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Concurrent autoimmune hemolytic anemia and immune thrombocytopenic purpura in a patient with budd-chiari syndrome post-direct intrahepatic portocaval shunt: A case report

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Abstract A rare liver condition known as Budd-Chiari syndrome (BSC) is characterized by hepatic vein blockage, which damages the liver. Immune-mediated hematological disorders that might exacerbate the clinical picture include immune thrombocytopenic purpura (ITP) and autoimmune hemolytic anemia (AIHA). In this case study, a patient with BCS who underwent a direct intrahepatic portocaval shunt (DIPS) operation developed AIHA and ITP. A 21-year-old female patient with Budd-Chiari syndrome and a straight intrahepatic portocaval shunt presents atypically in this case study. The case report details a patient's course of therapy, including diagnosis, preoperative planning, clinical testing, surgery, intensive care, and a successful recovery with appropriate postoperative care (Chandra et al., 2018).

Keywords: autoimmune hemolytic anemia, Budd-Chiari syndrome, intrahepatic portocaval shunt, immune thrombocytopenic purpura, hepatic vein obstruction, portal hypertension

1. Introduction

The important presence of immune thrombocytopenic purpura along with autoimmune hemolytic anemia coining the name Evans syndrome puts forth a whole lot of diagnostic and therapeutic hurdles. These two rare immune-mediated disorders carry a double complication in the clinical course when these are associated with some other underlying conditions like Budd-Chiari syndrome (Aydinli & Bayraktar, 2007).

The Budd-Chiari syndrome (BCS) is a rare vascular hepatic disorder due to obstruction of hepatic venous outflow. It usually presents with hepatomegaly, abdominal pain, and ascites. However, 20% of cases may be asymptomatic due to collateral circulation-forming channels (Aydinli & Bayraktar, 2007). This leads to delays in diagnosis. DIPS (direct-intrahepatic portocaval shunt) is an interventional procedure often performed to relieve portal hypertension and its complications in BCS (Chandra et al., 2018).

AIHA is characterized by autoantibodies directed against erythrocytes, which are responsible for hemolysis and anemia. Clinical symptoms of it include pallor, dyspnea, jaundice, and malaise. Diagnosis is by positive direct antiglobulin test or Coombs test an important diagnostic test founded in 1945 by Dr Robin Coombs to establish the autoimmune etiology of anemia (Hill & Hill, 2018).

In ITP, however, there is immune-mediated destruction of the platelets. The autoantibodies render platelets more susceptible to phagocytosis, predominantly in the spleen. The syndrome is associated with purpura, mucosal bleeding, and isolated thrombocytopenia. ITP is rather infrequent, with an annual incidence of 1 to 6 annual cases per 100,000 individuals, typically following viral infections in children and young adults (Aboud et al., 2017). The diagnosis is made clinically with findings of bleeding and low platelet counts and by ruling out secondary causes of thrombocytopenia (Jaime-Pérez et al., 2018).

This report presents a rare co-existing AIHA and ITP case in a 21-year-old female patient with a background of BCS managed by DIPS procedure-needs to highlight the diagnostic challenges, therapeutic modalities, and clinical outcome implications in this population environment, emphasizing the importance of multinational management (Bianco et al., 2021).

2. Case Presentation

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A 21-year-old female patient presented to the hospital, with complaint of bilateral lower limb swelling, left hypochondrium pain, and dark stools for two months. She also reported regular fevers that subsided with medication. Her medical history included Budd-Chiari syndrome, previously treated with trans jugular intrahepatic portosystemic shunt (TIPS) followed by a direct intrahepatic portocaval shunt (DIPS) (Chandra et al., 2018).

Clinical Examination:

On examination, the patient was seemingly pale in color due to a yellowish discoloration of eyes. Swelling of the face also appears to be noticeable. The vital signs indicated are normal limits:

- Temperature: 38.5°C.
- Blood pressure: 140/80 mmHg.
- Heart Rate: 110 bpm.
- Respiration Rate: 18 breaths/min.
- Oxygen Saturation: 98% on room air.

