those who responded partially evolved to liver cirrhosis, with a similar follow-up time in both groups (105 vs 113 months). 5 of the 6 patients who did not have a biochemical response developed liver cirrhosis. 7 of the 13 patients with cirrhosis (54%) already presented clinical or histological data of cirrhosis in the initial evaluation

Conclusions: PBC patients have frequently other autoimmune diseases such as Autoimmune Hepatitis, Sjögren syndrome or Scleroderma so we must actively seek the presence of these pathologies. The treatment with ursodeoxycholic seems to be useful in all patients but it is important to make an early diagnosis

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AB1144

HISTOLOGY OF ROSAI-DORFMAN DISEASE IN A SUBSET OF PATIENTS WITH ERDHEIM-CHESTER DISEASE: A DISTINCT ENTITY MAINLY DRIVEN BY MAP2K1

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Background: Diagnosis of Erdheim-Chester disease (ECD) is based on characteristic imaging of bone, retroperitoneal and/or cardiovascular involvement.¹ Biopsy is mandatory to exclude other diagnoses and confirm infiltration of histiocytes, but histology is not specific.² By contrast diagnosis of Rosai-Dorfman disease (RDD), a rare histiocytosis, is based on histology, which is characterised by infiltration by CD68 +CD1 a- S100+ histiocytes with large nuclei and abundant lesions of emperipolesis.² Up to 70% of ECD have *BRAF* or *MAP2K1* mutations,³ which are rare in RDD.

Objectives: We investigated patients harbouring an ECD phenotype but RDD histology.

Methods: We reviewed records of ECD patients followed in Pitié-Salpêtrière hospital (Paris, France) and Memorial Sloan Kettering Cancer Centre (New-York, NY, USA) between 2007 and 2018. Biopsy samples of the patients were systematically investigated for mutations of genes of MAP kinase pathway.

Results: Among 209 patients with ECD, we found 10 (4.7%) patients who had RDD histology. These 10 patients had typical ECD clinical and radiological presentation, in particular bones (n=7), vascular (n=5) and peritoneal (n=6) involvements. Patients also had typical neurological involvement of ECD (n=6). All patients except one had at least one biopsy with a compatible histology of ECD at diagnosis. ECD biopsies showed non-specific fibrosis (n=5), foamy CD 68 +CD1 a- histiocytes (n=3) and/or Touton cells (n=1). Biopsies disclosing RDD histology were performed during the course of the disease involving testes (n=5), stomach (n=1), tibia (n=2), cheek (n=1) and omentum (n=1). All tissues showed lympho-plasmocytic infiltrate with large histiocytes infiltration. Histiocytes were CD68 +CD1 a- S100 +with large nuclei and abundant lesions of emperipolesis. Five patient harboured *MAP2K1* mutation and one patient had *PIK3CA* mutation. None of the patients had *BRAF* mutation.

Conclusions: Some patients with ECD may also present the iconic histological lesions described by Rosai and Dorfman. Overlap forms of such distinct histiocytoses between ECD and RDD is mainly driven by *MAP2K1* but not by *BRAF*.

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AB1145

ACUTE POSTERIOR MULTIFOCAL PLACOID PIGMENT EPITHELIOPATHY: CLINICAL PATTERN AND MANAGEMENT OF 79 PATIENTS

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Background: Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE) is an uncommon inflammatory disease causing acute-onset chorioretinal bilateral disease. It typically affects the posterior pole of both eyes leading to visual blurring or scotomas. Although it is thought to be benign, APMPPE has been associated with central nervous system (CNS) manifestations: cerebral vasculitis, meningoencephalitis and cerebral vascular disease.

Objectives: The aim of this study was to define clinical features, systemic manifestations, treatment and outcomes of a review of 79 patients with APMPPE.

Methods: We retrospectively analysed the epidemiology, potential triggers, prodromes, clinical data, ophthalmological study, extraocular manifestations, treatment and outcomes of 79 patients collected through an extensive review of the literature from the first description of Gass JD up to the present time.

Results: A total of 79 patients were reviewed (47 male). Mean age at diagnosis was 30 years (with a range of 8 to 58 years old). 27 patients (34.2%) presented with a previous triggering being flu-like illness the most frequent.¹⁵ However, the complete serological study was only requested in 28 patients (and the immunological study just in 22) Median time from trigger to overt APMPPE was 9 days. Main clinical symptoms were: decreased visual acuity/blurred vision (100%), headache (51.8%) and photophobia (12.2%). Average decreased visual acuity was 13/20. APMPPE is defined by the presence of multiple white-yellowish plaques in funduscopy, and early hypofluorescent areas with late hyperfluorescence in fluorescein angiography. The fundus was pathological and compatible with APMPPE in all cases (100%), as was fluorescein angiography in those that had been performed (59). CNS involvement appeared in up to 50.6% (40 patients). The CNS manifestations were divided into language disorders (11 patients), motor deficit,²⁰ sensory deficit²⁰ and other CNS manifestations.²³ The mean time from visual deficit to neurological manifestations was approximately 2 weeks. Cerebrospinal fluid was studied in 37 patients, with a predominance of lymphocytic pleocytosis (mean of 46 cells/mm3) and elevated proteins (mean of 111 mg/dl). Within the neuroimaging studies carried out (58) up to 69.7% were pathological. 67 patients (84.8%) received treatment with corticosteroids. 14 patients (17.7%) also received other immunosuppressants (mainly azathioprine and cyclophosphamide), especially if CNS involvement. Regarding the evolution, 55 patients (69.6%) presented improvement, 12 (15.2%) relapsed and 6 (7.5%) died due to APMPPE.

Conclusions: APMPPE is a rare inflammatory disease which primarily affects the retina. However, the CNS involvement could be more frequent than what is classically described. Also, it seems that there might be a trigger effect either inflammatory or infectious. Steroids and immunosuppressants should be considered in patients with CNS involvement from the beginning.

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AB1146

THE ASSOCIATION OF COMMON MEFV GENE MUTATIONS WITH AXIAL SPONDYLARTHROSIS IN FMF PATIENTS: A RETROSPECTIVE STUDY

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Background: Familial Mediterranean fever (FMF) is an autoinflammatory disease seen with autosomal recessive inheritance and is characterised by recurrent and self-limiting attacks with peritonitis, pleuritis, arthritis or fever alone. The association of spondylarthrosis and FMF is reported in some studies. There are few studies evaluating the association of MEFV gene mutations with axial spondylarthrosis in FMF patients.

Objectives: The aim of this study is to identify patients with FMF associated spondylarthrosis retrospectively and to compare the frequency of common MEFV gene mutations in FMF patients with and without axial spondylarthrosis.

Methods: We have reviewed 138 charts of FMF patients. The data of 116 patients (70 female, 46 male) who met the diagnosis of FMF with Tel Hashomer classification criteria and have the results of MEFV gene mutation were examined. Patients' age, sex, MEFV gene mutations were recorded. The presence of