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LETTER TO THE EDITOR

Diagnosis of primary biliary cholangitis in a ''teen'' male patient

Sir,

We report the case of a young man (19-years-old) referred to our centre for the study of autoimmune liver disease because he had persistently high values of liver enzyme tests. His medical history was negative until two years before when an autoimmune thyroiditis was diagnosed, a substitutive therapy has been given and a regular follow up has been started. The hypothyroidism was well compensated and no autoimmune diseases were reported in his family history. At presentation his liver levels were as follows: gamma glutamil transpeptidase 321 U/L (normal limit < 61 U7L), alkaline phosphatase 254 U/L (normal limit < 130 U/L), serum aspartates aminotrasferase 43 U/L (normal value < 40) and serum alanino trasferase 83 U/L (normal value < 40), IgG 1446 mg/dL (normal value 700—1600) and IgM 422 mg/dL (normal value 40—230). All the other liver tests were normal (bilirubin, albumin, INR). All the causes of liver disease (viral, genetic, metabolic, drugs) have been rule out by appropriate tests. In particular deficit of alfa-1 antitrypsin and ceruloplasmin have been rule out, while the research for gene *ABCB* mutation was not performed because no reduction of MDR expression at biopsy level was revealed. Both the ultrasound examination and cholangio RM were normal, in particular they did not show any abnormality of biliary tree. At this regard, the patient underwent also to a ERCP. Under clinical point of view the patient was completely asymptomatic and neither fatigue or pruritus were reported.

The research for non-organ specific autoantibodies performed by indirect immunofluorescence revealed a seropositivity for anti-mithocondrial pattern on rat tissues and both multiple nuclear dots and rim-like/membranous patterns on HEp-2 cells; all these reactivities have been confirmed by a commercially liver blot (Euroimmun, Lubecca, Germany) that documented a AMA positivity and the seropositivity for both anti Sp100 and anti gp210. The liver biopsy showed a mild activity hepatitis with a porto-portal septum, initial bile duct injury and a portal granuloma; the histological picture was compatible with a low PBC stage (grading 7, staging 3 Ishak' score).

As shown in the detailed way all the major criteria (intrahepatic cholestasis, AMA and ANA specific PBC seropositivity and histological assessment) [1] were satisfied and the diagnosis was well-defined. Ursodeoxycholic acid therapy at 15 mg/kg has been started and after 1 year of therapy cholestatsis enzymes came back into the normal limit and the IgM serum levels were only slightly elevated (274 mg/dL); at this moment both cholestasis enzymes and IgM are within the normal ranges.

The calculation of liver stiffness by transient elastography after one year of therapy was similar to the diagnosis one (4.9 vs 5.2 kPa, respectively). The patient's follow up is continuing (to date the follow up is of 38 months) without any liver complication or sign of disease progression.

PBC is not a typical disease of young men, so our first suspect was addressed to a primary sclerosing cholangitis and we performed both cholangio RM and ERCP; only after the documentation of the normality of biliary tree the patient has been undergone to a liver biopsy while serological tests were running.

The peculiarity and the interest of our clinical report is related to the epidemiological setting; PBC is a characteristic autoimmune liver disease typically affecting the perimenopausal age; to this regard there is a very recent epidemiological study from Switzerland where the lowest age of diagnosis was 42 years [2]; the research for similar cases to ours revealed only two other reports where a PBC diagnosis has been made at similar age and in both cases the female gender was affected [3]; there is then an other case of interest where the patient, a three years female child with a diagnosis of type 2 AIH had a transient seropositivity for antimithocondrial antibody, without however the development of PBC [4]; furthermore in the past a controverse case of a newborn with a connatal liver damage and a subsequent detection of AMA positivity has been described [5]: here we described the most improbable combination under the epidemiological point of view: a ''teen'' male with a well-defined diagnosis of PBC; to our knowledge this is the first report of a diagnosis of PBC in a teen male patient, and on this basis in front of a chronic cholestatsis also in young men a PBC diagnosis has to be suspected.

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Disclosure of interest

The authors declare that they have no competing interest.

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