Previously reported medical history showed that she had been diagnosed with Budd-Chiari Syndrome (BCS) which was treated with tranjugular intrahepatic portosystemic shunt (TIPS) followed by a direct intrahepatic portocaval shunt (DIPS) (Aydinli & Bayraktar, 2007).

3. Investigations

The provisional diagnosis of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP) is based on laboratory tests that indicate severe anemia and thrombocytopenia. The diagnostic findings are summarized in Table 1 (Zanella & Barcellini, 2014).

Low RBC mass and macrocytes, ovalocytes, and tear-drop cells were found in the smear from peripheral blood (Hill & Hill, 2018). The Coombs test is direct and the indirect Coombs test is performed. Direct Coombs returns positive results to confirm AIHA.

Abdominal imaging shows evidence of hepatomegaly, distended gallbladder with cholelithiasis, and other findings suggestive of DIPS placement.

3. Imaging Studies

Cardiac Assessment: Two-dimensional echocardiography revealed normal sinus rhythm with normal valves, LVEF was at 60% with no clots or vegetation (Orloff & Johansen, 1978).

Abdominal and pelvic imaging: CECT showed hepatomegaly, cholelithiasis, a thickened hepatic colon wall, and free fluid in the pelvis (Figure 1) (Bianco et al., 2021).

Chest X-ray: Post-DIPS placement (Figure 2) (Chandra et al., 2018).

4. Treatment strategy

4.1. Medication regimen

The patient received treatment with immunosuppressive and corticosteroid therapies and adjunct treatments as follows:

- Methylprednisolone high dose IV and oral.
- Azathioprine (50 mg/d).
- Ursodeoxycholic acid and rifaximin oral.
- Blood and platelet transfusion to correct anemia and thrombocytopenia.
- Antibiotics: Piperacillin-tazobactam (4.545 g IV TDS for 3 days) and ceftriaxone (1 g IV BD).
- Supplementation with protein powder.

4.2. Gynecology Management

Irregular menstrual cycles made the gynecology evaluation suspect Hb electrophoresis sickling. She was given Pause-XT (tranexamic acid, tribasic calcium phosphate, menadione) for promoting coagulation and reducing bleeding (Berentsen & Sundic, 2015).

4.2. Supportive Measures

Patient educated regarding medication adherence and potential complications. Follow-up required, including imaging (MRI or ultrasound) for liver function and DIPS status (Zanella & Barcellini, 2014).

4.3. Differential Diagnosis

Excluded primary hematological malignancies and infections, using:

- Bone marrow aspiration and biopsy: No malignancy evident (Hill & Hill, 2018).
- Peripheral smear: No hemiparasites and other infectious causes found (Berentsen & Sundic, 2015).

4.4. Outcome and follow up

The patient reacted positively to treatment, as evidenced by increasing hemoglobin and platelets (Table 1). After treatment, she reported not having any more episodes of epistaxis or dark stools. There would still be follow-ups for the two: on disease recurrence and assessment of liver function (Jaime-Pérez et al., 2018).

Table 1 Blood investigation test report on admission and after treatment.

