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Case Study

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EXTRAMEDULLARY PLASMACYTOMA OF THE TRACHEA: A RARE CASE REPORT

Dr. Joy Augustine[1], Dr. Sumi Thomas*[2], and Dr. Divya S.[3]

[1]Professor and HOD, Department of Pathology, Amala Institute of Medical Sciences, Thrissur.
[2]MD Resident, Pathology, Amala Institute of Medical Sciences, Thrissur.
[3]Associate Professor, Department of Pathology, Amala Institute of Medical Sciences, Thrissur.

***Corresponding Author: Dr. Sumi Thomas**

MD Resident, Pathology, Amala Institute of Medical Sciences, Thrissur.

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ABSTRACT

Trachea is an extremely rare site of Extramedullary plasmacytoma(EMP). We report an incidentally detected solitary tracheal plasmacytoma in a 43 -year old man. Bronchoscopy revealed lobulated lesions around the carina obstructing the lumen. Pathologic analysis revealed EMP. Additional investigations excluded multiple myeloma.

KEYWORDS: Extramedullary plasmacytoma, trachea, multiple myeloma.

INTRODUCTION

Extraosseous (extramedullary) plasmacytomas(EMP) are localized plasma cell neoplasms that arise in tissues other than bone. They constitute 1-3 % of all plasma cell neoplasms.[1] Approximately 80% of extraosseous plasmacytomas occur in the upper respiratory tract, including the oropharynx and nasopharynx but they may occur in numerous other sites as gastrointestinal tract, bladder, breast, thyroid, parotid and skin.[2] EMP of the trachea is an extremely rare entity.[3] In this case report, we are presenting an incidentally diagnosed case of EMP of the trachea.

CASE DETAILS

A 43-year old gentleman presented at pulmonology OPD of our hospital with cough and dyspnoea. Imaging of the chest showed a small polypoidal soft tissue density along the left lateral wall of the trachea (14 x 13 mm) at the arch of aorta level(Fig.1). Bronchoscopy revealed 1.5 cm lobulated single vascular lesion seen on left lower lateral tracheal wall, 1.5 cm above the carina. Another identical lesion was seen just below secondary carina(Fig.2). Bronchoscopic biopsy done showed multiple fragments of tissue measuring 0.4 cm in greatest dimension. Microscopy revealed sheets of cells below the epithelium. The cells showed plasmacytoid morphology(Figs 3,4). With the differential diagnosis of Plasmacytoma and neuroendocrine neoplasm, we proceeded with the immunohistochemistry. Cells were positive for CD 138 (Fig.5). Since reactive collection of plasma cells are common in this site, Kappa and Lambda markers were done to determine the clonality. The neoplastic cells showed lambda light chain restriction (Fig.6,7). Meanwhile clinical, biochemical and radiological investigations were done to rule out multiple myeloma. Serum electrophoresis was done and no M band was detected. Urine was negative for Bence Jones Protein. Bone marrow aspirate and biopsy were within normal limits. Radiologically no other lytic lesionswere identified. This excluded the possibility of multiple myeloma with extraosseous deposits. Finally, a report of extramedullary plasmacytoma of trachea was given. The patient was started on radiation therapy (45 Gy in 25 fractions). Since the patient was non-responsive to conventional radiotherapy and the symptoms worsened, he was sent to interventional pulmonologist. The tracheal mass was removed. Microscopy revealed the same picture (Fig.8). Bone marrow aspirate and biopsy repeated. The results were negative.

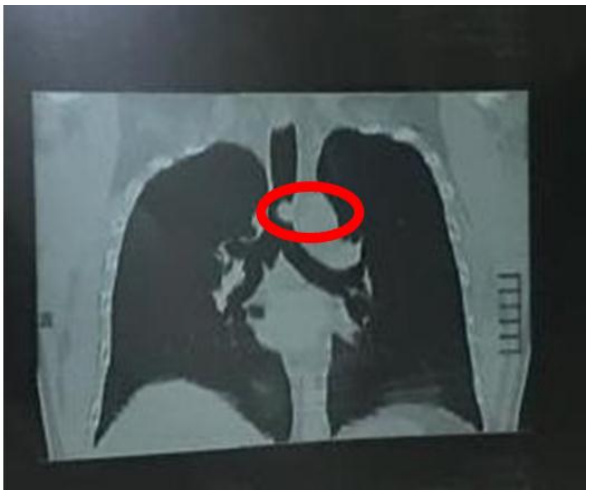

Fig. 1: Computerized tomography showing mass lesion in trachea.

