main-tenance doses, controlling the seizures. On physical examination, hypertension 143/112 mmhg, mild palpebral oedema, disorriented in time, space and person, Glasgow 12/15 (e4 m5 v3), isocoric hyporeactive pupils, total blindness and de-creased ostoentendular reflexes.

Results: Laboratory results: Moderate anemia: hemoglobin 9.9 g/dl, thrombo-cytopenia: 122,000/mm³, acute renal failure: urea 52 mg/dl, creatinin 1.63 mg/dl, hematuria, proteinuria, anti-nuclear and anti-dsDNA antibodies: positive, hypocomplementemia: C3:26 mg/dl, C4:3 mg/dl, negative antiphospholipid an-tibodies, ANCA C and P: negative, SLEDAI: 28 (severe activity).

Brain magnetic resonance imaging (MRI) showed bilateral cortical-subcortical hypertensive lesions on occipito-parietal lobes, right parietal and left frontal convexity; another lesion is observed in the left thalamus which suggests sub-acute-acute ischemia, characteristic of severe PRES (lesions located in pos-terior and anterior territory, also ischemic lesion in thalamus).

Symptomatic treatment was immediately established with anticonvulsants and anti cerebral oedema drugs, as well as control of causal factors; high blood pressure (enalapril, amlodipine, atenolol, minoxidil), severe lupus flare and lupus nephritis (immunomodulatory drugs, immunosuppressive).

After 15 days, follow-up brain MRI showed a nearly complete resolution of the above lesions. The diagnosis was confirmed with the resolution of the clinical and imaging abnormalities.

Conclusions: 1.-PRES is related to different pathologies and drugs. In our pa-tient, the predisposing factors were severe lupus activity, uncontrolled hyperten-sion, lupus nephritis and immunosuppressive agents use.

2.-The pediatric SLE-PRES association is infrequent, is based on clinical-radiological diagnosis, and must be considered in the differential diagnosis in patients who develop neurological symptoms: seizures, visual abnormalities and alterations of the mental state.

3.-Early diagnosis and treatment of triggering factors are key to avoid lethal complications, usually reversing the clinical and imaging abnormalities.

4.- Therapeutic approach is multidisciplinary: pediatrics, rheumatology, neurology, and nephrology.

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JOINT INVOlVEMENT IN JUVENILE DERMATOMYOSITIS (JDM)
Claudia Malagón1, and Karen Jiménez1. 1Universidad El Bosque, Bogotá, Colombia.

Objectives: To determine the types of joint involvement at onset and during the course of the disease.

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