Sr No	Investigation	Observed Value (On Admission)	Observed Value (After treatment)	Reference value
1.	Hemoglobin	3.1 gram per deciliter	8.1 gram per deciliter	For men, 13.2 to 16.6 grams per deciliter. For women, 11.6 to 15 grams per deciliter.
2.	Mean corpuscular volume	79.2 femtoliters	91.3 femtoliters	Adult/elderly/child: 80-95 fL. Newborn: 96-108 fL.
3.	Mean corpuscular hemoglobin	21.2 picogram per cell	30.9 picogram per cell	27 to 31 picograms per cell
4.	Total red blood cell count	1.46 million red blood cells	2.9 million red blood cells	Male: 4.7 to 6.1 million cells per microliter (cells/mcL) Female: 4.2 to 5.4 million cells/mcL.
5.	Total white blood cell count	8300 per microliter	10200 per microliter	4,500 to 11,000 WBCs per microliter
6.	Total Platelet Count	0.07 per microliter	0.12 per microliter	150,000 to 450,000 platelets per microliter of blood.
7.	Hematocrit	11.6 % percentage	26.5 %	normal levels for men range from 41%-50%. For women, the normal range is slightly lower: 36%-44%.
8.	Urea	27 millimoles per liter	52millimoles per liter	1.8 to 7.1 mmol urea per liter
9.	Creatinine	0.5 milligram per deciliter	0.6 milligram per deciliter	0.7 to 1.3 mg/dL (61.9 to 114.9 µmol/L) for men and 0.6 to 1.1 mg/dL (53 to 97.2 µmol/L) for women
10.	Sodium	139 milliequivalents per liter	141 mEq/L milliequivalents per liter	135 to 145 milliequivalents per liter (mEq/L)
11.	Potassium	4.0 millimoles per liter	4.3 mmol/L millimoles per liter	Adult/elderly: 3.5-5.0 mEq/L or 3.5-5.0 mmol/L (SI units) Child: 3.4-4.7 mEq/L
12.	Alkaline phosphate	87 international units per liter	146 IU/L international units per liter	44 to 147 international units per liter (IU/L) or 0.73 to 2.45 microkatal per liter (µkat/L).
13.	ALT(SGPT) alanine transaminase (Serum Glutamic Pyruvic Transaminase)	15 units per liter	30 units per liter	7 and 56 units per liter.
14.	AST(SGOT) aspartate aminotransferase (serum glutamic-oxaloacetic transaminase)	26 international units per liter	47 IU/L international units per liter	5 to 40 units per liter of serum
15.	Total protein	5.1 grams per deciliter	4.7 G/dl grams per deciliter	6.0 to 8.3 grams per deciliter (g/dL) or 60 to 83 g/L.
16.	Albumin	2.6 gram per deciliter	2.6 g/dl grams per deciliter	3.4 to 5.4 g/dL
17.	Total bilirubin	8.3 micromole per liter	4.0 µmol/L micromole per liter	0.1 to 1.2 mg/dL (1.71 to 20.5 µmol/L)
18.	Bilirubin conjugated	4.7 milligram per deciliter	2.3 mg/dl	Less than 0.3 mg/dL (less than 5.1 µmol/L)
19.	Bilirubin unconjugated	3.6 milligram per deciliter	1.7mg/dl milligram per deciliter	0.2 to 0.8 mg/dL.
20.	Globulin	2.5 gram per deciliter	2.1 gram per deciliter	2.0 to 3.5 grams per deciliter (g/dL) or 20 to 35 grams per liter (g/L)
21.	Lactate dehydrogenase	561 units per liter	489 units per liter	135-214 units per liter

