Case Report

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Oncology Section

Primary Endobronchial Plasmacytoma with Mediastinal Lymph Nodes treated with Radical Radiotherapy: A Case Report with Review of Literature

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ABSTRACT

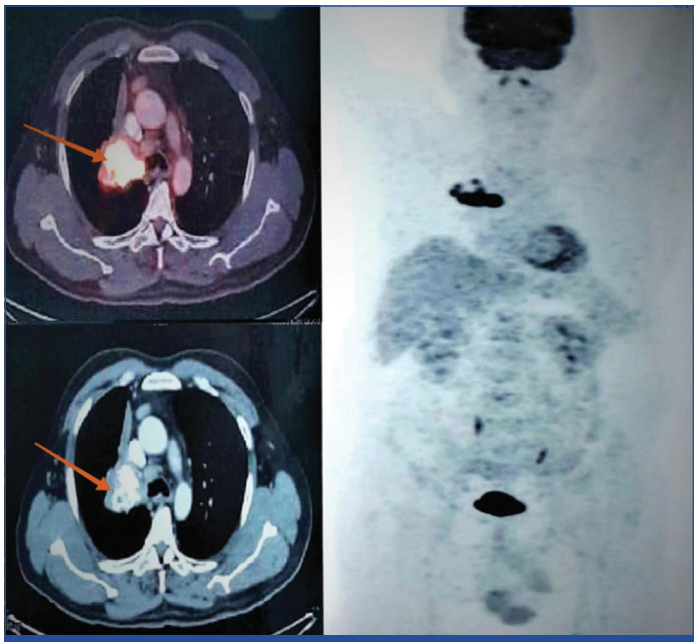
Malignant proliferation of plasma cells arising outside the bone marrow or osseous sites is known as extramedullary plasmacytoma. Solitary Extramedullary Plasmacytoma (SEP) are commonly located in upper respiratory tract and sinuses. Involvement of lower respiratory tract is rarely seen in case of SEP. Here, the authors report a case of endobronchial plasmacytoma in a 41-year-old male patient, without any medical risk factors presenting with complaint of blood stained sputum. Radiological imaging confirmed endobronchial mass in right main bronchus. Histopathology with immunohistochemistry confirmed the diagnosis of plasmacytoma. Work-up for multiple myeloma was negative. Thus, patient was subsequently planned for radical radiotherapy for the primary disease. The patient tolerated the treatment well and was asymptomatic at the time of last follow-up. Solitary Endobronchial Plasmacytoma (SBP) is a rare presentation of SEP. Radical radiotherapy yields excellent local control rates and symptomatic benefit. Close surveillance for development of multiple myeloma is required at periodic intervals.

Keywords: Malignant proliferation, Right bronchus, Upper respiratory tract

CASE REPORT

A 41-year-old male patient, without any medical risk factors presented with complaint of blood stained sputum since last two years. Computed Tomography (CT) of thorax revealed polypoidal endobronchial mass in right main bronchi and enlarged lymphnodes in pre-vascular, pre-tracheal, right tracheo-bronchial and right hilar region, largest at right para-tracheal measuring approximately 2.5×1.8 cm. Positron Emission Tomography-Computed Tomography (PET-CT) showed a large irregular metabolically active heterogeneously enhancing Fluorodeoxyglucose (FDG) avid (SUVmax: 28.47) soft tissue attenuation mass measuring approximately 6×5.6×3 cm (AP×TS×CC) in right hilar and supra-hilar location extending into right main bronchus, multiple FDG avid right para-tracheal, pre-vascular, pre-carinal and right internal mammary lymph nodes, largest measuring 2×1.4cm (SUVmax: 2.71) and no osteolytic lesions elsewhere.

[Table/Fig-1] Pre-treatment PETCT representing mass in right hilar and supra-hilar region (SUVmax: 28.47) left hand side showing axial section of thorax (upper one is coloured film and lower one is black and white film) and right hand side showing whole body PET-CT black and white film, all images showing mass in hilar and supra-hilar region.



