a normal laboratory testing, except for Mycoplasma serology that increased up to 1:5120 two months after the onset of symptoms. Only five other case reports describe a primo-infection with mycoplasma pneumoniae causing hepatitis with(out) cholestasis. We present a case of a recurrent Mycoplasma induced hepatitis and to our knowledge this has never been reported previously. Mycoplasma is a rare cause of cholestasis in children and should be recognized early to enable adequate and early treatment

- Y04 -

INTRAHEPATIC CHOLANGIOCARCINOMA WITH PREDOMINANT DUCTAL PLATE MALFORMATION PATTERN: AN UNUSUAL MIMICKER OF BENIGN LESION. F. Noel (1), O. Detry (2), C. Sampoux (3), N. Blétard (1) / [1] CHU of Liège, Belgium, Anatomie pathologique, [2] CHU of Liège, Belgium, Abdominal Surgery, [3] CHUV Lausanne, Lausanne, Switzerland, Pathology Department.

Case Report: Background Intrahepatic cholangiocarcinoma with predominant ductal plate malformation pattern is a recently discovered rare entity. This lesion has similar histological features seen in a ductal plate malformation, which resembling ductular reaction and would be developped from hepatic progenitor cells. Materials, methods and results A 57-year-old woman presented a liver mass in the right lobe. A diagnosis of biliary adenofibroma was suggested on the first biopsy. In a second stage, a right hepatectomy was performed. On gross examination, we found a lesion 5,5 cm long, whitish, firm, well-demarcated, homogeneous, located within segment 7. On histology, this lesion was unencapsulated and pushing out the rest of the non-tumorous liver parenchyma. The tumour is characterised by a scattered well-differentiated ductular proliferation within a dense fibrous stroma. Glandular structures with irregular dilated lumens are lined by a low-colmunar-to-cuboidal cells with high nucleocytoplasmic ratios and round vesicular nuclei. There was no marked pleomorphism. Usually, no mucin is visualised. Some immunohistochemistry was realised, Arginase 1, Hep-Par1 and S100 were negative. All tubules expressed CK7 and CK19 heterogeneously. EMA demonstrated an apical diffuse marking on tumor cells. EPCAM revealed an extended basolateral positivity and NCAM, a partial basolateral positivity. Immunohistochemistry for P53 was wild-type. The proliferation index assessed by ki67 was low, around 5%. Finally, a diagnosis of intrahepatic cholangiocarcinoma with predominant ductal plate malformation pattern was retained. Conclusion Intrahepatic cholangiocarcinoma with predominant ductal plate malformation pattern is an unusual entity that can mimic a benign lesion.

- Y05 -

AUTOIMMUNE HEPATITIS DEVELOPING AFTER SARS-COV-2 VACCINATION: SERIES OF THREE CASES. G. Rasschaert (1), S. François (1), A. Verbeeck (2), M. Schils (1), M. Aerts (1), P. Lefesvre (3), I. Colle (4), H. Reynaert (1) / [1] Universitair Ziekenhuis Brussel, Brussels, Belgium, Gastroenterology and Hepatology department, [2] Algemeen Stedelijk Ziekenhuis Aalst, Aalst, Belgium, Gastroenterology and Hepatology department, [3] Universitair Ziekenhuis Brussel, Brussels, Belgium, Pathology department, [4] Universitair ziekenhuis Gent, Belgium, Gastroenterology and Hepatology Department.

