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Regression of Nephrotic Syndrome with Colchicine Therapy Secondary to Amyloidosis with Associated Castleman's Disease

Dear Sir,

This report describes a patient with Castleman's disease (CD) of mixed hyaline vascular type at the mesentery of the jejunum associated with systemic amyloidosis. Potassium permanganate-sensitive amyloid deposition was found on the vascular walls of the rectal mucosa and mesenteric lymphoid mass. After surgical resection of the mesenteric mass, nephrotic syndrome (NS) continued. With colchicine therapy, clinical and laboratory manifestations improved. She is asymptomatic and disease free for 23 months.

CD was first described in detail by Castleman et al. [1] in 1956 and is also known as lymph node hamartoma, angiofollicular lymph node hyperplasia and giant lymph node hyperplasia [2].

Case Report

A 28-year-old woman was admitted to our outpatient clinic in November 1991 with a 7-month history of edema and ascites. She had been operated in another hospital 5 months previously for an intra-abdominal mass. Splenomegaly and lymphoadenopathy (4 × 4 × 4 cm) (LAP) were found at laparotomy and LAP was resected together with the jejunal loop. Ascites, edema and incisional cicatrix were found on physical examination. She reported 5 abortions in her past history.

Laboratory Findings

Routine hematological and biochemical tests were within normal limits except for hypoalbuminemia (2.1 g/dl), hyperlipidemia (i.e. total lipid 1,290 mg/dl), and proteinuria (10.5 g daily). Protein electrophoresis revealed albumin 44.9%, α_1-globulin 3.99%, α_2-globulin 22.5%, β-globulin 13.4%, and γ-globulin 15.3%. Lipid cylinders were found in the urinary sediment and the creatinine clearance was 162 ml/min. Antinuclear antibodies, rheumatoid factor and cryoglobulins were negative. IgG, C_3, PT, PTT, chest X-ray, ECG, and echocardiography were normal. Serologic tests did not show evidence of infection including EBV, HSV, CMV, HIV, HBV, *Salmonella*, *Brucella* and *Listeria*. Bone marrow aspiration and biopsy were normal and did not show amyloid deposits. Marrow as well as ascitic fluid cultures were negative for bacteria, mycobacteria and fungus. Bence Jones proteinuria was negative. Splenomegaly and ectopic right kidney were detected ultrasonographically and tomographically. Upper gastrointestinal radiography was found to be normal. Rectoscopy was negative and amyloid deposits were detected in a rectal biopsy and which was also sensitive to potassium permanganate. Lymph node morphology was reevaluated and we found a diffuse follicular pattern with the characteristic features of CD, and eosinophilic, amorphous material which stained positive with Congo red and was potassium permanganate sensitive.

The patient had severe proteinuria at admission and NS continued despite excision of the mass. Colchicine was given 1.5 mg daily. Nine months later, the urinary protein and serum albumin levels were found to be 1.5 g/day and 3 g/dl, respectively and she was pregnant. Colchicine was stopped. Her pregnancy was uneventful and she delivered a healthy full-term infant. Renal function was normal and daily proteinuria was less than 500 mg after the birth and at the last evaluation proteinuria was negative. She is disease free for more than 23 months. Repeated rectal biopsy sampling was negative for amyloid deposition.

Discussion

CD is a reactive process as judged by preservation of the lymph node architecture [3], polyclonal increase in plasma cells [4] and the benign course in most cases. It is not surprising that there are many systemic complications particularly when plasma cell infiltration is marked.

The renal pathology associated with CD is heterogeneous including minimal change disease, membraneous, mesangial-proliferative, cresentic, membranoproliferative glomerulonephritis, interstitial nephritis, and amyloidosis [5]. Proteinuria, hematuria or renal failure are the most frequent renal findings.

It is known that AA amyloidosis is associated with recurrent infections or some chronic inflammatory diseases, FMF, and some neoplasms. The therapy for CD is unknown and tumor resection is generally an accepted therapy and some chemotherapeu-

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ticals such as prednisolone, vincristine and melphalan are indicated in patients with multicentric disease. Tumor resection generally causes a regression of the systemic symptoms [6]. Colchicine was given as treatment in our case. After 9 months of colchicine therapy she was pregnant and edema free.

In conclusion, we diagnosed CD and AA amyloidosis in a patient with NS. NS disappeared with colchicine therapy. Although the effect of surgical resection of the mass on the regression of NS could not be excluded, colchicine was probably highly effective in our patient. Our case, which had had 5 abortions previously, delivered a full-term liveborn infant. It opened the question whether CD can be a cause at early abortion? She is well and disease free for 23 months.

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