Plasmacytoma of Bronchus Treated by Radical Radiotherapy-A Rare Case with Four and a Half Years Follow up

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Abstract: Plasmacytoma of the bronchus is a very rare plasma cell neoplasm affecting the bronchus. Here we report a case of plasmacytoma of the bronchus treated by radical radiotherapy in July 2002. The tumor responded very well to treatment and showed a slow but sustained regression in the size over two years. Presently, he has completed four and a half years of follow-up and is free of disease.

Key Words: Extra medullary plasmacytoma, bronchus, radical radiotherapy.

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Extramedullary plasmacytoma (EMP) constitutes 3% of all plasma cell neoplasms.[1] It mainly affects the areas of the upper aerodigestive tract, such as the nasopharynx, tonsils, paranasal sinuses, nasal cavity, and salivary gland.[1] Primary pulmonary plasmacytoma is very rare. More rare are those arising from the bronchus. Here we present a rare case of plasmacytoma of the bronchus treated by radical radiotherapy.

CASE REPORT

A 62-year-old man presented to us in June 2002 with history of dry cough for the past three months. He also had three episodes of hemoptysis. He was a nonsmoker. Physical examination revealed decreased air entry on the left side of the chest. His complete blood counts, liver and renal functions were within normal limits. ESR was 30 millimeters in 1 hour. Sputum was negative for acid fast bacilli. Chest X-ray (Figure. 1) showed a large left hilar mass. Bronchoscopy showed an infiltrative growth involving the left main bronchus and left upper lobe bronchus. Contrast enhanced CT scan (Figure. 2 and Figure. 3) showed a large hyper dense mass of size 9 × 7 × 8 centimeters encasing the left main bronchus reaching the chest wall. There was no hilar or mediastinal lymphadenopathy. Bronchoscopic biopsy showed proliferation of mature plasma cells in an amyloid background, composed of immunoglobulin light chain deposits (Figure 4). On special staining with kappa and lambda antigens, lambda light chain positivity was seen (Figure 4 insert). Serum protein electrophoreses showed a narrow M-band in the beta-gamma interzone with normal polyclonal gamma globulins. M-band was 9.2% (0.05 g/dl). Immunofixation studies showed the M-band to be composed of lambda light chains only. Urine electrophoresis was normal. Bone marrow study showed a nomocellular picture. Quantitative immunoglobulin levels were also within the normal range (IgG-1230 mg/dl, IgA-255mg/dl, IgM-261mg/dl) Serum LDH was 368 U/L. Beta-2 micro globulin was 2739 micrograms/liter (normal range-700–3400). C-reactive protein was 0.6 milligrams/deciliter. There were no bony lesions on skeletal survey. We made a diagnosis of Extramedullary Plasmacytoma involving left bronchus.

Patient was treated by Radical external beam radiotherapy, 50 Gray in 25 fractions over 5 weeks, by anteroposterior/posteroanterior parallel opposed portals on telecobalt machine. The target volume included the tumor and regional nodes with margins. During radiotherapy he had only Grade 1 dyspnea which lasted for one month post radiotherapy and was managed symptomatically. He tolerated the whole course of the treatment very well.

The patient was followed up with serial CT scans and serum protein electrophoresis. CT scans showed a slow but sustained regression in the size of the tumor over two years. Partial response was documented in the CT scan done one and a half month post radiotherapy and complete response was achieved at 3 years. The CT scan at 3 years (Figure 5) showed no tumor. Serum M-spike became negative one and half months posttreatment and remains negative till now. Presently he has completed four and a half years of follow-up and is free of disease.

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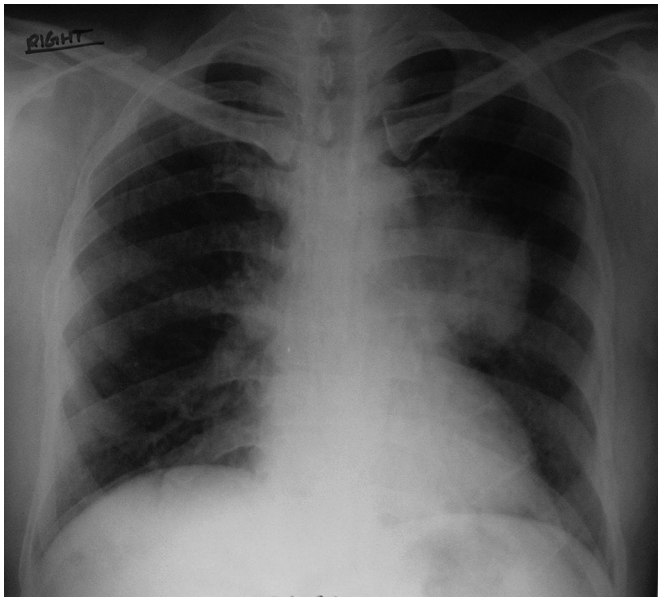

FIGURE 1. Chest X-ray of the patient before treatment.

FIGURE 2. Pretreatment CT scans of thorax (coronal view).

