of her right elbow. The patient denied any trauma or associated symptoms such as muscle weakness or joint stiffness. On physical examination, a large hard mass was present over her left trochanteric region, with hyperemic skin, and a similar one was palpable over her right elbow.

Results: Radiographs of the two sites showed well-defined calcified masses, one (9x5 cm) over the trochanteric region, and the other (3x2) cm over the posterior aspect of the distal end of the humerus extending up to the elbow. MRI confirmed the presence of calcified intramuscular lesions (Figure 1). Serologic evaluation showed increased levels of CRP and ESR, while autoimmune screening was negative. Calcium/phosphorus metabolism, creatinine kinase, aldolase and ferritin levels were within the normal limits. Therefore, a clinical diagnosis of idiopathic calcinosis was made. After 3 months, the lesion on the elbow had spontaneously reabsorbed, while the other one kept on growing, with 3-4 episodes per month of febrile painful swelling. A CT-guided biopsy was then performed, confirming the diagnosis of calcinosis. No infectious causes were detected. One year later, since there was no local amelioration, the lesion was surgically removed with apparent complete local remission. No other new lesions were detected during the 3 months follow-up period.

Conclusion: Idiopathic calcinosis is very rare and no specific medical therapies are available. In our patient no causative factors were identified and thus the diagnosis of idiopathic calcinosis was made. Prognosis seems to be variable, since one lesion resolved spontaneously while the other needed surgical excision.

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Abstract AB0971 Figure 1. MRI of the trochanteric region showing calcified intramuscular lesion.

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AB0972 ARTEFACTUAL SKIN LESIONS: A TRAP FOR RHEUMATOLOGISTS

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Background: Dermatitidis artefacta (DA) refers to deliberate and conscious self-inflicted skin lesions to satisfy an unconscious psychological or

emotional need. It is an unusual condition often described in women, and rarely reported in children. The disorder is often misdiagnosed and needs to be distinguished from organic illness, Munchausen's by proxy syndrome, and from illicit, nontherapeutic drugs or overdose of medications.

Objectives: We report three paediatric cases with a long medical history characterized by many laboratory and radiological exams, and mistaken etiologies.

Methods: *Case 1.* An 11-year-old female was referred to our Rheumatology Department for a second opinion about a suspicion of Behcet's disease based on her cutaneous manifestations. Clinical examination was unremarkable except for bluish ecchymoses, and linear cutaneous abrasions and scars located in accessible areas of her skin (Figure 1, Figure 2 and Figure 3). Extensive laboratory tests and radiological exams were normal.

Case 2. A 14-year-old female was admitted to our pediatric hospital for a 1 month history of fever, malaise and multiple purpuric lesions. The patient referred a 3 year history of similar recurrent episodes characterized by abrupt onset of fever, arthralgia and purpuric lesions for which she already received the diagnosis of urticarial vasculitis. The girl was in good general conditions, without fever. Violaceous ecchymotic lesions were evident on her left forearm. Exhaustive laboratory tests, chest X-ray, and abdomen ultrasound were within normal limits.

Case 3. A 14-year-old female was referred to our Pediatric Rheumatology department for a 6 month history of musculoskeletal pain, recurrent shoulder and hip dislocations, and spontaneous ecchymosis on her neck and face. Except for joint hypermobility (Beighton score 7/9), physical examination was normal. At a careful orthopedic evaluation and after shoulder and hip imaging the history of dislocations appeared to be inconsistent. Blood and radiologic tests, cardiologic and genetic evaluations for Elher-Danlos syndrome were negative.

Results: In *Case 1*, dermatitis artefacta was suspected and the patient was referred to our mental health colleagues that confirmed a severe psychological stress in a basal condition of psychiatric disorder. In *Case 2*, an in-depth psychological evaluation highlighted a depressive status for personal and familiar suffering. The patient was therefore referred to our mental health department for assessment and management. In *Case 3*, eventually a psychiatric interview oriented the diagnosis to a major depressive disorder.

Conclusion: DA is a rare condition that should be evaluated in the differential diagnosis, in the presence of unexplained cutaneous lesions without an apparent identifiable cause. Association with various psychological disturbances is the mainstay for this diagnosis, and a careful psychiatric history represents the essential diagnostic key to identify this disorder.

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Abstract AB0972 Figure 1

Abstract AB0972 Figure 2

Abstract AB0972 Figure 3

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AB0973 ATYPICAL KAWASAKI DISEASE SHOCK SYNDROME CASE REPORT: NOT ALL SHOCKS ARE SEPTIC OR TOXIC

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Background: Kawasaki disease (KD) is an acute, self-limited vasculitis of unknown etiology with peak incidence at 9-12 months of age. The term Kawasaki Disease Shock Syndrome (KDSS) was introduced in 2009 after reports of hemodynamic instability during the acute phase of the illness.

Objectives: Present and discuss a rare case of KDSS in an adolescent patient.

Methods: Review of patient's clinical records and scientific literature.