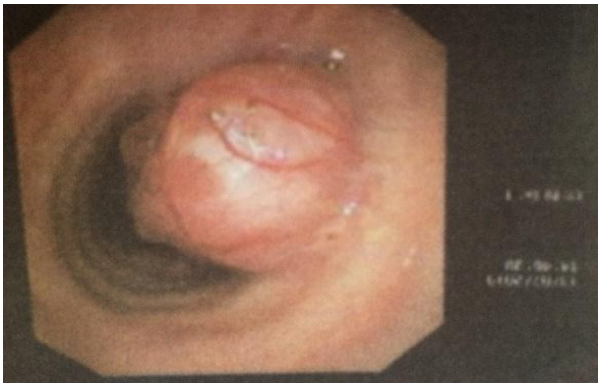

Fig. 2: Bronchoscopy showing pedunculated intraluminal mass narrowing the trachea.

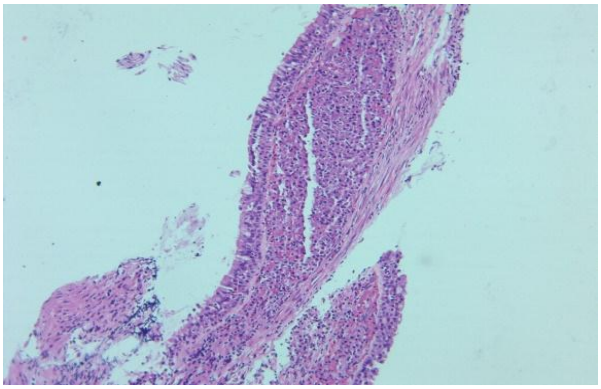

Fig. 3: Microscopy showing neoplasm beneath intact epithelium (H&E x40).

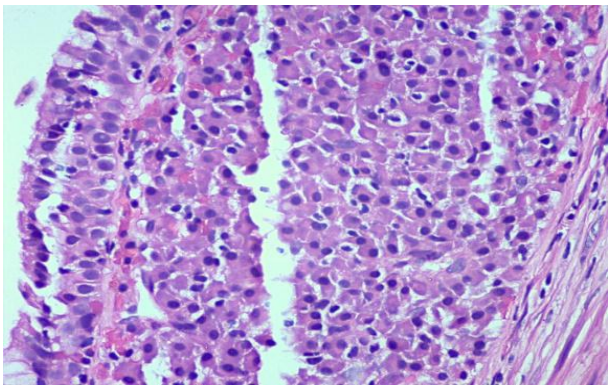

Fig. 4: Higher power showing sheets of plasma cells(H&Ex400).

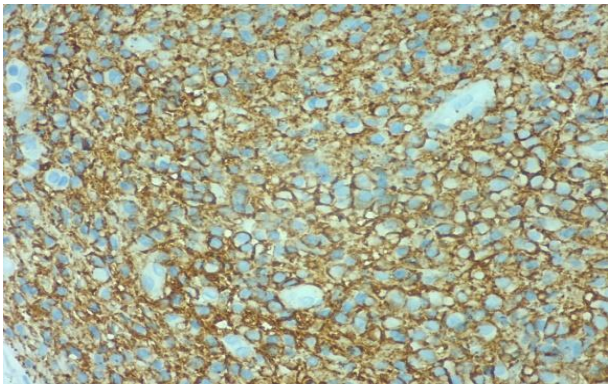

Fig. 5: Tumour cells with intense CD138 staining (H&Ex400).

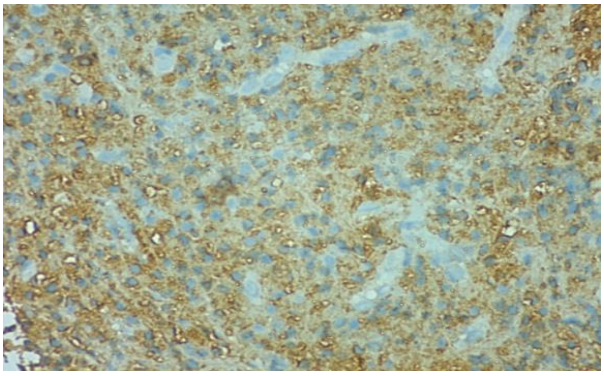

Fig. 6: Tumour cells with lambda light chain restriction (H&E x400).

