Research Square

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Association of Primary biliary cholangitis with idiopathic thrombocytopenic purpura

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Case Report

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Abstract

A 47 years old woman with seven years of pruritus and abdominal distension was diagnosed with primary biliary cirrhosis with chronic liver disease. A month after she developed bruises/petechie on hands and legs. The Platelet count abruptly increased with steroid therapy. She has multiple relapses of thrombocytopenia to a very lesser counts i.e. 6000.The association between these two conditions is rare and only few cases has been reported previously in Japan.

Conclusion: This case supports a rare autoimmune association between primary biliary cholangitis and idiopathic thrombocytopenic purpura after exclusion of viral, drug-related, hyper splenic, and marrow-related causes of thrombocytopenia.

INTRODUCTION

Primary biliary cirrhosis (PBC) is a condition in which biliary damage occurs as a result of an autoimmune process that affects both intra and extrahepatic bile channels. PBC is frequently related with other autoimmune conditions. It includes Polymyositis[1],Sjogren syndrome [2],autoimmune thyroid diseases [3] and autoimmune hepatitis [4]. Idiopathic thrombocytopenic purpura (ITP) is purely an autoimmune condition. The two conditions (PBC and ITP) are rarely associated, a few case reports has been published till now[5–7]. We present a case of an adult woman with Primary Biliary Cholangitis who remain undiagnosed for a long time, then developed thrombocytopenia. We will confirm their association after ruling out other possibilities.

CASE PRESENTATION

History:

In November 2023, a 47-year-old woman presented to us with a history of Pruritus for 7 years abdominal distension for a month, and bruises for 5 days. She does not have any history of infection/drug use.

Examination:

She had multiple bruises in pressure areas, injection sites, and hepatosplenomegaly.

Investigations:

Peripheral smear, liver biochemical tests, viral serology, platelet count, and autoimmune liver profile are summarized in Fig. 1. The serial platelet count response to treatment is shown in Fig. 2. Upper gastrointestinal endoscopy showed no varices (Fig. 3). Abdominal ultrasonography showed hepatomegaly with mildly echogenic liver texture, normal portal pressure, no splenomegaly, and no

ascites (Fig. 4). Bone marrow biopsy revealed normal cellular marrow with increased megakaryocytes, supporting peripheral platelet destruction.

Treatment:

Dexamethasone 8mg intravenously and Tab ursodeoxycholic 800mg orally daily were administered. Platelet counts were improved for some time but decreased to 8000 after a month. The shift from steroid to eltombopag was merely due to cirrhotic features. Eltrombopag 50mg was started for two weeks. Platelet counts were in normal range (graph 1) until the patient was unable to afford eltrombopag. Low-dose steroids (5mg prednisolone) (graph 1), Tab azathioprine 50mg per oral OD, and Tab Urso 400mg per oral BD were intiated. Alkaline phosphatase and platelet counts are near normal.

Follow up:

Patient is advised to screen for platelet counts every 2 weeks, liver function every 2 months and ultrasonography every 6 months. She does not require a liver transplant at the moment as MELD score is less than 12.

DISCUSSION

Primary biliary cholangitis is a disease that affects middle-aged females and is clinically diagnosed with > 1.5 times alkaline phosphatase, no biliary obstruction, and an AMA level more than 1:40.When other diseases exist, such as obesity, heavy alcohol consumption, or non alcoholic fatty liver disease, a biopsy is required. However, staging and prognosis of the illness remain important. Idiopathic thrombocytopenic purpura is a diagnosis of exclusion. Primary biliary cholangitis and autoimmune thrombocytopenic purpura is linked to autoimmune illnesses such as autoimmune hepatitis, Sjogren syndrome, and Hashimoto's thyroiditis. The relationship between the two disorders has already been described in Japan [5] and one case of Primary biliary cholangitis development after ITP [6].Few cases of idiopathic thrombocytopenic purpura, post-liver transplant has been reported (secondary to PBC) [7].

