A Case of Cardiac Light Chain Deposition Disease in a Patient with Solitary Plasmacytoma

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Conflict of interest: None declared

Patient: Male, 31
Final Diagnosis: Light chain deposition disease
Symptoms: —
Medication: —
Clinical Procedure: None
Specialty: Hematology

Objective: Rare co-existence of disease or pathology
Background: Light chain deposition disease is a systemic disease characterized by deposition of immunoglobulin light chains in various organs. Cardiac involvement of light chain deposition disease, also known as cardiac nonamyloidotic immunoglobulin deposition disease (CIDD), is a rare clinical entity, where clinical outcome is very variable and best treatment approaches are not well known.

Case Report: We present the case of a 31-year-old man with a solitary thoracic plasmacytoma and cardiac light chain deposition disease with evidence of congestive heart failure by echocardiography and cardiac markers. The patient underwent surgical resection of the plasmacytoma followed by systemic therapy with 50% VDT-PACE and then VRD with near-normalization of his heart function. A melphalan-based stem cell transplant is planned in this young patient to achieve the best possible long-term remission.

Conclusions: CIDD is a very rare disease, with previous reports showing diverse manifestations with variable outcome. A high level of clinical suspicion should be maintained in such cases and early intervention, as in our patient, can restore cardiac function. There is very little literature on the optimal management of these patients. A combination of surgery and chemotherapy were pursued in our patient with very good results.

MeSH Keywords: Antineoplastic Combined Chemotherapy Protocols • Heart Failure • Immunoglobulin Light Chains • Plasmacytoma

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/895762

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Background

Light chain deposition disease (LCDD) is a rare disorder associated with clonal proliferation of plasma cells or B lymphocytes, which synthesize abnormal monoclonal immunoglobulin light chains. Cardiac involvement [also termed cardiac nonamyloidotic immunoglobulin deposition disease (CIDD)] is characterized by immunoglobulin deposition in the myocardium and is a very rare entity with variable outcome. We report a patient with thoracic plasmacytoma who on further work-up was found to have light chain deposition disease of the heart manifested by decreased ejection fraction and increased NT-proBNP levels.

Case Report

A 31-year-old man was found to have an incidental pleural effusion on a routine chest X-ray obtained as a part of pre-employment screening. Further evaluation with computed tomography of the chest and magnetic resonance imaging of the thoracic spine showed a 10×7×4 cm lesion in the para-vertebral and prevertebral soft tissue, extending from T4 to T8 (Figure 1) with prominent erosion and destruction of the vertebral bodies (Figure 2), and with moderate-sized pleural effusion on the right side. CT-guided biopsy of the para spinal mass was consistent with plasma cell neoplasm (Figure 3 depicts sheets of plasma cells that stain for CD138 in Figure 4). No further lesions were detected on PET CT and diffusion-weighted image MRI.

The blood cell counts were as follows: hemoglobin 14.7 gm/dl, white blood count 6980/µL, and platelet count 264 000/µL. Biochemical assay includes a fasting blood sugar of 96 mg/dL, serum sodium 141 mEq/L, potassium 3.9 mEq/L, calcium 9.3 mg/dL, phosphorus 4.1 mg/dL, BUN 21mg/dL, creatinine 0.9 mg/dL total protein 7.5 g/dL, albumin 4.1 g/dL, NT-proBNP 5213 pg/mL, LDH 181IU/L, and beta -2 microglobulin 2.3 mg/L. Immunochemistry findings included IgG 1570 mg/dL with Kappa 3.45 mg/dl, lambda 13.65 mg/dl, and a monoclonal protein of 0.7 g/dL. IgA and IgM were in the normal range, with 307 mg/dL and 114 mg/dL, respectively. Urinalysis showed 300 mg/dL of proteinuria and urine electrophoresis showed 960 mg/dL of proteins with 81% albumin and negative monoclonal protein. Serum and urine immunofixation showed IgG lambda paraproteinemia.

