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Amyloid Got Your Tongue? Multiple Myeloma Presenting as Progressive Dysphagia

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Introduction

- Multiple myeloma is a well-known hematologic malignancy characterized by plasma cell proliferation, producing monoclonal immunoglobulin
- Typical symptoms of MM at presentation often include bone pain from lytic bone lesions, hypercalcemia, systemic signs of malignancy, or renal failure
- Amyloid deposition is a known sequela of MM, most often found in the kidney causing renal failure. Amyloid deposition is also known to occur in the heart, liver, tongue, esophagus, and other organs with organ-specific findings (i.e. heart failure, hepatomegaly)
- Amyloid deposition causing dysphagia is a poorly described sequela of MM, especially without other findings suggestive of MM such as kidney or bone disease, and amyloidosis
- Here we present a case of undiagnosed multiple myeloma presenting as progressive dysphagia in a patient without worsening kidney disease, bone pain, hypercalcemia, or other systemic signs of malignancy, with also the incidental finding of concurrent pancreatic cancer
- This case illustrates a diagnostic challenge that involved multiple subspecialties and unconventional imaging modalities. It also illustrates the psychosocial complexity of diagnosis and treating patients with malignancy.

Case Presentation

- 71 year old male with past medical history relevant for stage 3 chronic kidney disease attributed to hypertension and insulin-dependent diabetes
- Presenting with multi-month history of progressive dysphagia, starting with solids, now unable to tolerate any oral intake without significant difficulty
- Associated symptoms: ~50 pound weight loss, weakness, fatigue, lightheadedness, Patient denied: fever, chills, night sweats, pain at any location, enlargement of tongue or other oropharyngeal organs
- Outpatient swallow study: severe pharyngeal dysphagia resulting in aspiration of all consistencies and severe residue.
- Admitted directly from clinic due to clinical malnourishment, dehydration, and for further diagnostic further workup
- Admission studies: CBC unremarkable. BMP notable for creatinine 1.5, consistent with patient baseline
- Exam on arrival: Generally cachectic. Pooled secretions in mouth, no other visible oropharyngeal abnormalities. No lymphadenopathy. No pain on palpation of any location. Normal neurologic exam without focal deficits.

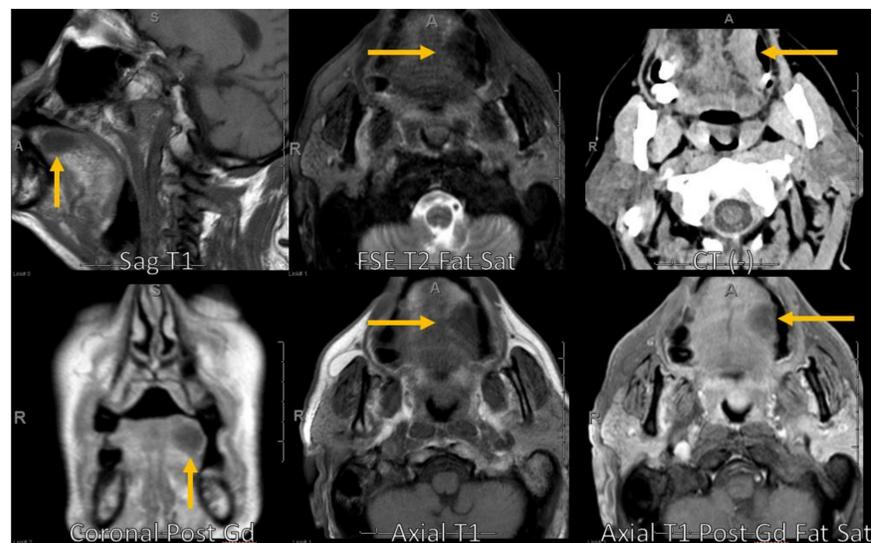


Figure 1. MRI Soft Tissue Neck indicating mass found to be amyloid. MRI read and graphic courtesy of Dr. John Corrigan, HFH Radiology

- Dysphagia workup:
 - Barium esophagram: presbyesophagus, no other physical or functional abnormalities
 - CT head and neck: irregular soft tissue edema of larynx without masses or lymphadenopathy. Incidental finding of possible pancreatic tumor
 - Direct visualization with flexible laryngoscopy: laryngeal edema, no other visible abnormalities.
 - Paraneoplastic panels and neuromuscular disorders workup: negative
 - MRI soft tissue neck: non-specific, hypointense mass near the tongue (fig. 1)
- Biopsy of the mass revealed amyloid deposition (fig. 2)
- Protein electrophoresis suggestive of multiple myeloma
- Diagnosis of Multiple Myeloma confirmed by Bone Marrow Biopsy
- Skeletal survey negative for bone lesions
- Patient received PEG tube and was started on chemotherapy for Multiple Myeloma
- Patient course complications:
 - Refeeding syndrome
 - Incidental discovery of Pancreatic Adenocarcinoma
 - New-onset Major Depression
 - Multiple aspiration events necessitating tracheostomy placement