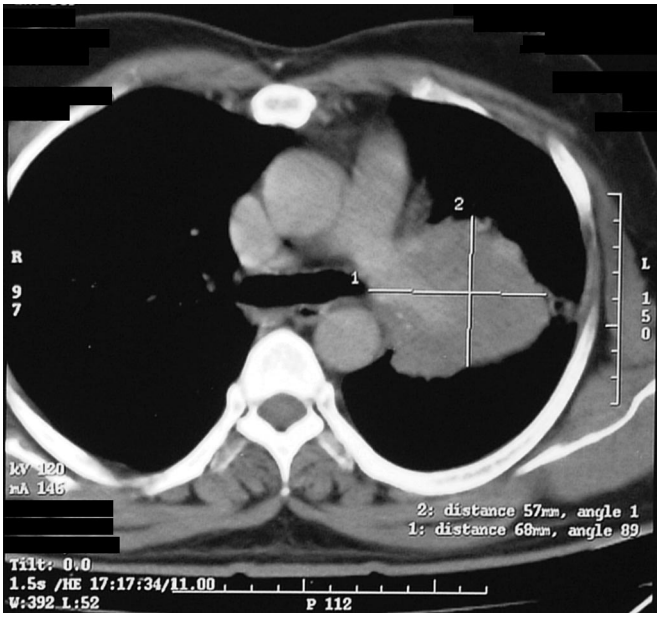

FIGURE 3. Pretreatment CT scan of thorax (axial section).

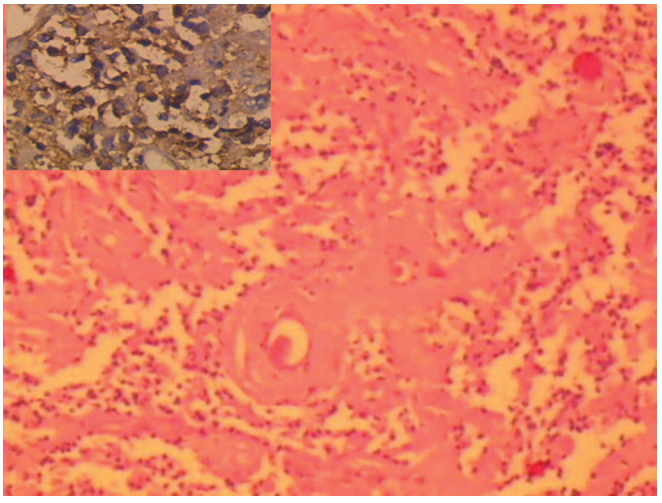

FIGURE 4. Histopathological picture H&E stain. (Inset: Lambda stain).

DISCUSSION

Primary pulmonary plasmacytoma is a very rare neoplasm (5% of EMP). Until now only 37 cases has been reported in the world literature.[2] More rare are those arising from the bronchus per se.[3,4] This is the fifth case of plasmacytoma of the bronchus to be reported.

Due to its rarity, there are no standard treatment guidelines for these cases. Surgical resection has been the mainstay of treatment in most of the reported cases. Some have tried local excision followed by radiotherapy. Our case is exceptional in that we have used radical radiotherapy treatment alone. EMP is known for the involvement of regional nodes. About 30–40% have first echelon nodes at diagnosis or at relapse. It is generally agreed upon that the regional nodes should also be addressed while treating EMP. Plasmacytomas are generally highly responsive to radiotherapy and chemotherapy. United Kingdom Myeloma forum recommends a radiotherapy dose of 40 Gray in 20 fractions for tumors less than 5 centimeters and up to 50 Gray in 25 fractions for tumors more than 5 centimeters.[5] In our case, the tumor was large, therefore requiring a dose of 50 Gray in 25 fractions over 5 weeks to the tumor with first echelon nodes.

In a review of literature on EMP, Etienne et al showed that complete or partial response following treatment is achieved in 65%.[2] In our case, we have seen a slow but sustained regression of the tumor. Chao et al treated 16 patients of EMP with radiotherapy and observed that local control was achieved in 100 % patients.[6] The predicted 10-year overall survival rate was 54%. Weber did an extensive literature review on EMP and saw that local control

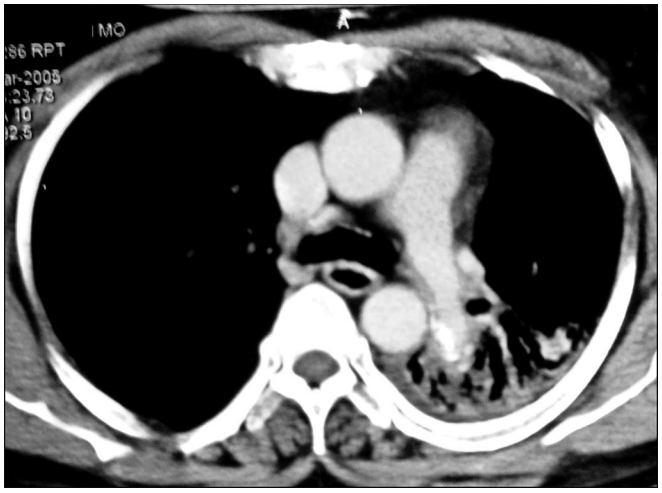

FIGURE 5. Posttreatment CT scan of thorax (axial section).

ranged from 80%–100% in different studies.[5] The 10-year disease free and overall survival ranged from 50–80%. The overall 2- and 5-year survivals of pulmonary plasmacytomas are 66% and 40%, respectively.[7]

Ozsahin et al. studied 258 patients of solitary plasmacytoma (bone = 206, extramedullary = 52).[8] Most (n = 214) of the patients received radiotherapy (RT) alone. The median follow-up was 56 months (range 7–245). The 5-year overall survival, disease-free survival, and local control rate was 74%, 50%, and 86%, respectively. Patients with EMP had the best outcomes, especially when treated with moderate-dose RT.

A significant proportion of cases of EMP do progress to multiple myeloma during follow up. This occurs after a median of 1.5– 2.5 years and their clinical course at progression is similar to those of patients diagnosed with de novo symptomatic myeloma. In the review of the literature by Etienne et al., 14% did develop multiple myeloma within 3 years following treatment.[2] In the Chao series treated by radiotherapy, the 5 year rate of conversion to multiple myeloma was 31%.[6] This mandates close follow up with serial serum protein electrophoresis even after complete remission of EMP.

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