Case Report: We present a series of three patients with an assumed diagnosis of autoimmune hepatitis (AIH) after SARS-CoV-2 vaccination. A 57-year-old woman was referred for progressive elevation of liver function tests (LFTs) four weeks following the first dose of Moderna mRNA-1273 vaccination. Medical history included Hashimoto's thyroiditis and SARS-CoV-2 infection six months earlier with a mild disease course. She did not take medication or herbal supplements. No substance abuse was recorded. LFTs were normal one month before vaccination. Physical examination was unremarkable. At referral, laboratories were significant for aspartate aminotransferase (AST) (22xULN), alanine aminotransferase (ALT) (38xULN), alkaline phosphatase (AP) (1.3xULN) and gamma-glutamyl transpeptidase (GGT) (3xULN). Hepatitis A, B, C and E virus markers, HIV, Cytomegalovirus, Epstein-Barr, Herpes simplex type 1 and 2 serology were negative. Ceruloplasmin and α1-antitrypsin levels were normal. Anti-mitochondrial, anti-smooth muscle, anti-liver-kidney microsomal antibodies were negative, while antinuclear antibody (ANA) was positive (1:320, speckled pattern). Total IgG was normal. Abdominal ultrasound was unremarkable. Histology showed interface activity including a mixed inflammatory infiltrate with predominant lymphocytes, focal rosetting was observed. According to the International Autoimmune Hepatitis Group (IAIHG) criteria, pre-treatment score was 19, accounting for definite AIH. Immunosuppression was not immediately started due to slow but progressive decrease of LFTs. However, six weeks later budesonide 9mg was initiated for an increase in LFTs (transaminases x10 ULN). Lack of efficacy necessitated a shift towards methylprednisolone 32mg after one month. Azathioprine 50mg was associated two weeks later. Currently she is doing well with marginal AST and ALT elevation (<1.5ULN) and ANA normalisation. In the future, we will continue to treat her according to the standard of care for AIH. A 53-year-old man consulted for silent icterus since four days, seven days after completing the schedule of Pfizer-BioNTech BNT162b2 mRNA vaccination. He denied smoking, alcohol and medication use and had a negative medical history. LFTs were normal six weeks before vaccination. There was no previous SARS-CoV-2 infection. Physical examination showed jaundice. Laboratories were significant for AST (48xULN), ALT (95xULN), AP (2xULN) and GGT (12xULN). Total bilirubin was 3.1g/dL (direct 2.8mg/dL). Viral serology was negative. Ceruloplasmin and α1-antitrypsin levels were normal. Auto-immune panel showed positive ANA (1:80, speckled pattern). Total IgG was normal. Abdominal ultrasound observed steatosis. Histology was similar to the first patient. According to IAIHG criteria, pre-treatment score was 13, accounting for probable AIH. Spontaneous LFT improvement was appreciated two weeks following presentation. A 39-year-old woman was referred for LFT alterations four weeks after her first dose of Pfizer-BioNTech BNT162b2 mRNA vaccination. She had a medical history of Hashimoto's thyroiditis and rheumatoid arthritis. LFTs were normal seven months before vaccination. She reported alcohol (5 units/week) and tobacco consumption (12 cigarettes/day). No over the counter drugs were reported. There was no documented previous SARS-CoV-2 infection. Physical examination was unremarkable. Laboratories were significant for AST (11x ULN), ALT (17x ULN), and GGT (1.2x ULN). Viral serology was negative. Ceruloplasmin and α1-antitrypsin levels were normal. The auto-immune panel was negative. Total IgG was normal. Abdominal ultrasound showed mild steatosis. Histology disclosed interface hepatitis and predominant lymphocyte infiltration without biliary changes. According to IAIHG criteria, pre-treatment score was 16, accounting for definite AIH. Immunosuppressive therapy with methylprednisolone (32mg) and azathioprine (100mg) was initiated after work up, about eight weeks after documentation of LFT alteration. At that time there was a continued increase of LFT alterations with a bilirubinaemia of almost 4.0g/dL. An adequate response was observed. With her treatment of methylprednisolone 8mg and azathioprine 100mg, LFTs almost completely normalised. Above, we described the onset of AIH after vaccination with different mRNA SARS-CoV-2 vaccines in three patients. It is accepted AIH can be triggered by a plethora of viruses and drugs. Additionally, past reports have attributed development of AIH to prior vaccination suggesting a potential role of both virus and vaccine in revealing AIH in predisposed individuals. So far there is no pathophysiological link between SARS-CoV-2 vaccines and AIH. One hypothesis is molecular mimicry. In vitro data demonstrates spike protein S1 antibodies (for which the mRNA codes) have high affinity against transglutaminase 3, transglutaminase 2, anti-extractable nuclear antigen, nuclear antigen and myelin basic protein. So far, only a handful of similar cases are reported in literature. Important questions are raised concerning the safety of booster vaccination.

