e - ISSN - 2349 - 8005

INTERNATIONAL JOURNAL OF ADVANCES IN CASE REPORTS

Journal homepage: www.mcmed.us/journal/ijacr

ENIGMATIC EVOLUTION OF THE ASSOCIATION OF TUBERCULOSIS PERITONITIS AND PRIMARY AMYLOIDOSIS

Haider A Omer[1], Mohammed Assiri[2], Mohammed A Alzahrani[3], Mohammed Y Alasmary[4], Abdullah S Alghamdi[5], Awad M Al-Qahtani*[6]

[1]Assistant Professor and Consultant Nephrologists, King Khalid University, Abha, Saudi Arabia.
[2]Resident in Internal Medicine, Aseer Central Hospital, Abha, Saudi Arabia.
[3]Assistant Professor and Medical Consultant, King Khalid University, Abha, Saudi Arabia.
[4]Demonstrator Najran University, Medical Department, Internal Medicine Program, ACH Abha, Saudi Arabia.
[5]Professor and Consultant Cardiologists, King Khalid University, Abha, Saudi Arabia.
[6]Senior Registrar of Family Medicine, Department of Family& Community Medicine, Najran university, Saudi Arabia.

Corresponding Author:- **Awad Al-Qahtani, SBFM**
E-mail: dr.awad2009@hotmail.com

Article Info
Received 01/12/2015
Revised 04/12/2015
Accepted 08/01/2015

Key words:
Tuberculosis, Peritonitis, Amyloidosis, Enigamatic Evolution.

ABSTRACT
Amyloidosis is a group of pathologies characterized by extracellular deposition of fibrillar proteins having a beta-pleated sheet tertiary structure. The disease can be categorized as primary or secondary. Secondary amyloidosis can occur as sequel of chronic inflammation such as Tuberculosis. However, this is not the case with our patient. We report a 47 years old female patient who was admitted to our hospital for investigation for ascites and nephrotic range proteinuria. A diagnosis of TB peritonitis was made and anti-tuberculosis was started. After the completion of anti-TB course, the nephrotic syndrome did not improve. Several investigations were done for nephrotic syndrome and histological examination of renal tissue revealed AL amyloidosis. Coexistence of TB peritonitis and primary amyloidosis must also be considered in such cases.

INTRODUCTION

Amyloidosis is a rare group of pathologies characterized by extracellular deposition of fibrillar proteins having a beta-pleated sheet tertiary structure. Amyloid proteins are abnormal proteins that the body cannot break down and recycle, as it does with normal proteins. When amyloid proteins clump together, they form amyloid deposits.

The buildup of these deposits damages a person's organs and tissues. Amyloidosis can affect different organs and tissues in different people and can affect more than one organ at the same time. Amyloidosis most frequently affects the kidneys, heart, nervous system, liver, and digestive tract. The disease can be categorized as primary or secondary. Secondary amyloidosis can occur as sequel of chronic inflammation such as Tuberculosis [1-5].

CASE REPORT

This study describes 47 years old, female patient with easy fatigability, and vague abdominal pain and distension of one month duration. Examination revealed emaciated, patient with ascites, enlarged liver and bilateral lower limb edema. Laboratory findings included: protein urea (24 hours urine protein: 3.6 gm/day), hypochromic microcytic anemia (HB: 9.4); low SAAG (ascites); high serum cholesterol (247 mg/dl); triglyceride levels (219 mg/dl); serum creatinine (0.7 mg/dl); serum albumin (1.6 gm/dl); blood urea nitrogen (25 mg/dl); Na (133 mg/dl); and Ca levels (6.3 mg/dl). Ultrasonography revealed hepatomegaly and moderate ascites. She underwent laparoscopic laboratomy and peritoneal biopsy showed cazeating granuloma. TB peritonitis was confirmed and anti-TB was started. After the completion of anti-TB course, the nephrotic syndrome did not