The association between HLA alleles and susceptibility to PBC and ITP has been studied because immunogenetic variables are thought to play a role in the development of autoimmune disorders [8–11].Platelet degradation by anti-glycoprotein I b/Ⅸ antibodies may contribute to ITP development. It has also been shown that autoantibodies from a patient with PBC and ITP react with glycoprotein IIb/IIIa of autologous and allogeneic platelets, bind to an epitope of rat 70-kDa mitochondrial protein M2 [12].PBC with thrombocytopenia was compared to controls, and more than half of the cases were related with anti-platelet antibodies (GP IIb/IIIa and GP I b/IX) using the MAIPA assay [13].

Our patient has responded abruptly to steroid therapy and doesn't have splenomegaly (a sign of cirrhosis) i.e. splenic congestion that would lead platelet destruction. Bone marrow results are equivocal with thrombocytosis and normal physiology of other cell lines. Unfortunately, we cannot perform immunogenetic factors due to limited resources. Written consent was taken from the patient and also an approval from ethical review committee of our institution.

CONCLUSION

1. Middle-aged female with generalized itching should be screened for Alkaline phosphatase levels and ultrasound hepatobilliary system.
2. More research is required for cheaper and more comprehensive treatment modalities for both conditions.
3. Today's research should be focused on immunogenic correlations between the two conditions, as it may help us in screening the condition beforehand.

SUMMARY

A 47-year-old female having a long history of itching has also developed bruises and abdominal distension. On investigation she has increased liver enzymes and thrombocytopenia. Antimitochondrial antibodies are strongly positive. Ultrasound reported an echogenic liver with a normal-sized spleen and no significant pathology on biopsy. Hence, her diagnosis of primary biliary cholangitis and Idiopathic thrombocytopenic Purpura is confirmed. She has responded to all of the treatment modalities well enough including steroids, eltrombopag, and ursodeoxycholic acid. It is reasonable to select drugs keepingin view both conditions. It is momentous to look for immunogenic associations of the conditions.

Declarations

Ethics Approval and Consent to Participate

This case report was reviewed and approved by the Institutional Review Board/Ethics Committee of Khalifa Gul Nawaz Teaching Hospital, Bannu, affiliated with Bannu Medical College, Bannu, Pakistan. Written informed consent to participate was obtained from the patient. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and all accompanying clinical information and images. A copy of the written consent is available for review by the Editor-in-Chief of the *Journal of Medical Case Reports* upon request.

Competing Interests

The authors declare that they have no competing interests.

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Author Contribution

Dr. Amjid Ali Khan contributed to the conception of the case report, clinical evaluation and management of the patient, supervision of the work, and critical revision of the manuscript.Asmat Saeed contributed to clinical data collection, literature review, drafting of the case presentation, and revision of the manuscript.Shad Muhammad Khan contributed to collection and organization of clinical information, review of investigations, literature review, and manuscript drafting.Ahmad Sanan contributed to literature review, manuscript writing, reference organization, and revision of the final draft.Muhammad Fayyaz contributed to clinical data review, interpretation of relevant investigations, literature review, and revision of the manuscript.Syed Muhammad Ahmad and Dr Abdul Muiz contributed to manuscript preparation, coordination among authors, editing, formatting, correspondence, and final revision of the manuscript.All authors read and approved the final manuscript.

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Availability of Data and Materials

All data relevant to this case report are included in the published article. Additional clinical information is not publicly available because of patient confidentiality but may be made available by the corresponding author on reasonable request, subject to institutional and ethical approval.

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Figures

LABORATORY INVESTIGATIONS

TESTS	VALUE	Normal Range	TESTS	VALUE	Normal Ra
Hb	11.3 g/dl	11.5-16	serum albumin	3.24 g/dl	3.6-5
MCV	90	80-101	cholesterol	280 mg/dl	10-200
TLC	9700 /cm^2	4-11k	HDL	19 mg/dl	35-100
Platelet	6000 /cm^2	150k-400k	LDL	144 mg/dl	0-150
Bilirubin	1.96 mg/dl	0-1	triglyceride	570 mg/dl	0-200
ALT	84 U/L	0-41	serum calcium	8.8 mg/dl	8.4-10.2
ALP	2613 U/L	40-265	vit D3	63 ng/ml	30-40
PT	15	14	HBV	-ve	
INR	1		IgA level	301 mg/dl	60-400
appt	36	34	IgG level	1255 mg/dl	700-160(
HCV	-ve	------	HIV	-ve	---------