- Y06 -

USP53 GENE: THE NEW KID ON THE BLOCK CAUSING LOW GGT CHOLESTASIS. A. Ravindranath (1), R. Wadhwa (2), M. Wadhwa (3) / [1] Apollo BGS Hospital, Mysore, India, Pediatric Gastroenterology, [2] Apollo BGS Hospital, Mysore, India, Gastroenterology, [3] Accura diagnostics, Mysore, India, Pathology. Department

Case Report: A 14-year-old boy presented with history of pruritus since infancy for which he was being treated with topical medications at home. He also had history of recurrent wheeze for which he was receiving regular nebulisations and inhalational steroids. He developed jaundice with high colored urine for 2 months with worsening of pruritus. On preliminary evaluation his liver functions showed Total bilirubin: 18mg/dL, direct bilirubin: 12mg/dL, aspartate amino transferase: 248IU/L, alanine amino transferase: 467IU/L, total protein: 6.7g/dL, albumin: 3.8g/dL, alkaline phosphatase: 562IU/L, gamma glutamyl transpeptidase (GGT): 28IU/L. His prothrombin time and hemogram were normal. Anti HAV IgM, Anti-HEV IgM, Anti HBc IgM were negative. There was no history of intake of hepatotoxic medications. Serum bile acid level was 127mg/dL. Ultrasonogram of the abdomen showed coarse liver echoes, normal portal vein diameter and no ascites or splenomegaly. Esophagogastroduodenoscopy did not show any varices. He was born of second-degree consanguineous marriage. His father had history of pruritus and jaundice from adolescence. He developed decompensated liver disease at the age 32 years and died. Etiological work-up of father's liver disease was not available. Mother and two brothers are asymptomatic. Since he had low GGT cholestasis initial possibilities kept were progressive familial intra-hepatic cholestasis and bile acid synthetic defects. Since serum bile acid level was high bile acid synthetic defect was less likely. Liver biopsy showed bland cholestasis with mild periportal fibrosis. To confirm the etiology, exome sequencing was performed. The sequences obtained were aligned to human reference genome (GRCh38.p13) using Sentieon aligner and analyzed using Sentieon for removing duplicates, recalibration and re-alignment of indels. Sentieon haplotype caller was used to identify variants. A homozygous single base pair insertion in exon 10 of the USP53 gene (chr4:g.119268407_119268408insG; Depth: 111x) that results in a frameshift and premature truncation of the protein 5 amino acids downstream to codon 427 (p.Lys427GlufsTer5; ENST00000450251.5) was detected. On performing Sanger's sequencing in the mother, she was found to be heterozygous. The child was started on ursodeoxycholic acid and rifampicin sequentially. After 2 months of treatment pruritus subsided completely, liver functions improved. At 6 months of treatment, liver functions normalised. At a follow-up of 1 year child has gained weight of 3 kilogram and remained asymptomatic.
Discussion: Ubiquitin-specific peptidase 53 (USP53) is one of the recent genes described to be mutated in children with low GGT cholestasis. Although the exact function of USP53 is not known it is postulated that it interacts with Tight junction proteins 1 and 2 and regulates protein turnover by modulating deubiquitination. Mutated USP53 gene can affect tight junction scaffolding. Hence, USP53 mutated cholestasis has many overlapping features with that of TJP defect including wheezing and hearing loss. Since USP53 related cholestasis is only a recent addition to the set of low GGT cholestasis, long term outcomes are not definitely known. Many previously described reports have shown that the cholestasis is mild and self-limiting. In the given case, the child has been predominantly symptomatic with pruritus since infancy and visible jaundice appeared in adolescence. He has wheeze; hearing and speech are normal. Since