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BEHÇET'S DISEASE IN CHILDHOOD. A CASE REPORT

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Objectives: To describe a case of recurrent aphthous stomatitis, genital ulceration, and uveitis, compatible with Behçet's disease (BD) and be aware of it and 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 arthritis and malaise were present.

Results: This patient had received various treatment schemes, with no resolution of her symp. Referred to pediatric rheumatology, requesting complementary studies WBC: 7,800/mm3, Hemoglobin: 12,6g/dL, hematocrit: 37%, Platelets: 315,000/mm^{3}CRP Negative, urine test: not pathologic, glucose: 104 mg / dl Urea: 17,3 mg / dl, Creatinine: 1,04mg/dl, SGPT: 9 U/L SGOT: 17.5 U/L, Amylase: 64.6U/L; ANCA, APS antibodies: negative; Tuberculosis serology negative; C3 169 mg/dL, C4 28 mg/dL (normal value); ANA: 1.93 positive, anti DNA negative. Oral wound secretion culture: Streptococcus sp, HIV IgG and IgM: not reactive, vaginal secretion culture: Candida albicans. HLA B5-B51 requested but could not be carried out due to lack of resources. Pathergy test: positive. Gathering data, recurrent oral and genital ulcers, ocular involvement, a probable diagnosis of systemic vasculitis: BD was made. Started on oral corticoids, colchicine and calcium.

Pediatric neurology ruled out neurological compromise by brain nuclear magnetic resonance, ophthalmology assessment indicates anterior uveitis without retinal involvement.

Conclusions: BD has an intermittent protean clinical picture, and the diagnosis could be delayed, due to the rarity of the condition and its non-specific presentation, specially in pediatric populations. The diagnosis of BD is challenging for pediatricians and is important to keep in mind that besides mucocutaneous symptoms patient can have both ocular, neurologic, gastroenterologic or vascular involvement with devasting complications, and even death.

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

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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.

Acute lupus pneumonitis and diffuse alveolar hemorrhage (DAH) are one of the most dreaded complications. Patients usually present with 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%. However, these complications are seldom diagnosed early, because of their abrupt onset and rapid progression.

Methods: 9-year-old female. Refers a 7-day history of fever, cough, malaise, weight loss, hyporexia and maculo-erythematous lesions on the face, scalp and abdomen. Remarkable examination, erythematous and crusted non-bleeding forehead and scalp lesions, rales and bilateral basal crepitus, SpO2 97%. No adenopathy, no hepatomegaly or splenomegaly. Admitted as pneumonia, with initial complementary studies WBC 7000/mm^3, neutrophils 73%, lymph 24%, platelets 105,000/mm^3, Hemoglobin 8.2g/dL, hematocrit 26%, Erythrocyte sedimentation rate (ESR) 59mm, CRP positive. Chest radiograph showed isolated bilateral alveolo-interstitial infiltrates. Despite antibiotic regimen, fever persisted, becoming more frequent. Antibiotic scheme was changed every 72 hours, not ameliorating clinical symptoms, but keeping good appearance. Meanwhile negative viral serologys were obtained. Chest CT scan shows a 2.59x2.18cm apical segment right lung field consolidation, air bronchogram and central bronchiectasis a 4.8x3.5cm apical-posterior segment left lung field consolidation. Consolidation image in posterior basal segment. Pleural effusion with 1.07cm of detachment at the posterior level.

Results: After 2 weeks, hematology ruled out infiltrative process. Painless oral ulceration and facial erythema were detected, pulmonary symptoms worsen suddenly with desaturation, respiratory distress, haemoptysis, and ill appearance. WBC 1500/mm^3, platelets 90,000mm^3; Hemoglobin 7.3. Patient was admitted to the Pediatric intensive care unit; bleeding was evident through the orotracheal cannula. Chest radiograph showed diffuse bilateral alveolar infiltrates, compatible with alveolar hemorrhage, Hemoglobin 5.1g/dL and hematocrit 18.1%. Negative cultures. ANA positive 101 (<50), anti DNA positive by Crithidia luciliae, hypocomplementemia C3 0.37 (0.8-1.57), C4 0.086 (0.12-0.39), Anti Sm 23.5 mg/dL (<8), IgG 32.9 (6-14.5), negative VDRL, Coombs, ANCA and antiphospholipid antibodies profile. Therefore, pediatric SLE and pulmonary involvement diagnosis was made. No renal o cardiac involvement was detected. Methylprednisolone boluses plus cyclophosphamide were started. The response was partial, but relapsing bleeding make it difficult to wean the patient. Human immunoglobulin, biweekly cyclophosphamide and mofetil mycophenolate were initiated, with good response. Hemoglobin 11.4, hematocrit 34.8, and platelets 358.000/mm^3.