Results: A 12 year-old boy was admitted in the pediatric intensive care unit (ICU) with a 5-day history of high persistent fever, abdominal pain, vomiting and headaches. On examination, he looked ill, was tachycardic and hypotensive with delayed capillary refill, had non-exsudative oropharyngeal erythema, cracked lips, terminal nuchal rigidity, jaundice and small palpable supraclavicular lymph nodes. On day 3 a generalized morbilliform rash and diffuse swelling of the hands became apparent. Blood gas showed metabolic acidosis. He had decreased platelet count of 72,000/mm³, AST 122 U/L, ALT 192 U/L, LDH 366 U/L, triglycerides 207 mg/dl, total bilirubin 85,5 µmol/l (direct 68,4). Renal function, urinalysis and fibrinogen were normal. ESR was 42 mm/h, CRP was markedly high (268 mg/l) and ferritin 544 ng/ml. Lumbar puncture revealed mild sterile pleocytosis. Due to progressive hemodynamic instability he required fluid resuscitation, inotropic drugs and mechanical ventilation. Broad-spectrum antibiotherapy was started due to suspected septic shock/streptococcal toxic shock syndrome. IVIG was empirically started 24h later due to non-

response. Viral screening, all blood and urine cultures and ASO titers were negative. Myelogram revealed no signs of hemophagocytosis, neoplastic changes or growth of microorganisms in culture. Fine needle biopsy of a supraclavicular lymph node showed nonspecific inflammatory changes. Thoracoabdominal scan revealed moderate bilateral pleural effusion, mild hepatosplenomegaly, retroperitoneal/mesenteric lymphadenopathy. Brain MRI and echocardiogram were normal as well as ophthalmologic evaluation. In the presence of prolonged fevers, mucosal changes, extremity edema and non-specific rash in a severely-ill adolescent with no response to broad spectrum antibiotics and negative cultures, the possibility of KD was raised.

Due to ongoing fevers and inflammation after 36h of first IVIG, he received a second IVIG dose 2g/Kg, methylprednisolone IV pulses for 3 days and aspirin, with rapid clinical and laboratorial improvement. Within 24h the patient was off inotropic support, 48h off mechanical ventilation and 3 days later he was discharged from the ICU. His general condition continued to improve gradually, with increased platelet counts, normalization of liver function and CRP. In this phase, skin desquamation (buttocks/perineal region) was noted. At follow-up, thoracoabdominal angioMRI and serial ecocardiograms were normal. He was weaned off corticosteroids and aspirin without recurrence of symptoms.

Conclusion: In this case, despite the atypical age and lack of some classical signs/symptoms, broad-spectrum antibiotic refractoriness and the described clinical presentation raised the hypothesis of KDSS. Diagnosis can be difficult, especially if shock occurs in incomplete forms of KD, but must be suspected early and treatment promptly started in order to ensure a good prognosis. Clinicians should be aware that thrombocytopenia and hepatitis are risk factors for refractory severe KD. MAS must always be excluded in cases of hemodynamic instability.

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AB0974 A CASE OF ADENOSINE DEAMINASE 2 DEFICIENCY (DADA2) WITH AN UNCOMMON CLINICAL PRESENTATION AND RESPONSE TO IV IG

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Background: DADA2 is an autoinflammatory disease with autosomal recessive inheritance characterized by a heterogeneous clinical phenotype ranging from multisystemic inflammation (fever, polyarteritis nodosa, cerebral stroke, livedo reticularis etc.) to immune-dysregulation and immunodeficiency.

Objectives: To extend the clinical spectrum of DADA2 reporting a case of isolated nonspecific systemic inflammatory syndrome associated with signs of immune-dysregulation in a patient with a novel ADA2 mutation.

Methods: In a patient with nonspecific inflammatory phenotype associated to susceptibility to viral infections, Next Generation Sequencing (NGS) panel was performed; mutations detected were confirmed by Sanger analysis. ADA2 enzymatic activity was analyzed in monocyte isolated from the patient and incubated with adenosine and an ADA1 inhibitor.

Results: The girl, adopted and of Asian origin, began to suffer from non-specific systemic inflammatory symptoms (persistent fever and arthralgias) at the age of 6. In past history recurrent respiratory infections and impaired immunological response to viruses (CMV related hepatitis, measles after vaccination) were reported. After few months the patient developed clinical and laboratory findings of HLH (Hemophagocytic Lympho-Histocytosis), confirmed on bone marrow samples; treatment with intravenous (IV) high dose (HD) steroids was started, with prompt response. During steroids tapering fever and systemic inflammation reappeared; anti-IL1 treatment (anakinra) was not effective. Immunologic assessment demonstrated mild hypogammaglobulinemia and moderate NK deficiency on lymphocyte subsets. HD IV Immunoglobulins (IG) (2 g/kg every month) allowed to achieve a complete control of the clinical picture; the frequency of administration was progressively reduced to every 4 months due to persistent wellbeing. At the age of 9, after switching IG to the