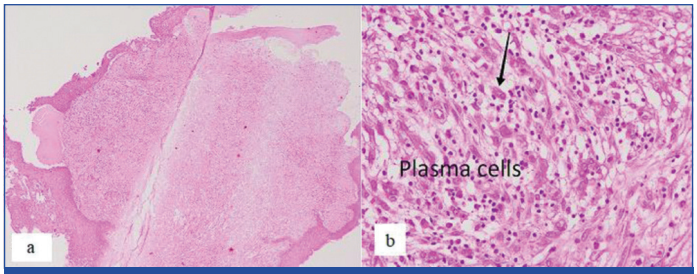
[Table/Fig-1]: Pre-treatment PETCT (whole body) representing mass in right Hilar and Supra-hilar region.

Haemoglobin (14.7 gm/dL), serum calcium (8.6 mg/dL), creatinine (0.7 mg/dL), beta 2 microglobulin (2328 ng/mL) and lactate dehydrogenase (150 U/L) were within normal limits. Serum protein electrophoresis with immunofixation (IgG -1360 mg/dL, IgM- 343 mg/dL, IgA- 117 mg/dL) did not reveal any M band and serum free light chain ratio was 1.1 (Kappa/Lamda). Bone marrow biopsy showed mild hypercellular bone marrow with 1% plasma cells and mild eosinophilia. Above findings points towards solitary plasmacytoma.

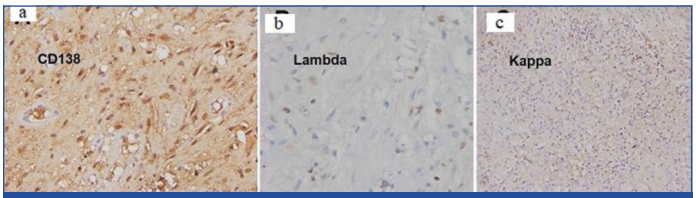
Bronchoscopy revealed right upper lobe mass lesion obstructing main bronchus lumen. Histopathology with immunohistochemistry showed plasma cells expressing Cluster of Differentiation (CD) 138, Epithelial Membrane Antigen (EMA), CD79a with lambda expression and limited kappa, cells are negative for CD38, Anaplastic Lymphoma Kinase (ALK), Synaptophysin, and chromogranin confirming the diagnosis of plasmacytoma. Pan Cytokeratin (CK), Leukocyte Common Antigen (LCA), Synaptophysin, chromogranin and kappa light chain were negative. Based on findings of bone marrow biopsy, biopsy of the specimen and IHC final diagnosis of SBP was made.

[Table/Fig-2a,b]: Histopathology findings depicted Photomicrographs of the polyps showing sheets of plasma cells (H&E Original magnification X40) and [Table/Fig-3] depicts Immunohistochemistry findings with photomicrographs showing CD 138 positive plasma cells [Table/Fig-3a], Lambda positive cells [Table/Fig-3b] and Kappa negative plasma cells [Table/Fig-3c].

Patient was treated with radical radiotherapy to the primary disease and involved lymph nodes to the dose of 45 Gray in



[Table/Fig-2]: Histopathology findings: Photomicrographs of the polyps showing sheets of plasma cells (H&E original magnifcation, X40).

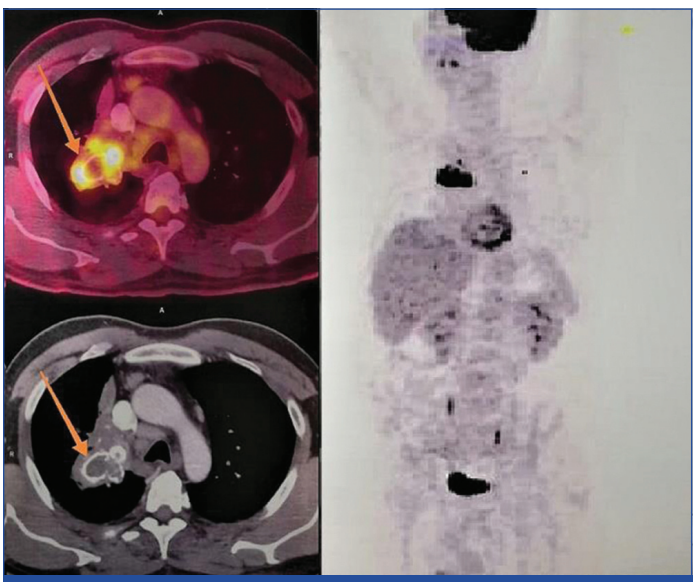


[Table/Fig-3]: Immunohistochemistry findings: photomicorgraphs showing CD138 positive plasma cells(3a). Lambda positive plasma cells(3b). Kappa negative plasma cells(3c) original magnification x100 (3a-3c).

