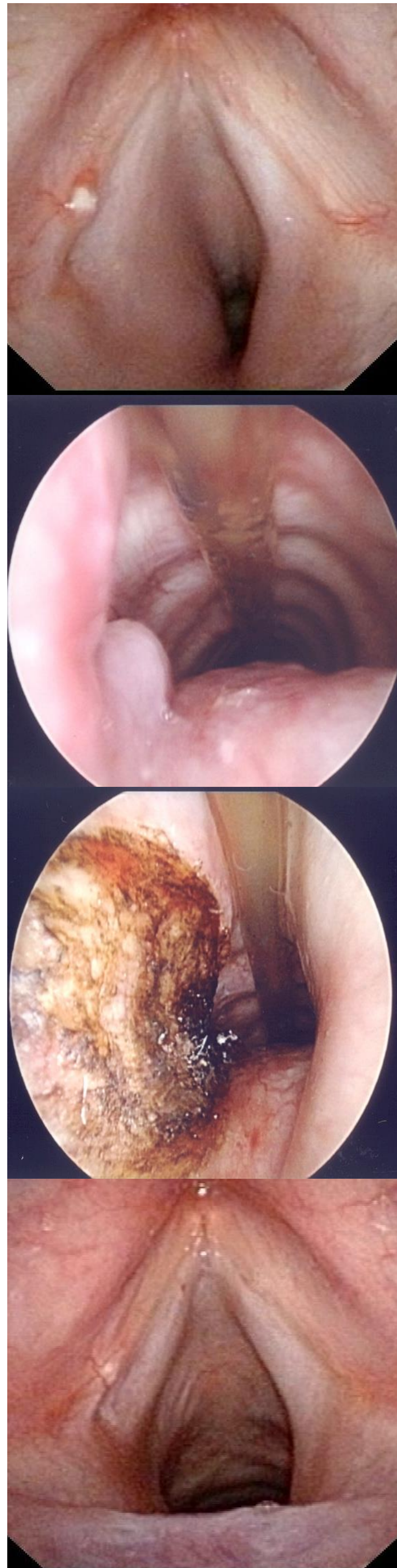


Subglottic extramedullary plasmacytoma with multiple myeloma masquerading as adult-onset asthma

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In sequence, initial consult, pre laser treatment, post laser treatment and four months post treatment

Introduction

Plasmacytoma is a frequent complication of multiple myeloma either at diagnosis or with disease progression. Extramedullary plasmacytoma (EMP) represents less than 1% of head and neck malignancies, however more than 90% of EMPs are diagnosed in the head and neck. (1) It is important to distinguish EMP from other plasma cell dyscrasias such as multiple myeloma for the purposes of prognosis and treatment.

Case Report

- A 55 year old gentleman with subglottic plasmacytoma with multiple myeloma is described.
- The patient initially presented with shortness of breath with expiratory wheeze, and was diagnosed and treated presumptively for asthma with response to steroid therapy.
- A subsequent relapse 6 months later prompted a specialist Otolaryngology consult.
- Preliminary findings showed a benign-looking nodular lesion at the subglottis. Biopsy of lesion was reported as a squamous cell carcinoma at an outside institution.
- Work up at our institution for newly diagnosed squamous cell carcinoma revealed an FDG avid left subglottic lesion with multiple bone metastases on a staging PET/CT scan.
- The patient underwent a panendoscopy and laser excision of the subglottic lesion with subglottic jet ventilation. Histology was reported as an extramedullary plasmacytoma.
- Further work up revealed presence of kappa light chain multiple myeloma with adverse cytogenetics.
- Patient was treated systemically with lenalidomide, bortezomib and dexamethasone for 4 cycles with rapid improvement in his symptoms.

Discussion

Solitary EMP of the larynx with associated multiple myeloma is rare but has been reported previously.

- Werner in 1991 reported a total of hundred and eleven cases of laryngeal plasmacytoma of which twenty one had multiple myeloma. (2).
- Wein in 2002 reviewed twelve reports of primary plasmacytoma of the subglottis. Six of the twelve presented with or later went on to develop MM. Progressive onset of shortness of breath and/or hoarseness were presenting symptoms in most cases. Majority of patients (58%) required tracheostomy for airway stabilization and underwent local radiation as primary therapy. Interestingly, much like the case presented, three of twelve cases noted a false negative in initial biopsy results. (3)
- Since 2002 there has been one further case report of EMP of the subglottis, which was treated with radiotherapy and one cycle of chemotherapy. (4)
- **We present the first case of subglottic plasmacytoma with multiple myeloma managed locally via laser excision and systemic therapy.**

Proper management of extramedullary plasmacytoma with MM remains controversial.