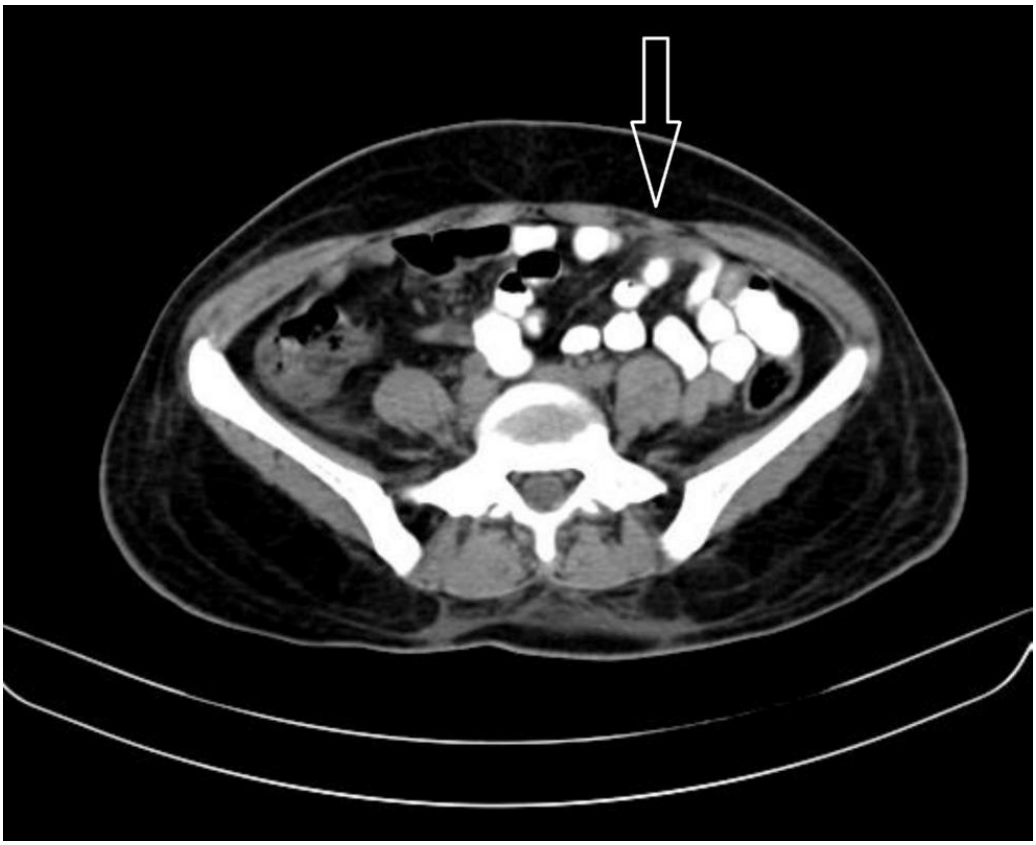

Figure 1 The image shows cholelithiasis with hepatomegaly.

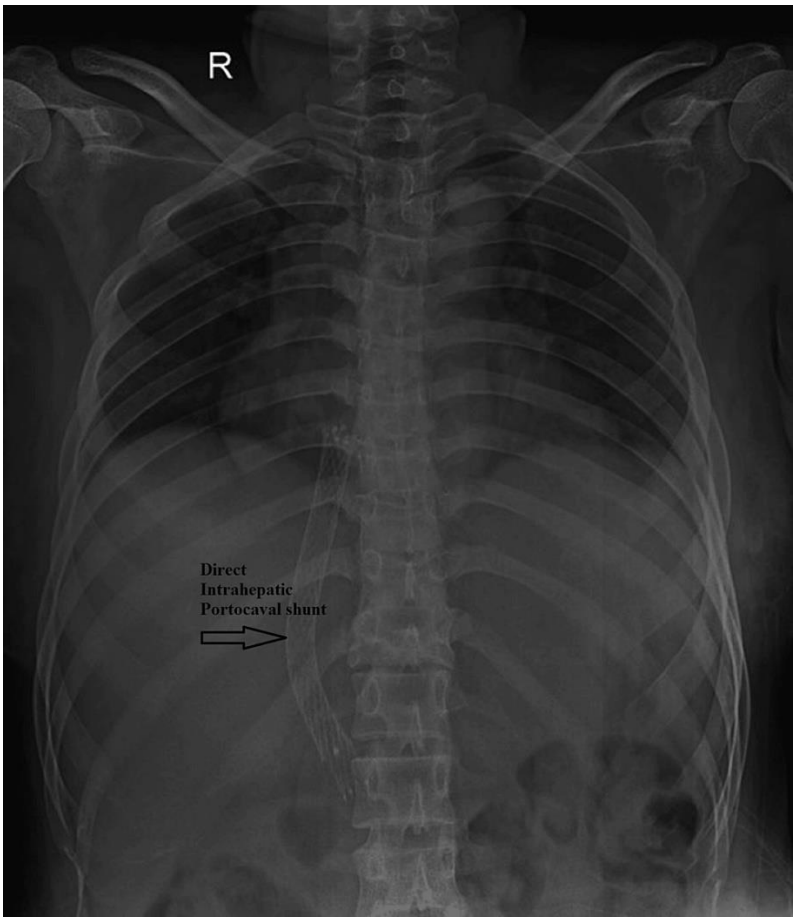

Figure 2 The image shows post-DIPS by IPS.