25 fraction over five weeks at 1.8 Gray per fraction followed by boost to primary disease to a dose of 5.4 Gray in 3 fraction over three days with 3-dimensional conformal radiotherapy on a linear accelerator. Gross primary disease as evident on CT imaging was taken as Gross Tumour Volume Primary (GTV-P) and nodal disease as GTV-N. A margin of 1.5 cm was given to GTV-P to generate primary Clinical Target Volume (CTV-P) and 1 cm margin was given to GTV-N to generate CTV-N. A margin of 1 cm each was given to CTV-P and CTV-N to generate the respective Planning Target Volume (PTV). Haemoptysis subsided after 10 fractions of radiotherapy.

Patient is on regular follow-up every three monthly and monitored for multiple myeloma at every six months interval. PET-CT done at five months showed a large irregular metabolically active heterogeneously enhancing Fluorodeoxyglucose (FDG) avid (SUVmax: 11.7 vs 28.47) soft tissue attenuation mass measuring approximately 5.1X4.4×6 cm in hilar region showing favourable response to therapy.

[Table/Fig-4]: Post-treatment PET-CT representing mass in right hilar and supra-hilar region (decrease in SUV uptake from SUVmax: 28.47 to SUVmax: 11.7), left hand side showing axial section of thorax (upper one is coloured film and lower one is black and white film) and right hand side showing whole body PETCT black and white film, all images showing mass.



[Table/Fig-4]: Post treatment PETCT (whole body) representing mass in right Hilar and Supra-hilar region (decrease in size compared with previous PETCT).

At the time of last follow-up on 36 months, patient was asymptomatic with no respiratory complaints.

DISCUSSION

Malignant proliferation of plasma cells arising outside the bone marrow is known as extramedullary plasmacytoma. SEP are solitary non-osseous lesion, comprising approximately 3-5% of all plasma cells neoplasms and in approximately 80% of cases, it is located in the mucosa of nasopharynx and upper respiratory tract arising from sub-mucosal lymphoid tissues [1,2]. For diagnosis of extramedullary plasmacytoma, following points should be considered: the presence of a biopsy proven plasma cell tumour, bone marrow specimen showing fewer than five percent plasma cells, systemic signs and symptoms associated with multiple myeloma such as anaemia, hypercalcaemia, and bone pain should be absent [2]. Here, the authors report a rare case of SBP in a middle aged man presenting with haemoptysis.

SEP which involves the lung are uncommon form of plasma cell neoplasms that present outside of the bone marrow. Upper respiratory tract and nasopharynx is involved in approximately 85% of SEP, and they commonly presented with epistaxis, rhinorrhoea or a nasal obstruction [1]. SEP is more common in males than females with median age of presentation is 55 years, almost 10 years earlier in comparison to multiple myeloma [1]. In aero-digestive system, most common sites include pharynx, nasal cavity, oral cavity and paranasal sinuses [1]. Less than 15% of overall SEP involve non head and neck region and are associated with poor survival [3].

In a similar case report on SBP, diagnosis was made based on blood test which was normal. Imaging revealed mass obstructing left bronchus, biopsy revealed atypical tumour cells with an abundant basophilic granular cytoplasm and eccentrically located round nuclei, CD38 positive, serum and urine protein electrophoresis were normal, skeletal bone examination was also normal, bone marrow biopsy revealed a normocellular pattern with no increase in plasma cells [4]. Differential diagnosis included carcinoid, adenoid cystic carcinoma and, less likely, primary lung cancer.

