low levels of T+ CD4 cells and recurrent infections. Intestinal histoplasmosis may have contributed to the development of malakoplakia trough disturbances of the phagocytic abilities of the machrophage. The immunological disorder investigation for this patient continues in progress.

A055
Systemic granulomatosis disease in common variable immunodeficiency – case report

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Background: At least 5% of common variable immunodeficiency (CVID) patients develop non-caseating granulomas on the skin, solid organs, or lymphoid tissues. These cases have an unpredictable outcome and the prognosis is generally worse.

Case Report: The authors present the case of 34-years-old male patient, with a diagnosis of CVID since 2003. One year after the diagnosis leucopenia (2600/uL) and thrombocytopenia (110000/uL) were detected. The abdominal CT scan showed a homogenous splenomegaly. The high resolution thoracic CT scan exposed a nodular lesion with a spiculated edge, as well as several mediastinal adenopathies. The bronchoalveolar lavage revealed a high number of total cells (50/uL), with 50% lymphocytes and a normal CD4+/CD8+ ratio (1 : 13). The lung nodule biopsy showed non-caseating granulomas, indistinguishable from those in sarcoidosis. The mantoux test, as well as the search for mycobacteria and fungi were negative. Functional respiratory tests were normal. Anti-nuclear antibodies, platelet and erythrocyte autoantibodies were negative. Infections by HIV 1 and 2, HCV and HBV were excluded.

Results: The patient initiated treatment with 40 mg prednisolone/day (0.8 mg/Kg/day), associated with intravenous immunoglobulin (IVIG) at 3 week interval at the appropriate dose to achieve pre-treatment levels of IgG higher than 8 g/L. After 3 months, an improvement in splenomegaly, as well as a reduction in the pulmonary lesion was observed, with posterior normalization of the haematological values, with 8500 leucocytes/uL and 165.000 platelets/uL. The systemic corticosteroids were progressively tapered and were suspended in June/2006. Since then, the leucocytes values have been between 3.200 and 5400/uL, platetets values between 111.000 and 157000/uL and pre-infusion IgG between 6,59 and 12 g/L. The high resolution CT scan (2007) revealed residual fibrotic lesions. The abdominal ultra-sonography was normal. The patient maintains episodes of cough and purulent sputum, with need of antibiotherapy 2–3 times/year.

Conclusion: The optimum treatment for systemic granulomatous disease in CVID patients isńt well defined. In this case, the association of corticotherapy with higher doses of IVIG led to a reduction of the nodular lung lesion, splenomegaly and to an improvement in haematological values. Patients with CVID and systemic granulomatosis disease appear to have a worse prognosis, essentially due to the increased risk of infection, autoimmune disease and recurrence of granulomatous lesions.

A056
Wiskott–aldrich syndrome; clinical outcome and immunological features of 13 patients

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Wiskott–Aldrich syndrome (WAS) is a rare X-linked immunodeficiency, characterized by microthrombocytopenia, recurrent infections and eczema. Patients are at high risk for the development of malignancy and autoimmune diseases. Here, we present clinical manifestations, laboratory findings, immunological features and the events that occurred in 13 WAS patients from seven different families followed between 1982 and 2008. In five patients, the diagnostic evaluation was performed because of a family member known to be affected. All patients admitted to the hospital for bleeding and petechial rash. Eczematous lesions and ecimotic areas were detected in all. Ten patients had history of frequent mild-severe infections. Summary of laboratory evaluation and clinical outcome of the patients are seen in the table.

Table 1. Laboratory evaluation and clinical outcome

Age at diagnosis (Median/Range)	4 years/(6 months-6years)
Platelet Count (Mean/Range)	24000 (11000-53000 mn/3)
Platelet Size (Mean/Range)	5.2 fl (4.2-7 fl)
Immunological Studies Decreased IgG Elevated IgA Decreased IgM Decreased CD4+ cell level Diminished lymphocyte response to PHA	2/13 patients 9/13 patients 8/13 patients 6/13 patients 7/13 patients
Autoimmunity	2/13 patients (Leukocytoklastic vasculitis)
Lymphoproliferative disorder	2/13 patients (Non-Hodgkin's lymphoma and EBV-related Lymphoproliferative disorder)
Chronic colitis	1/13 patients
Bronchiectasis	2/13 patients
Treatment	HSCT (Matched related donor) 3/13 patients IVIG and TMP-SMX (Prophylactic) 12/13 patients
Outcome	Alive 11/13 Dead 2/13 (Sepsis-Bleeding)
Follow up time	2.5-23 years