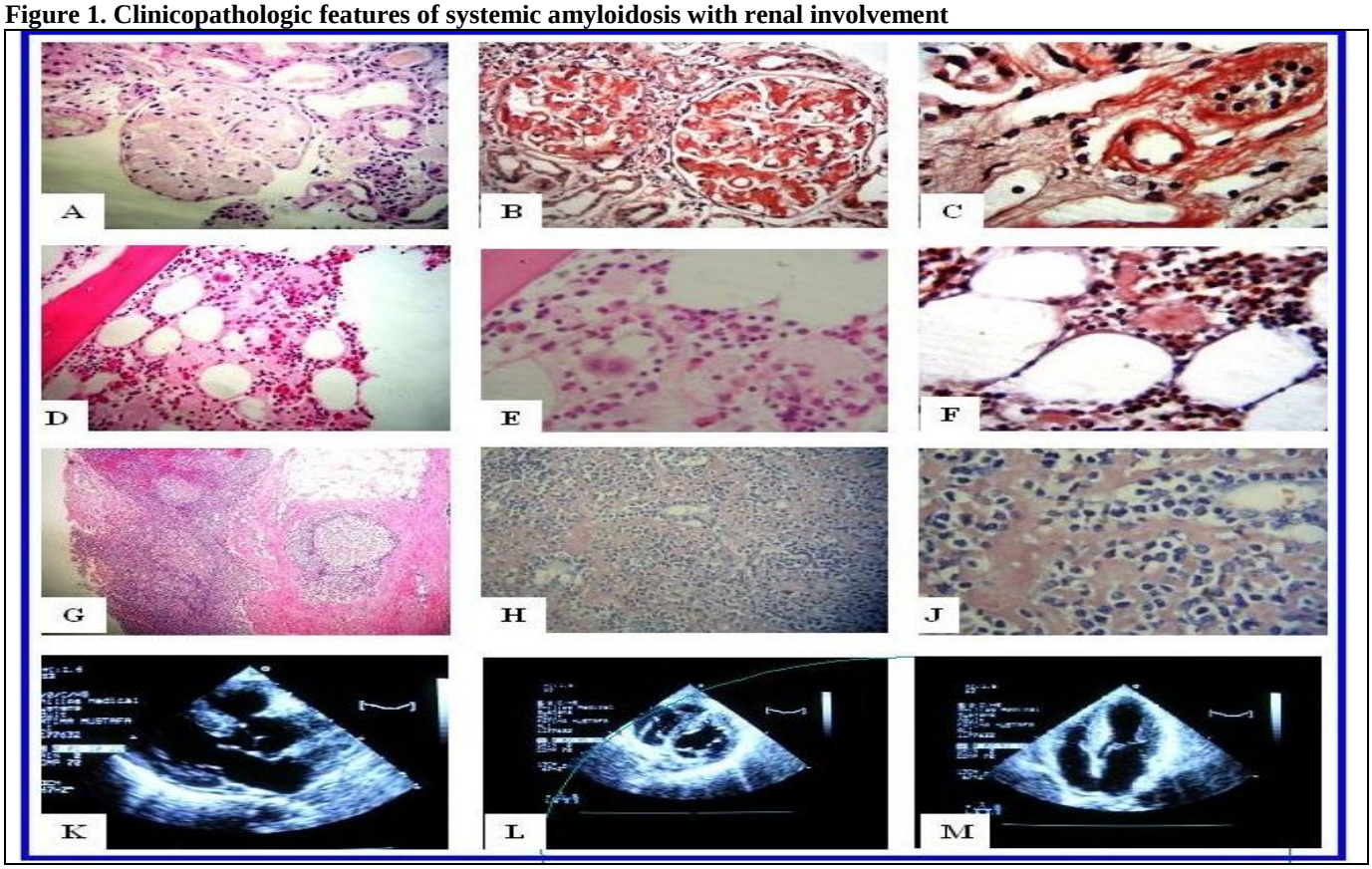
improved. Several investigations were done for her and the findings included: i) the cardiac biomarker NT-pro-BNP was grossly high pathognomonic of Al-amyloidosis; ii) sensitive serum free light chain assay showed significant LAMBDA excess 376 mg/L (NR 10-26); iii) Echo findings were compatible with cardiac amyloidosis and; iv) renal biopsy findings were consistent with amyloidosis. The diagnosis of nephrotic syndrome associated with extensive AL-amyloidosis was established. The general condition of the patient and the renal functions deteriorated rapidly and the patient expired within 18 months.