Non head and neck SEP is treated by surgical resection of tumour and head and neck SEP is treated by combination of surgery and radiotherapy or radiotherapy alone [1,3]. Conservative surgical resection has been done for benign endobronchial tumours with occasional requirement of thoracotomy. For the treatment of small tracheo-bronchial tumours, bronchoscopic intervention has been found to be safe and effective. Since, SBP is very rare, no standard guidelines are available. [Table/Fig-5] represents 10 cases of SBP that has been in literature [1,2,4-11].

Two cases were treated by bronchoscopic removal using Nd-YAG laser ablation [6,7], four cases underwent surgical resection [5,8-10], three were treated by radical radiotherapy [1,4,11] and in one case both surgery followed by radiotherapy was done [2]. In the present case, patient was treated by radical radiotherapy. As these tumours are radiosensitive, for non-resectable tumour, treatment of choice is radiotherapy with a curative intent to a dose of 40-50 Gy over a period of four weeks as stated by United Kingdom Myeloma forum. If tumour is less than 5 cm then radiotherapy should be given to dose of 40 Gy for 20 fractions and if more than 5 cm, then 50 Gy in 25 fractions should be given [12].

Variable response rates have been reported in literature. Etienne G et al., reported complete or partial responses in 64%, while 14% cases developed multiple myeloma within three years [13]. Chao MW et al., treated 16 patients of Embden-Meyerhof-Parnas pathway (EMP) with radiotherapy and observed local control in all patients. Multiple myeloma developed in five patients within five years of plasmacytoma diagnosis with 10 year overall survival (OS)

S. no.	Author, year of publication	Age/sex	Presenting features	Treatment	Outcomes
1.	Brackett LE et al., 1994 [6], providence, RI	68/M	Cough, dyspnoea	Nd-YAG laser	Not reported for follow-up
2.	Terzi A et al., 1996 [8], Italy	65/M	Cough, dyspnoea	Subtotal resection of tracheo-bronchial carina	Remained on follow-up for 63 months, outcome not mentioned
3.	Piard F et al., 1998 [9] France	70/M	Dyspnoea	Left lower lobectomy	Not reported for follow-up
4.	Edelstein E et al.,2004 [7] Israel	47/M	Cough and shortness of breath	Bronchoscopic debulking and YAG laser	Remain on follow-up for 8 months with no recurrence.
5.	Haresh KP et al., 2007 [11], Delhi	62/M	Dry cough	Radical radiotherapy	On follow-up for 4.5 year remains disease free.
6.	Wei S et al., 2012 [10], china	42/M	None	Radical left lower lobectomy and lymphadenectomy	On follow-up for 15 months showed no local recurrence.
7.	Woo Park C et al., 2013 [2], Republic of Korea.	47/F	Blood tinged sputum	Surgical resection+Radiation	On follow-up, outcome not mentioned
8.	Agrawal SR et al., 2015 [1], Maharashtra	50/M	Cough	Denied Surgery Radiation dose 40 Gy over 4 weeks	Non progressive disease at 4 months
9.	LeNoir B et al., 2019 [5] South Carolina	54/F	Shortness of breath, wheezing	Surgical resection	No disease recurrence 1 year after treatment
10.	Park JI et al., 2021 [4] South Korea	86/M	Productive cough, dyspnoea	Radiation therapy	No disease recurrence 6 months after treatment
11.	Present case, 2023, Lucknow	41/M	Blood stained sputum	Radical radiotherapy	On follow-up for 36 months with favourable response to treatment.

[Table/Fig-5]: Table showing presenting features, treatment and outcome in reported cases of SEP [1,2,4-11].
Nd-Yag: Neodymium-doped Yttrium aluminium garnet; Gy: Gray

of 54% [14]. Weber D reported 10 year OS ranged as 50-80% [15]. Patient having extramedullary SP results in best outcomes when treated with moderate-dose RT.

CONCLUSION(S)

The SBP is a rare presentation of SEP. Radical radiotherapy yields excellent local control rates and symptomatic benefit. Close surveillance with long term follow-up (atleast 5 years) is recommended as they can develop into multiple myeloma and this conversion can be insidious. Although SBP is rare, SEP should be considered in the differential diagnosis of an endobronchial tumour because non-surgical treatment options exist. At 3 years of follow-up, the present case in the report remained without any recurrence.

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