5. Discussion

AIHA and ITP are very rare autoimmune diseases of the blood, so there are sometimes tremendously confusing and troublesome diagnosis and therapy situations. (Hill & Hill, 2018). This case features the unusual coexistence of such conditions in a patient suffering from Budd-Chiari syndrome (BCS) following a direct intrahepatic portocaval shunt (DIPS) (Aboud et al., 2017).

AIHA is defined by the existence of autoantibodies to red blood cells, anemia, and hemolysis. It may occur either as primary disease or secondary to infections, other associated autoimmune disorders, or hematological malignancies. In contrast, ITP results from destruction of platelets through antiplatelet antibodies resulting in thrombocytopenia and a tendency for bleeding.

The patient presented with overlapping disease symptoms of AIHA and ITP after DIPS due to BCS. Budd-Chiari Syndrome (BCS) is brought about by hepatic venous obstruction and is known to lead to hepatomegaly, a congested liver, and sometimes liver failure when it is severe. The alteration of the immune response because of liver congestion and portal hypertension may make these patients susceptible to the development of autoimmune cytopenia's. This case proves how the liver dysfunction might relate to immune-mediated hematologic disorders (Aydinli & Bayraktar, 2007).

5.1. Role of DIPS Procedure with Immune Dysregulation

DIPS procedure is the favored strategy to relieve portal hypertension in BCS. The possible role of DIPS in triggering or worsening autoimmune conditions, however, remains unclear (Aydinli & Bayraktar, 2007). Literature demonstrates that vascular hepatic conditions, together with interventions like DIPS, may act to increase immune dysregulation in the pathogenesis for AIHA and ITP - autoimmune disorders (Chandra et al., 2018; Bayraktar et al., 2007).

5.2. The Management Approach

This case was managed multi-disciplinarily. High-dose corticosteroids and azathioprine suppress immune-mediated cytopenia's, IVIG therapy and transfusions per se stabilize hematologic parameters. The established treatment plan included dealing with the underlying hepatic dysfunction by means of anticoagulation and hepatoprotective agents. Such overall management emphasizes the importance of treating primary therapy in relation to the underlying hepatic pathology as well as secondary autoimmune manifestations (Zanella & Barcellini, 2014; Berentsen & Sundic, 2015).

5.3. Limitations and future perspective

There have been no genetic or immunologic investigations that potentially could have shown the underlying predispositions toward autoimmune disease; hence, this case report is limited. Another consideration would include long-term follow-up to ascertain if recurrence occurs in the future or if therapeutic outcomes are sustained. A further investigation is warranted to determine the immunologic implications of DIPS and similar interventions in patients with BCS (Jaime-Pérez et al., 2018).

6. Conclusions

This case is a single case report managing a 21-year-old female with Budd-Chiari syndrome (BCS) who developed concurrent autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP) following direct intrahepatic portocaval shunt (DIPS). The patient received blood and platelet transfusion, high-dose steroids, intravenous immunoglobulin (IVIG) therapy, and several other supportive measures. He showed clinical improvement, with the resolution of symptoms and stabilizing hematological parameters.

The complexity of her case rooted in treating all possible causes with a multi-disciplinary approach between hematology, hepatology, and supportive care teams was promising for her progress. Feedback from her was positive, satisfied with the overall experience with care, and thus reinforced the individualized management concept for such conditions. She had regular follow-up visits for liver function tests, monitoring for possible re-emergence of autoimmunity complications, and securing better long-term health outcomes.

It further reflects the need for close monitoring, early diagnosis, and individualized therapeutic strategies in complex cases involving autoimmune and hepatic diseases. It underscores the need for a multidisciplinary team in the management of overlapping rare disorders, as it optimizes outcomes (Aydinli & Bayraktar, 2007; Chandra et al., 2018; Hill & Hill, 2018).

Ethical considerations

Written informed consent was obtained from the patient. This study was conducted following ethical guidelines. No additional ethical approval was required.

Conflict of Interest

The authors declare no conflicts of interest.

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