Conclusions: The respiratory symptoms in our patient mimicked an infectious disease, but ancillary studies were not consistent, and improvement was not seen with antibiotic treatment. Abrupt respiratory deterioration, bleeding and desaturation broaden diagnostic possibilities, and positive antibodies allowed SLE to be diagnosed. We cannot forget that DAH and acute pneumonitis are characterized by nonspecific symptoms such as hemoptysis, dyspnea, cough, sputum, hypoxia, and acute diffuse lung infiltration on chest radiographic images, carrying potentially serious prognosis and even death if they are not appropriately detected.

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CHARACTERIZATION OF THE PEDIATRIC POPULATION REFERRED TO THE VIDEOCAPILLAROSCOPY SERVICE AT THE CLINICA UNIVERSITARIA BOLIVARIANA MEDELLIN- COLOMBIA. 2015-2018

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Objectives: Nailfold videocapillaroscopy (NVC) is a technique that allows evaluating the microcirculation. In pediatrics, it is useful in the approach to Raynaud´s phenomenon (RP) and diseases of the spectrum of systemic sclerosis (SS). While in initial studies in children, it was spoken as a "sclerodermiform" capillaroscopic pattern, the classification of adults in early, active, and late patterns has been used, with favorable results. To date, the use of NVC in Colombian children has not been reported. The objective was to describe the features of the patients referred to an NVC service in Medellin, Colombia.

Methods: Descriptive study. Patients younger than 19 years old referred to the NVC service at the Clinica Universitaria Bolivariana were included (2015-2018). NVC reports and referral notes were analyzed. Variables included: sociodemographic, clinical, laboratory, and capillaroscopic findings. Qualitative variables were expressed as absolute and relative frequencies and quantitative ones with mean and standard deviation (SD) or median and interquartile range (IQR) according to their distribution.

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 included 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%) nonspecific alterations (NSA), 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), patients with SSP had a lower CD (median: 5) than patients with SSLP (median: 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 population is highlighted.

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POLYARTERITIS NODOSA: A COHORT OF COLOMBIAN PEDIATRIC PATIENTS

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Objectives: Polyarteritis nodosa (PAN) is a necrotizing vasculitis caracterized 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. Patientes 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 reference centers from Medellín-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 66,7% and Systemic PAN (sPAN) in 33,3%. 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 arthralgia 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 patients. Erythrosedimentation rate (ESR) and C reactive protein (CPR) 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 frecuent than sPAN, as has already been reported in the literature. All patient presented fever and cutaneous manifestations and although nodules were the most frequent manifestation, 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 frecuently reported in childhood PAN patients, also could be present. Despite cPAN has been considered a benign disease, 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

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Objectives: To describe an unusual presentation of posterior reversible encephalopathy syndrome (PRES) in a pediatric patient with systemic lupus erythematosus (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 Quevedo-Los Rios, in August 2017 diagnosed with systemic lupus erythematosus, with renal involvement (nephrotic syndrome), receiving prednisone 30 mg, hydroxychloroquine 200 mg, atorvastatin 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 approximately 2 minutes in duration, receiving phenytoin initially and then at maintenance doses, controlling the seizures. On physical examination, hypertense 143/112 mmhg, mild palpebral oedema, disoriented in time, space and person, Glasgow 12/15 (e4 m5 v3), isocoric hyporeactive pupils, total blindnes and decreased osteotendinous reflexes.

Results: Laboratory results: Moderate anemia: hemoglobin 9.9 g/dl, thrombocytopenia: 122,000/mm^3, 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 antibodies, ANCA C and P: negative, SLEDAI: 28 (severe activity).

Brain magnetic resonance imaging (MRI) showed bilateral cortical-subcortical hyperintense lesions on occipito-parietal lobes, right parietal and left frontal convexity; another lesion is observed in the left thalamus which suggests subacute-acute ischemia, characteristic of severe PRES (lesions located in posterior 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 presure (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 previous lesions. The diagnosis was confirmed with the resolution of the clinical and imaging abnomalities.

Conclusions: 1.-PRES is related to different pathologies and drugs. In our patient, the predisposing factors were severe lupus activity, uncontrolled hypertension, 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)

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