AUTO IMMUNE LIVER PROFILE

Serological antibodies	Value	Normal Range
Anti AMA-M2 antibodies	103	05
Anti M2-3A antibodies	68	05
Anti LC-1 antibodies	0	05
Anti PML antibodies	2	05
Anti RO-52 antibodies	1	05
Anti SLA/LP antibodies	1	05
Anti Sp100 antibodies	0	05
Anti gp210 antibodies	2	05
anti-LKM antibody	3.01U/ml	12.00 U/ml

Urine R/E (pale yellow, acidic pH, glucose and protein Nil, pus cells and red cells 0-01/hpf.

*ANA, anti-nuclear antibody; Anti-LKM1, anti-liver-kidney microsomal antibody; Anti-LC anti-liver cytosol antibody type 1; Anti-PML, anti-promyelocytic leukemia antibody; Ant SLA/LP, anti-soluble liver antigen/liver-pancreas antibody; Anti-gp210, anti-glycoprotein-21 antibody

Figure 1

Laboratory investigations and autoimmune liver profile

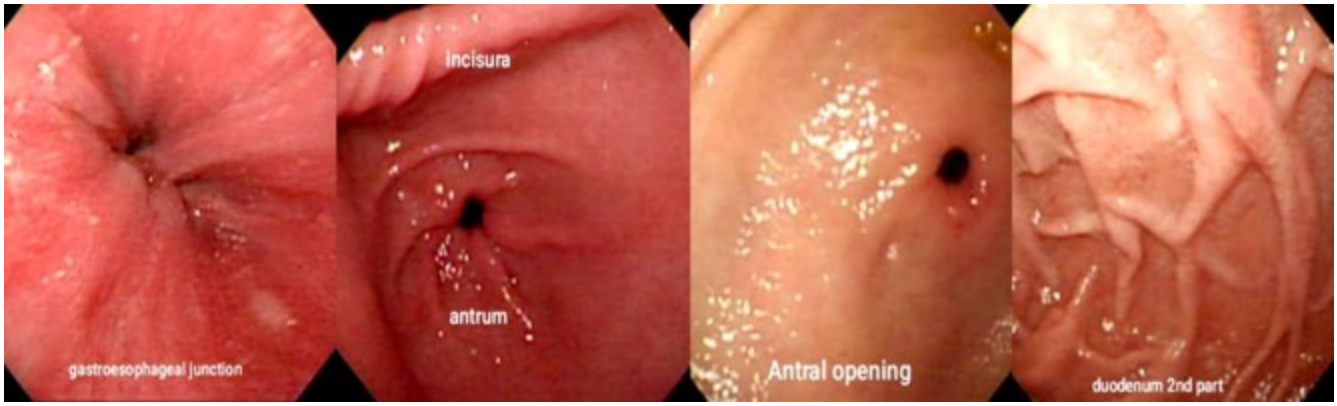


Figure 2

Effect of treatment on platelet count over time

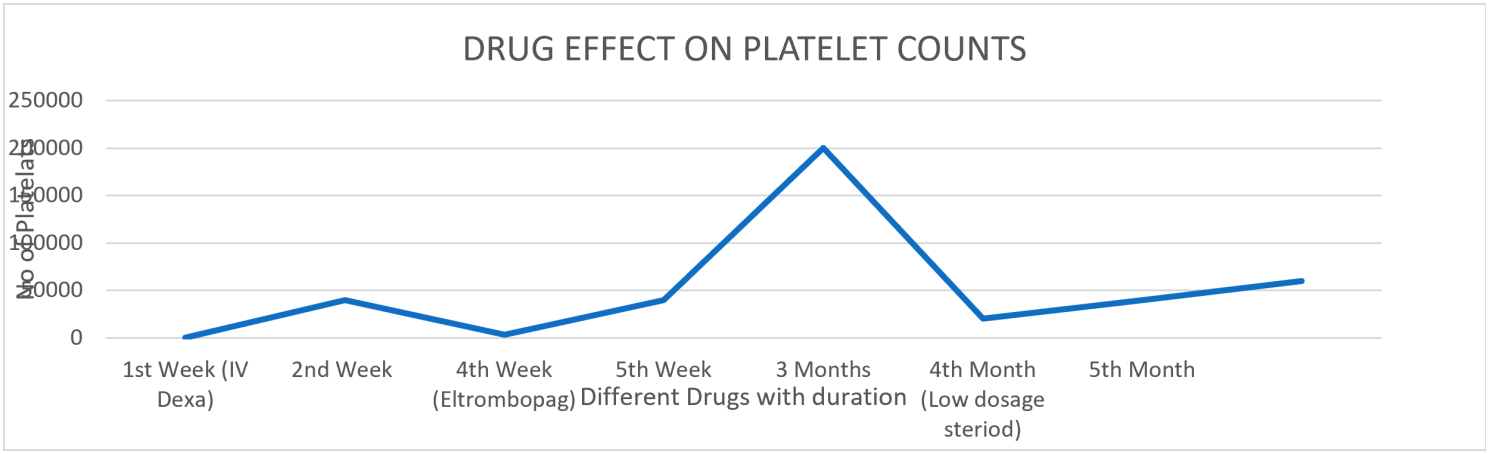


Figure 3

Upper gastrointestinal endoscopic findings

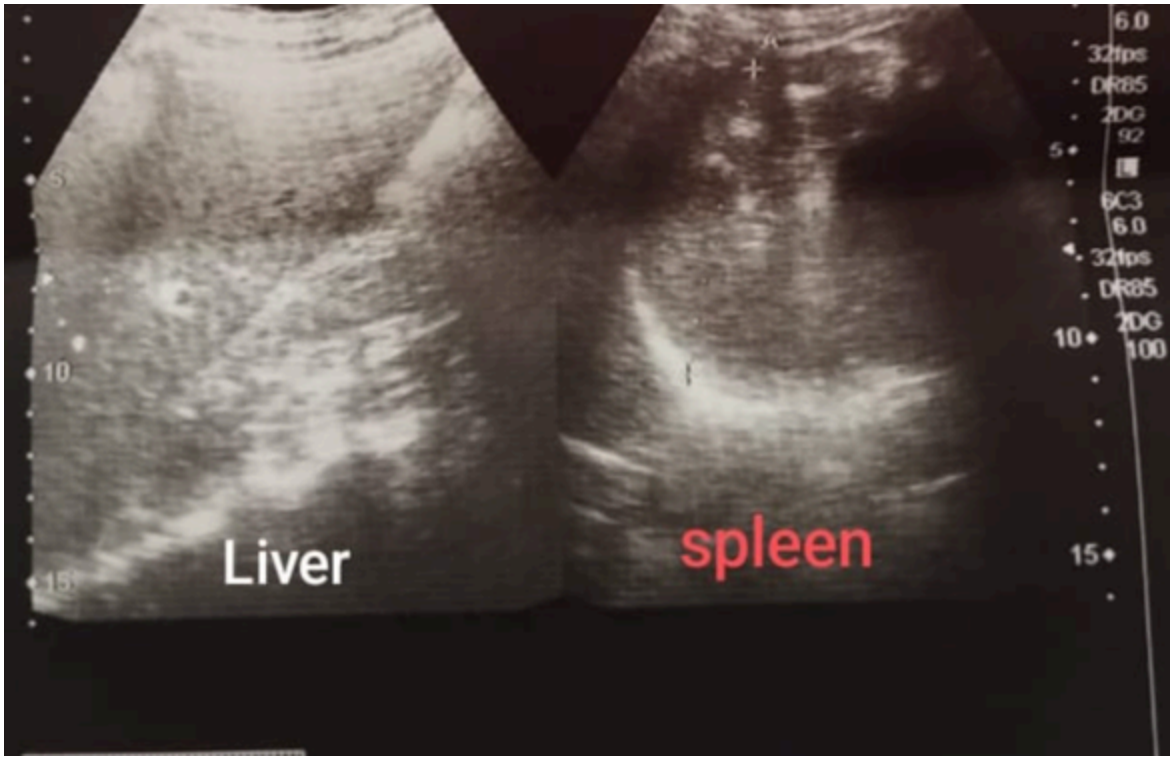


Figure 4

Ultrasonographic images of liver and spleen

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