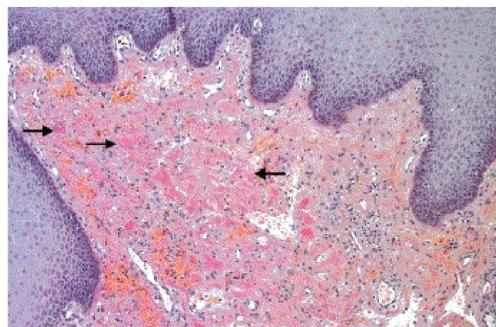
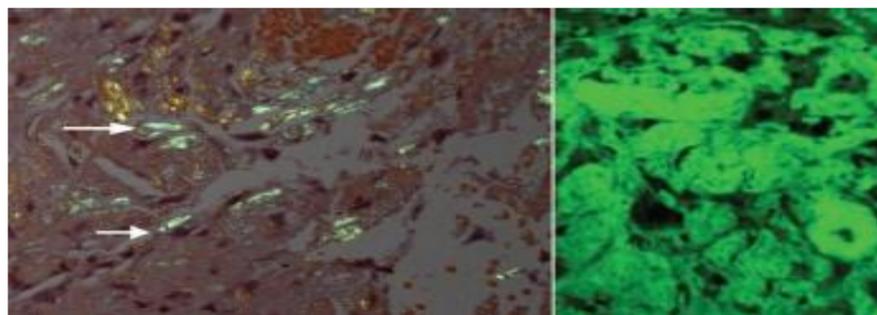


Figure 2. Histopathologic analysis of amyloid deposition in the tongue of a patient with multiple myeloma found to have a tongue tumor. Pink color shown with Congo Red staining above, apple-green birefringence through polarized light in the bottom left image, and Thioflavin-T staining showing immunofluorescence in the bottom right image (1)



Discussion

- Multiple Myeloma is a malignancy with a well-documented pattern of “classic” presentation symptoms, such as bone pain, kidney dysfunction and hypercalcemia
- Cases of MM-induced amyloid deposition causing dysphagia have been seen, but usually in the context of later-stage disease with systemic signs, bone lesions and lab abnormalities (2)
- Dysphagia as the primary presenting symptom leading to MM diagnosis is exceedingly rare, especially without any other typical MM findings on presentation
- Amyloid deposition, often in the setting of MM or in other amyloidosis settings, typically causes the following (3):
 - Kidney (most common) – may range from asymptomatic proteinuria to nephrotic syndrome or renal failure
 - Heart – many possible complications including systolic then diastolic dysfunction, arrhythmias, conduction abnormalities, coronary artery disease

- GI tract (esophagus, duodenum, colorectal) – may cause mucosal bleeding, obstruction or dysmotility, may be evaluated with random biopsies
- Also may cause macroglossia, bleeding disorders, or nervous system compromise
- Standard dysphagia workup was negative, leading to the consideration of atypical imaging modalities – MRI soft tissue neck is rarely used outside of known head/neck malignancy (4)
- Especially in the setting of new diagnosis of malignancy, special care should be paid to the specific deficits a patient is suffering from as these may be the most important issues for the patient, the biggest triggers for any mood disturbance or mental health difficulties, and most impactful barriers to compliance with care
- This patient’s most trying symptom, later identified as the primary trigger for major depression, was their inability to safely eat and enjoy food and drink, and they suffered multiple aspiration events and infections secondary to difficulty coping with this change.
- As many as 60% of cancer patients may develop depression related to their cancer diagnosis (5)
- The incidental finding of coinciding pancreatic cancer caused significant delay in workup for his dysphagia, as well as adding significant psychological stress to the patient and family
- Incidence of discovering multiple primary cancers in a single patient typically occurs when studies to evaluate one entity reveal the other, as in our patient, and appears to be increasing in day to day practice
- This association may be due to increased power of screening and surveillance techniques, and may cause significant complications when finalizing treatment plans (6)

Conclusion

- The common presenting symptoms of multiple myeloma, such as kidney dysfunction, hypercalcemia and bone pain are well known; however, MM may present in many manners without the typical findings, presenting a diagnostic challenge
- Amyloid deposition in various organs, including the tongue leading to enlargement or the esophagus, are known sequelae of multiple myeloma, progressive dysphagia is an exceedingly rare manner of presentation, especially without identifiable bone lesions, pain, worsening kidney function, or other findings typically for multiple myeloma.
- This case illustrates the importance of complete dysphagia workup including potential utilization of atypical imaging modalities and appropriate utilization of specialty service consultation
- The care of patients with debilitating malignancy necessitates significant attention paid to the psychological and social aspects of individual patients, including involvement of support systems and careful monitoring for mental health changes, for optimal outcomes
- Multiple unrelated cancers presenting in one patient is a rare but increasingly documented phenomenon that may possibly be attributed to improvements in screening techniques and imaging, and certainly contributes to the above-described patient complexity

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