Routine cardiac work-up prior to chemotherapy was performed; electrocardiography (ECG) showed regular sinus rhythm and left atrial enlargement with no specific T wave changes in the inferior lead. The echocardiography revealed ejection fraction of 35% with moderate diffuse hypokinesis with left ventricular concentric hypertrophy with decreased diastolic compliance consistent with restrictive cardiomyopathy. There was minimal pericardial effusion without any evidence of constriction. Cardiac MRI showed systolic dysfunction and grade III diastolic dysfunction. Left ventricular concentric hypertrophy was noted with marked enlargement of the left atrium, mild thickening of the right ventricle, and increased inter-atrial septal thickness. A patchy non-territorial nulling abnormality of the
myocardium was also noted. Cardiac biopsy of the septum showed diffuse linear lambda light chain staining by immuno-
fluorescence around each muscle fiber, with no amyloid depo-
sition by Congo red and thioflavin-t stains confirming the diag-
nosis of light chain deposition disease lambda type (Figure 3).
Bone marrow examination showed no morphological or im-
munophenotypic evidence of plasma cell neoplasm or amyloid
deposition. Lymph node biopsy did not show any evidence of
amyloidosis or light chain deposition disease. The fat pad bi-
opsy was negative for amyloid. Cytology from the pleural ef-
fusion was negative for malignancy.

The patient had to undergo T1–T11 posterior fusion with re-
section of the plasmacytoma at T6 level for concerns of spi-
nal instability. Histopathological examination demonstrated
sheets of atypical plasma cells infiltrating the soft tissue and
the bone consistent with plasma cell neoplasm (Figures 4, 5
showing CD138 stain). Follow-up cardiac evaluation with re-
peat echocardiogram after surgical removal of his plasmacy-
toma showed restoration of his EF to 55% with improvement
diastolic function from Grade III to Grade II. NT-proBNP lev-
els decreased from 5213 pg/dL to 1300 pg/dL. Further evalua-
tion showed a reduction of monoclonal protein from 0.7 g/dl
to 0.4 g/dl and in Lambda light chain from 13.65 mg/dL to
4.38 mg/dL. Given the size of the plasmacytoma and persis-
tent residual disease after surgery, we decided to start ag-
gressive systemic therapy in this young patient to achieve
best long-term disease control. The patient was subsequent-
ly treated with 50% VTD-PACE (Velcade 1 mg/m², thalido-
mide 200 mg D1–4, dexamethasone 40 mg po D1–4, cisplatin
5 mg/m² D1–4, adriamycin 5 mg/m² D1–4, cyclophosphamide
200 mg/m² D1–4, and etoposide 20 mg/m² D1–4), followed
by stem cell collection. He was subsequently started on VRD
(Velcade 1 mg/m² weekly, Revlimid 25 mg D1–D14 out of 21
days, and Dexamethasone 20 mg weekly). Four months after
initiation of systemic treatment, the patient’s EF remains sta-
ble at 55% and his NT-proBNP has improved significantly to
207 pg/mL. Monoclonal paraproteinemia has resolved and im-
munofixation in serum and urine is negative. Proteinuria has
decreased significantly from 960 mg/dL to 180 mg/dL, with
all of the protein being due to albuminuria. For best long-term
results, the plan is to proceed to a Melphalan-based stem cell
transplantation followed by maintenance. The presentation
is consistent with IgG lambda solitary thoracic plasmacy-
toma and nonamyloidotic light chains deposition disease of the
heart, which is now almost completely reversed after surgical
removal of his primary plasmacytoma and systemic therapy.

**Discussion**

LCDD is a disorder described in association with abnormal clonal
proliferation of plasma cells or B lymphocytes, which synthesize
abnormal monoclonal immunoglobulin light chains. This dis-
ease is rare and most often associated with kappa light chain
excess. In heart biopsies, main differential diagnosis is amyloid

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**Figure 3.** Immunofluorescence staining of the myocardium showing diffuse linear lambda light chain around each muscle fiber.

**Figure 4.** Sheets of atypical plasma cells.

**Figure 5.** CD138 staining of the plasmacytoma.
Cardiac involvement is very rare and occurs about 20% of LCDD [11]. Outcome of CIDD and its response to treatment are not well known, with some reports suggesting reversibility [12] and others showing aggressive disease with no response to treatment [13]. Clinical manifestations range from clinically asymptomatic disease to overt heart failure and dysrhythmias. Immunofluorescence and electron microscopy are diagnostic [14,15].

Our patient presented with cardiomyopathy due to CIDD in the setting of newly diagnosed solitary thoracic plasmacytoma with no clonal plasma cells in a bone marrow sample.

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