

# An Ankylosing Spondylitis Case Which Accompanies Portal Hypertension and Lichen Amyloidosis

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**ABSTRACT** In this paper, we present a case that has been followed with Ankylosing Spondylitis (AS) for forty years and developed diabetes, iridocyclitis, lichen amyloidosis and hypertension over time. A 73-year-old male patient presented to our hospital with dyspnea and massive ascites with portal hypertension in the last one year. Cardiac and hepatic venous pressure measurements revealed mild pericardial thickening and intrahepatic sinusoidal portal hypertension in the liver. An infiltrative reason as metabolic, viral, autoimmune to develop portal hypertension was not found and systemic inflammation and vascular involvement due to AS was thought. TIPS was applied to the patient due to resistant ascites. After this operation, albumin was not needed, acid regressed and general condition improved. We aimed to present a very rarely seen association of AS, lichen amyloidosis and portal hypertension, and the curative result of TIPS operation in this case.

**Keywords:** Spondylitis, ankylosing; hypertension, portal; amyloidosis IX

Ankylosing spondylitis (AS) is an inflammatory disease that predominantly results in the loss of movement, which leads to vertebral eruption beginning before the age of 45 years. There are often (HLA) - B27 gene carriers. It usually begins with chronic back pain and holds one or more hinges or hinges around the vertebra. Different diseases such as uveitis (25-30%), psoriasis (10%), inflammatory bowel disease (IBD) (5-6%) can be added to this manifestations.<sup>1</sup>

In AS, which is a systemic inflammatory disease, cardiovascular complications (acute coronary syndrome, stroke, venous thromboembolism, pulmonary embolism, pericarditis, conduction disorder, aortic root and valve occlusion) can be seen besides vertebral and articular involvement. In addition, restrictive changes in the lung and renal amyloidosis in the kid-

neys, IgA nephropathy, non-specific glomerulonephritis may develop.<sup>2</sup>

Portal hypertension (PH) is a condition that develops with high pressure in the portal vein (normal pressure: 7 mmHg) and develops ascites, splenomegaly, porto-caval anastomosis and esophageal varices over time. There may be prehepatic causes leading to obstruction or narrowing in the vein port. The most important of the intrahepatic causes (>90%) is liver cirrhosis. In addition, idiopathic portal hypertension (IPD) should be considered in the differential diagnosis of intrahepatic presinusoidal PH. Posthepatic causes should be differentiated between vena cava inferior, right heart and pericardial pathologies.<sup>3</sup>

Lichen amyloidosis (LA) is the primary localized type of amyloidosis involving the skin. This type

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of fibrillary amyloid material only holds the skin without systemic arrest. The anterior aspect of the arms and legs is characterized by itchy hyperkeratotic papules on the back and hips. Although the cause of LA is unknown, degenerated keratin caused by chronic irritation of the skin is the return to amyloid material.<sup>4</sup>

## CASE REPORT

A 73-year-old male patient complained of abdominal distension, back and low back pain, itching, and dyspnea in March 2017.

The patient's anamnesis included history of 25 years of hypertension, 20 years of diabetes mellitus, 10 years of lichen amyloidosis, 5 years of iridocyclitis and hyperuricemia. He has been admitted due to increased abdominal distention (ascites) for one year. It was learned that he had undergone cholecystectomy ten years ago and he has no family history. On physical examination, paleness, vertebrae and chest motion limitation, marked kyphosis, in the leg, arm and back, widespread and marked lichen and itching marks, tethered free ascites in the abdomen, splenomegaly were seen.

The patient was further investigated for differential diagnosis of portal hypertension due to excess ascites and dyspnea as well as AS and LA. In laboratory tests; haemoglobin (g/dL) 8.9, platelets ( $\times 10^3/\mu\text{L}$ ) 285, serum albumin (g/dL) 2.1, C-reactive protein (mg/dL) 24, serum amyloid A protein (SAA): 8.50 mg/L (N: 0-7) and liver function tests were within normal limits.

Hepatic and portal vein color doppler: Liver parenchyma echogeneity increased heterogeneously. The diameter of the portal vein increased. The current is hepatopedal. Hepatic vein and intrahepatic branches were open. Normal hepatic vein findings.

Computed tomography of the abdomen: Liver contours show microlobulation. The portal vein was wide, and venous collaterals were observed around the splenic vein and around the stomach cardia. Common acid in the west.