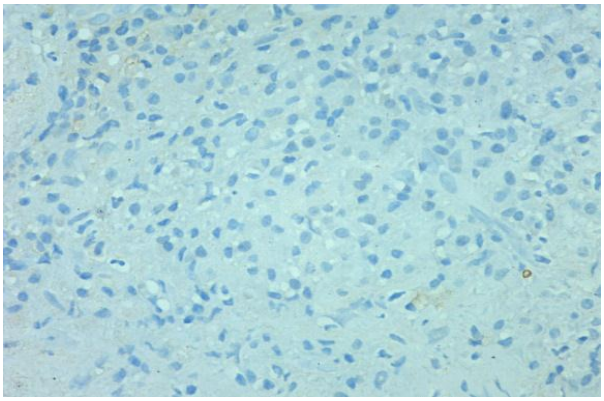

Fig. 7: Tumour cells staining negative for kappa (H&E x400).

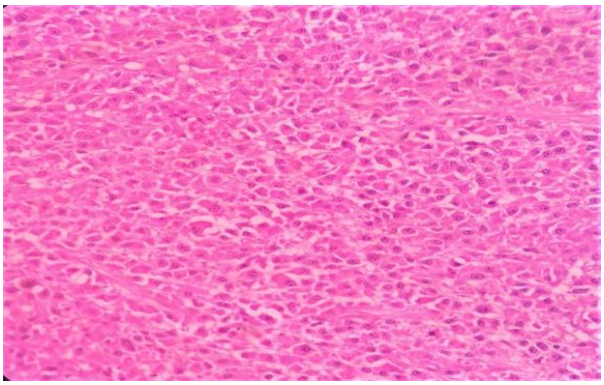

Fig 8: Tumour resection specimen (H & E x400).

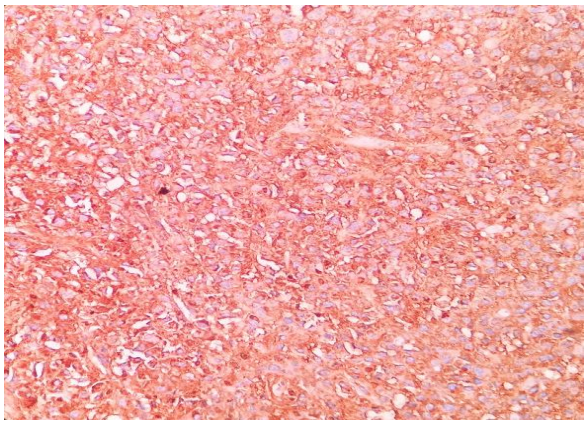

Fig 9: Tumour resection specimen with Lambda restriction (x400).

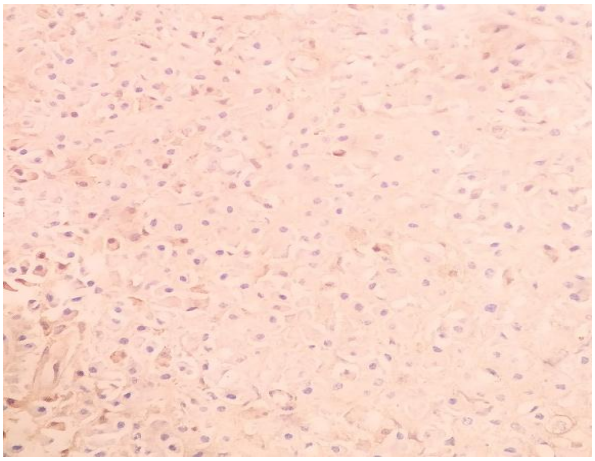

Fig 10: Tumour resection specimen negative for Kappa (x400).

