Amyloid Tumor Behaving as Localized Amyloidosis in a Patient with Long History of Asymptomatic Light Chain Myeloma

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Amyloidosis includes a heterogeneous group of diseases characterized by the deposition of various proteins in fibrillar form. Historically, amyloidosis was classified as primary and secondary as well as systemic and localized. Amyloidosis derived from immunoglobulin light chain is the only type of systemic amyloidosis associated with a monoclonal protein [1]. Tumor-like presentation of amyloidosis (amyloidoma) is a rare condition and is usually associated with systemic deposits of amyloid. Patients with systemic forms of amyloidosis have a short expected survival.

The localized form of amyloidosis is a distinct entity with a much more favorable prognosis. The treatment consists usually of local surgical excision. By definition, localized amyloidosis implies that there are no findings of systemic disease; a positive test for urinary light chains will exclude this diagnosis. Localized deposition of amyloid may occur in a variety of organ systems, but evolution into systemic amyloidosis has not been reported [2].

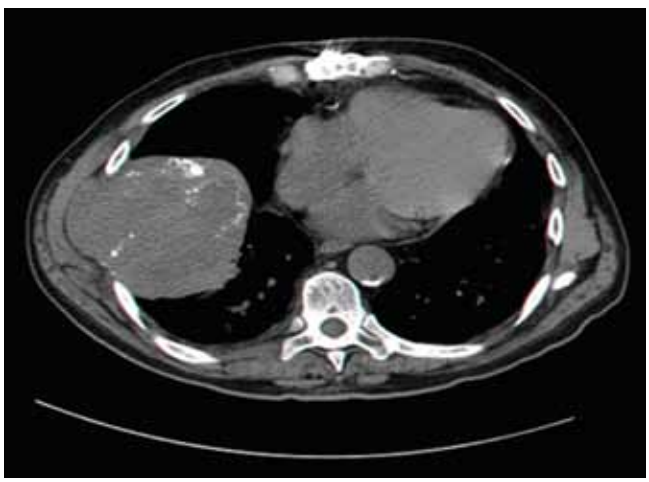
We describe a 73 year old man with a history of light chain multiple myeloma who presented with a huge solitary amyloidoma in his right chest 15 years after being diagnosed with multiple myeloma.

PATIENT DESCRIPTION

A 73 year old Caucasian man presented with a 2 week dry cough but no any other complaints. The physical examination was unremarkable, but a computed tomography scan of the chest demonstrated a 10 x 10 cm tumor in his right chest, originating most probably in the right seventh rib, compressing the lung, liver and chest wall [Figure]. Due to the calcifications within the tumor, chondrosarcoma was the suspected radiological diagnosis.

The patient's medical history was remarkable for lambda light chain multiple myeloma diagnosed 15 years previously. On diagnosis he had a pathological fracture due to right clavicle plasmacytoma, bone marrow involvement of about 30% monoclonal lambda plasma cells, and positive urinary lambda Bence-Jones protein. Otherwise, he had normal serum protein electrophoresis and immunofixation, normal electrolytes and blood count, and no other lytic lesions except for the right clavicle on skeletal survey. He was treated with local radiotherapy followed by 1 year of systemic therapy with monthly methylprednisolone and cyclophosphamide given intravenously and melphalan per os. During therapy, urinary Bence-Jones protein turned negative. During 15 years of follow-up he was completely asymptomatic and did not need further therapy.

He was under observation on a 6 month schedule. In the last 2 years, Bence-Jones urinary protein turned positive and gradually increased to a maximum 0.410 g/day. Serum free light chains kappa/lambda ratio was normal during the follow-up but turned abnormal at the time of tumor diagnosis, 0.21 (normal 0.26–1.65). The patient underwent computed tomography-guided biopsy from the chest tumor.