DISCUSSION

Amyloidosis is a rare disease and characterized by extracellular deposition of fibrillar proteins having a beta-pleated sheet tertiary structure. The resistance of amyloid material to digestion causes it to accumulate within tissues. It also interferes with their normal physiological functions, and destroys several organs. Amyloid is composed of a fibrillary protein and nonfibrillary glycoproteins. The nonfibrillary glycoprotein comprises amyloid P component (serum amyloid P, SAP), glycosaminoglycans, and apolipoprotein E. Serum amyloid P contributes to stability of amyloid deposits. The radioactive iodine labeled SAP is used to assess amyloid deposition [3,4].

Renal amyloidosis causes nephrotic syndrome and may be either a part of primary or secondary amyloidosis. The primary amyloidosis is called AL type because the amyloid is composed of monoclonal light chains (usually lambda). Only 10% to 20% of these patients have overt myeloma; the remainders have only a monoclonal spike in the serum and/or urine 1. About 40% of patient with AL type develop nephrotic syndrome. The poor survival in primary amyloidosis is due to development of acute leukemia or secondary malignancies. The secondary amyloidosis is called AA type. It occurs in patients with long-standing chronic inflammatory diseases such as tuberculosis or neoplasms. The main amyloid component is protein A, derived from proteolytic cleavage of serum amyloid A protein. 90% of patients have renal insufficiency or nephrotic syndrome at diagnosis. There are other forms of amyloidosis such as Beta 2 microglobulin type that is associated with long-term hemodialysis or peritoneal dialysis [3-5].In the view of active TB peritonitis and primary amyloidosis, a causal relationship is not defined. So, coexistence of TB peritonitis and primary amyloidosis must also be considered in such cases.

Figure 1. Clinicopathologic features of systemic amyloidosis with renal involvement



A-C): renal amyloidosis with deposition of salmon orange amorphous (A, X 200, H&E stain), Congo red positive deposits (B-C, X400, Congo red stain) in mesangium and subendothelium that obliterate glomeruli; also deposits in blood vessel walls, interstitium and around tubules ;
D-F): systemic amyloidosis with deposition of amyloid (D, X 200 and E, X400, H&E stain), (F, X400, Congo red stain) amid bone marrow elements ;
G-J): lymph node with deposition of amyloid (G-H, X 200, H&E stain), (J, X400, Congo red stain) amid lymphoid cells in the paracortex; and
K-M): Echocardiographic findings revealed sprinklings in the left ventricular wall with mild diastolic dysfunction.

REFERENCES

1. Falk RH, Skinner M. (2000). The systemic amyloidoses: an overview. *Adv Intern Med*, 45, 107-137.
2. Beers MH, Porter R, eds. (2006). The Merck Manual of Diagnosis and Therapy. 18th ed. Whitehouse Station, NJ: Merck Research Laboratories, 1310-12.
3. Falk RH, Comenzo RL, Skinner M. (1997). The systemic amyloidoses. *N Engl J Med*, 337, 898–909.
4. Gillmore JD. (1997). Amyloidosis: A review of recent diagnostic and therapeutic developments [Review]. *Br J Haematol*, 99, 245–256.
5. Kyle RA, Gertz MA. (1995). Primary systemic amyloidosis: clinical and laboratory features in 474 cases. *Semin Hematol*, 32, 45–59.