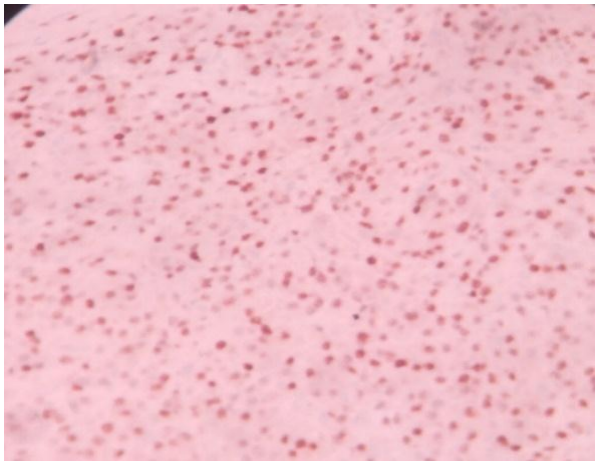

Fig 11: Tumour resection specimen positive for MUM 1 (x400).

DISCUSSION

The plasma cell neoplasms result from the expansion of a clone of immunoglobulin (Ig)-secreting, heavy chain class-switched, terminally differentiated B cells that typically secrete a single homogeneous (monoclonal) immunoglobulin called a paraprotein or M- protein. Plasma cell tumours consist of multiple myeloma (MM), extramedullary plasmacytoma (EMP), and solitary bone plasmacytoma (SBP).[4] MM can present as multiple bony lesions, plasma cell infiltration of the bone marrow and abnormal proliferation of monoclonal immunoglobulin in the blood. It can damage multiple organ systems and lead to organ failure.[5] Solitary lesions, occurring in the bone, are called plasmacytoma of bone, which most frequently occurs in the axial skeleton.[6] When solitary lesions occur in the soft tissues, it is called EMP.

Majority of the extraosseous plasmacytomas occur in the upper respiratory tract including the oropharynx, nasopharynx, sinuses and larynx. Symptoms are generally related to the tumour mass and include rhinorrhoea, epistaxis and nasal obstruction. Extramedullary plasmacytoma of the trachea is a rare entity with a very few cases reported in literature.[3,6,7] Two thirds of patients are male; median age at diagnosis is about 55 years. In our case, patient was a male aged 43 years. It represents around 3% of malignant tracheal tumours.[8,9] The pathogenesis of tracheal EMP is still unclear, but viral pathogenesis and chronic irritation may contribute to those lesions.[10] Airway obstruction is the most common symptoms of tracheal tumours. Clinically, in the early stages of development, patients may present with nonspecific symptoms such as chronic cough, dysphonia, hoarseness, hemoptysis, stridor, wheezing and dyspnoea, but dyspnoea becoming evident when the narrowing of the airway is over 75%.[7] Our patient had similar history.

Imaging plays a key role in depicting these tumours and assessing tumour extent within the lumen and airway wall. Chest radiography has many limitations and is often considered unremarkable in patients with tumours of the central airways.[11] CT scan is an essential diagnostic approach, which allows the lumen, airway wall, and mediastinal structures to be evaluated. Multiplanar reconstructions can be applied to evaluate the type, degree, and longitudinal extent of the airway narrowing as well as the location of the tumour and its distance from the cricoid cartilage to the carina.[11] Bronchoscopy can show an endoluminal view of the tumour. However, in extraosseous sites, distinction between lymphomas that exhibit extreme plasma cell differentiation and plasmacytoma may be difficult.[7] The final diagnosis is made through histopathological and immunohistochemical features. In our case tumour cells showed CD138 positivity with Lambda monoclonal light chain restriction. The diagnosis of EMP of the soft tissue has been based on the following criteria: pathological tissue evidence of monoclonal plasma cells involving a single extramedullary site; no bone marrow involvement; negative skeletal survey results; no anaemia, hypercalcemia, or renal impairment caused by plasma cell dyscrasia; and low serum or urinary levels of monoclonal immunoglobulin.[4]

In most cases the lesions are eradicated with local radiation therapy.[4,6,13] Regional recurrences develop in up to 25% of patients and occasionally there is metastasis to distant extraosseous sites.[4] Progression to plasma cell myeloma is infrequent, occurring in 15% of cases.[13,14]

CONCLUSION

The novelty of this case is that plasmacytoma of trachea is rare; in our case being an incidental finding. But it may cause significant respiratory distress. Clinical symptoms and imaging lack specificity. Diagnosis depends on histopathology and CD138 staining.

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