Chest CT showing a huge tumor in the left chest, with lung and chest wall compression

HISTOLOGICAL FINDINGS

Core needle biopsy of the mass demonstrated large deposits of amorphous eosinophilic material, which was Congo red-positive and exhibited green birefringence under polarized light, confirming the amyloid nature of the material. The amyloid deposits were surrounded by multinucleated foreign body-type giant cells. In the background there were infiltrates of plasmacytoid cells. These cells and the eosinophilic material were strongly positive for monoclonal lambda light chains, whereas a stain for kappa light chain was negative.

After the diagnosis of amyloidoma an extensive workup was conducted to seek systemic amyloidosis. Neurological examination was normal, without signs of peripheral neuropathy. The bone marrow biopsy showed normocellular marrow, with no increased number of plasma cells and no deposition of amyloid. A rectal biopsy was also negative for amyloid. A cardiac echocardiogram showed normal left ventricular function, with no signs of cardiac amyloidosis.

Skeletal survey and whole-body CT were negative for plasmacytomas. Laboratory tests showed the following: normal hemoglobin, calcium, creatinine, total protein/albumin, lactate dehydrogenase, serum protein electrophoresis, and as mentioned earlier, positive lambda light chains proteinuria of about 0.410 g/24 hr, with slightly abnormal free light chains kappa/lambda ratio. The patient underwent surgical extirpation of the tumor, along with excision of the right 7th rib; the recovery was uneventful. However, a follow-up chest CT scan 3 months after surgery showed a new 2.5 cm mass in the same area. A diagnostic biopsy showed the same pattern of amyloid tumor and the workup showed no other signs of amyloidosis. The patient is currently receiving local radiotherapy.

COMMENT

Localized and systemic amyloidosis are two distinct forms of the disease with different prognoses and management. The progression to disseminated disease is common in amyloid light chain amyloidoma [3,4]. Amyloidoma associated with plasmacytoma and without evidence of systemic amyloidosis is extremely rare [5].

We present a case of isolated amyloidoma that occurred in an otherwise asymptomatic patient with a long history of lambda light chain myeloma without need for treatment. Although by definition this tumor is part of a systemic disease, in our patient the clinical course was similar to the localized form. Due to the early relapse after surgical excision, systemic therapy as well as close follow-up may be warranted, after development of new signs and symptoms of systemic amyloidosis. To our knowledge, this is a unique case in which systemic amyloidosis behaved as a localized form.

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Capsule

Cytokine levels and histopathology in chronic HBV and HCV

The changes in balance of cytokine profile may result in either recovery or persistence of hepatitis B virus (HBV) and hepatitis C virus (HCV) infections. Akcam and co-scientists aimed to reveal a possible correlation between cytokine levels, i.e., tumor necrosis factor-alpha (TNFα), interferon-gamma (IFNγ), interleukin (IL)-10, IL-18, and transforming growth factor-beta (TGFβ); and Ishak score or fibrosis in patients with chronic hepatitis B (CHB) or chronic hepatitis C (CHC). Fifty patients with CHB (n=25), CHC (n=25), and the control group of subjects with negative hepatitis B and C serology (n=30) were included in the study. Patients who did not agree to participate in the study were excluded. Serum cytokine levels were measured by ELISA. Liver biopsies from the patients were also taken for pathological analyses by the same pathologist. The serum levels of TNFα, IL-10 and IL-18 in the hepatitis C group were significantly high compared with those of the control group ($P = 0.017$, $P = 0.001$, and $P = 0.004$ respectively), but only IL-10 levels in the hepatitis B group were significantly high ($P = 0.001$). These groups did not show any significant difference with respect to IFNγ or TGFβ levels. In patients with CHB or CHC, there was a significant correlation ($P = 0.000$) between TNFα and Ishak score or fibrosis, but no such correlation was found with IFNγ, IL-10, IL-18, or TGFβ. Results showed that cytokine activities were important indicators of clinical severity and progression of HBV and HCV infections. Further investigation on possible effects of cytokines on hepatocellular damage and fibrosis should be undertaken with new immunopathological approaches to viral hepatitis.

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