Upper esophago-gastro-duodenoscopy: First degree of esophageal varicose veins.

Heart and hepatic vein catheterization was performed for the differential diagnosis of constrictive pericarditis and portal hypertension: Supererated hepatic vein wedge pressure: 24 mmHg, inferior hepatic vein wedge pressure: 21 mmHg, right atrium pressure: 8 mmHg, pulmonary artery mean pressure: 26 mmHg. Conclusion: Intrahepatic sinusoidal portal hypertension + mildly constrictive pericarditis.

The patient who did not respond to diuretic treatment and high volume paracentesis and required continuous albumin replacement was administered TIPS. The first 24-48 hour follow-up of the patient was performed in the hospital. "Speed 100 / sec, Stent Potent" was reported in the control.

In this period, the patient had a temporary increase in serum ammonia level and mild coma symptoms (grade 1). With the necessary medical follow-up and albumin support. The patient's general condition improved after 48 hours. During the first month of follow-up, he needed paracentesis once a week. Sufficient diuresis was provided with diuretic treatment. The serum albumin level gradually increased and albumin replacement was not needed after the first month.

The patient is still followed up with aldactazide 50 mg daily and furosemide 20 mg twice a week.

An informed written consent was obtained from the patient for reporting this case.

## DISCUSSION

The case, who had been treated with AS for 40 years and had been suffering from LA, iridocyclitis, hypertension and diabetes for 10 years, was hospitalized due to portal hypertension and resistant acid which became prominent especially in the last year.

The patient was primarily investigated for the etiology (prehepatic, hepatic and posthepatic) of portal hypertension. No prehepatic cause was found. The etiology of viral (HBV, HCV), metabolic (hemochromatosis, Wilson), autoimmune, steatohepatitis and infiltrative disease (amyloidosis) which could lead to chronic liver disease were investigated. Viral and autoimmune signs were negative. Hemochromatosis and Wilson disease were excluded. Liver biopsy

could not be performed due to excessive ascites and risk of bleeding.

It was thought that AS is a systemic inflammatory disease and especially venous vessels and venous thromboembolism can cause venous thromboembolism. This is confirmed by the fact that long-term liver tests, especially ALT and AST, and the number of PLTs are normal and that no coma symptoms and high ammonia levels are seen despite excessive portal pressure elevation. Increased inflammatory and subsequent fibrotic progression was thought to lead to coarse liver.<sup>5,6</sup>

As a result, LA and portal hypertension are a rare association in AS, which is a systemic inflammatory disease. In this case, in addition to the actual disease, TIPS application for the treatment-resistant acid was successful and there was no need for albumin and paracentesis. It was found appropriate to report the case.

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#### Conflict of Interest

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

#### Authorship Contributions

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## REFERENCES

1. Reveille JD, Weisman MH. The epidemiology of back pain, axial spondyloarthritis and HLA-B27 in the United States. *Am J Med Sci.* 2013;345(6):431-6. [[Crossref](#)] [[PubMed](#)] [[PMC](#)]
2. Stolwijk C, Essers I, van Tubergen A, Boonen A, Bazelier MT, De Bruin ML, et al. The epidemiology of extra-articular manifestations in ankylosing spondylitis: a population-based matched cohort study. *Ann Rheum Dis.* 2015;74(7):1373-8. [[Crossref](#)] [[PubMed](#)]
3. Bloom S, Kemp W, Lubel J. Portal hypertension: pathophysiology, diagnosis and management. *Intern Med J.* 2015;45(1):16-26. [[Crossref](#)] [[PubMed](#)]
4. Fernandez-Flores A. Cutaneous amyloidosis: a concept review. *Am J Dermatopathol.* 2012;34(1):1-14. [[Crossref](#)] [[PubMed](#)]
5. Sato Y, Sawada S, Kozaka K, Harada K, Sasaki M, Matsui O, et al. Significance of enhanced expression of nitric oxide synthases in splenic sinus lining cells in altered portal hemodynamics of idiopathic portal hypertension. *Dig Dis Sci.* 2007;52(8):1987-94. [[Crossref](#)] [[PubMed](#)]
6. Morikawa H, Tamori A, Nishiguchi S, Enomoto M, Habu D, Kawada N, et al. Expression of connective tissue growth factor in the human liver with idiopathic portal hypertension. *Mol Med.* 2007;13(5-6):240-5. [[Crossref](#)] [[PubMed](#)] [[PMC](#)]