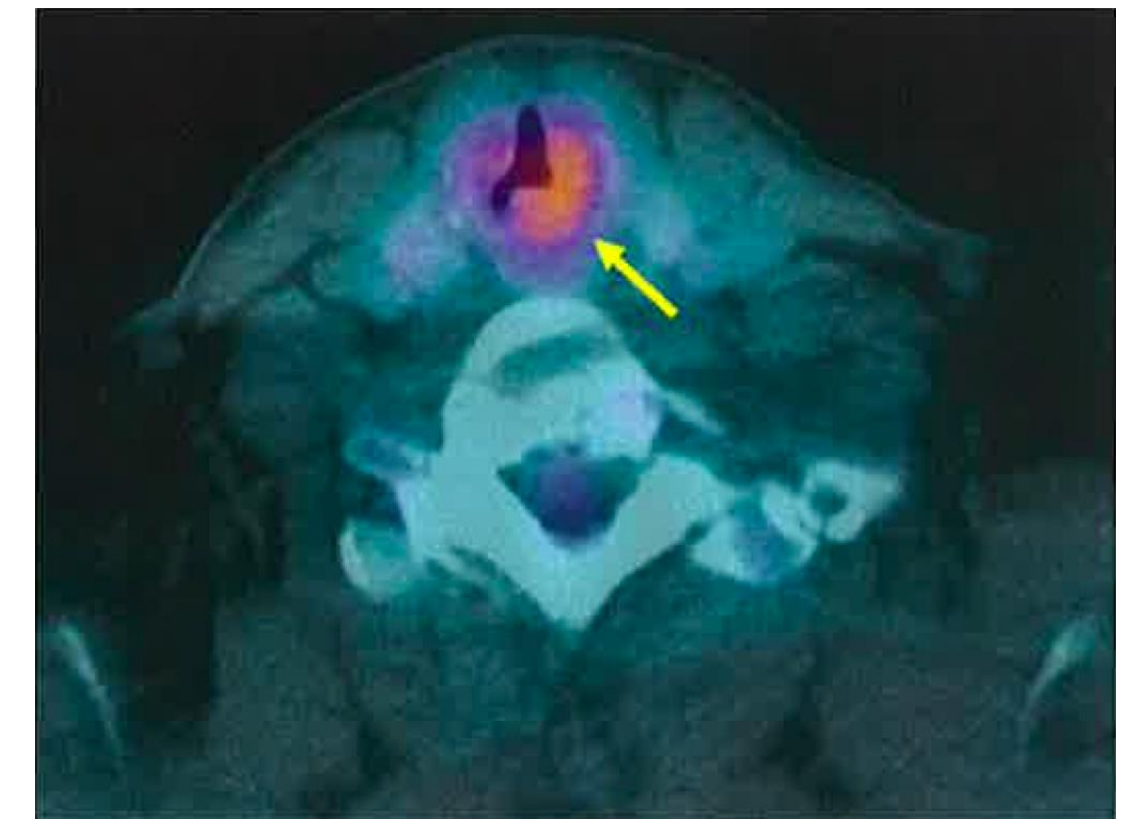
- The clinical course of EMP with multiple myeloma is often aggressive and associated with both early progression and short survival.
- Response to conventional chemotherapy, thalidomide or/and HDT is poor. (5) Focal radiation therapy often in combination with dexamethasone is the treatment of choice for local control.
- Newer agents such as lenalidomide and bortezomib are associated with high response rates in treatment of both multiple myeloma and plasmacytoma. This is particularly useful to avoid radiation induced toxicities when treating the head and neck area.
- However there is a lack of prospective well-powered studies to decide the best treatment of EMP with multiple myeloma at initial diagnosis.

We also highlight in this case report the unfortunate delay in diagnosis due to a presumptive diagnosis of asthma. Patients presenting with shortness of breath and especially soft stridor, warrant an immediate or early ENT referral.

References

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PET/CT scan



A mild to moderately FDG avid left subglottic lesion is seen