

Case Description of Head and Neck Alveolar Rhabdomyosarcoma in an Adult Patient

McKenzie Hargis¹, Hannah Malinosky¹, James Watson², MD, Deanna Huffman³, DO

¹Louisiana State University Health Sciences Center School of Medicine,

²Department of Surgery, ³Department of Medicine, Section

Hematology/Oncology



Introduction

- Rhabdomyosarcoma (RMS) is a soft tissue tumor of myogenic lineage that is typically seen in the pediatric population and is a remarkably rare diagnosis in adults
- The three subtypes of RMS include embryonal, alveolar, and pleomorphic
- The alveolar subtype (ARMS) predominates in older age groups and carries a worse prognosis
- Presentation in the head and neck region is uncommon and often misdiagnosed as small cell carcinoma due to its rarity and histopathological characteristics
- Genetic detection of characteristic chromosomal translocations aid in making a definitive diagnosis

Methodology

- Electronic medical record review of notes, imaging and pathology results

Case Summary

- 44-year-old female presented for two weeks of rapidly progressive right sided fixed and tender cervical lymphadenopathy, weight loss and night sweats
- CT scan of her head and neck revealed enlarged right cervical and intraparotid lymph nodes
- Fine needle aspiration (FNA) of the masses were positive for malignant cells
- Excisional biopsy of cervical lymph node was consistent with alveolar rhabdomyosarcoma with positive myogenin and PAX3/FOXO1 fusion gene
- Maxillofacial CT revealed large soft tissue mass centered at the right ethmoid air cells and infiltrating the entire right paranasal sinuses
- Repeat biopsy of the primary mass by laryngoscopy confirmed the diagnosis and patient was staged as stage 1, T1aN1M0, clinical group 3, intermediate risk ARMS
- Four cycles of chemotherapy with vincristine, dactinomycin, and

Imaging

Initial

Imaging

Figure 1. CT Maxillofacial Scan from 10/5/2022. Large soft tissue mass centered about the right ethmoid air cells infiltrating the entire right paranasal sinuses with extension into the right cavernous sinus. Bilateral cervical lymphadenopathy, right greater than left.

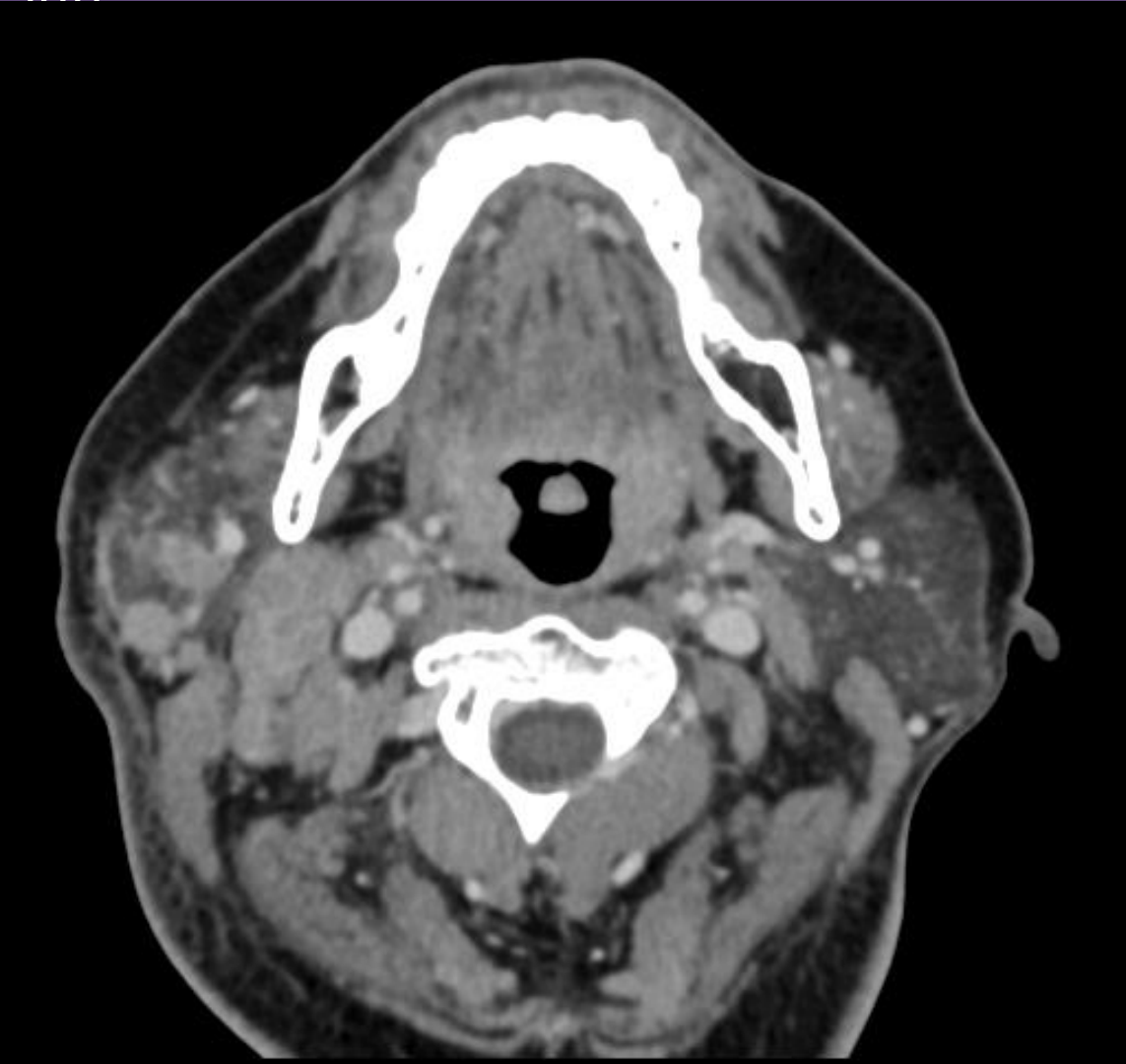


Figure 2. MRI Face and Neck with and without contrast 10/6/2022. Irregular enhancing mass centered about the right inferior concha with bulky head and neck lymphadenopathy most consistent with metastatic nasopharyngeal carcinoma.

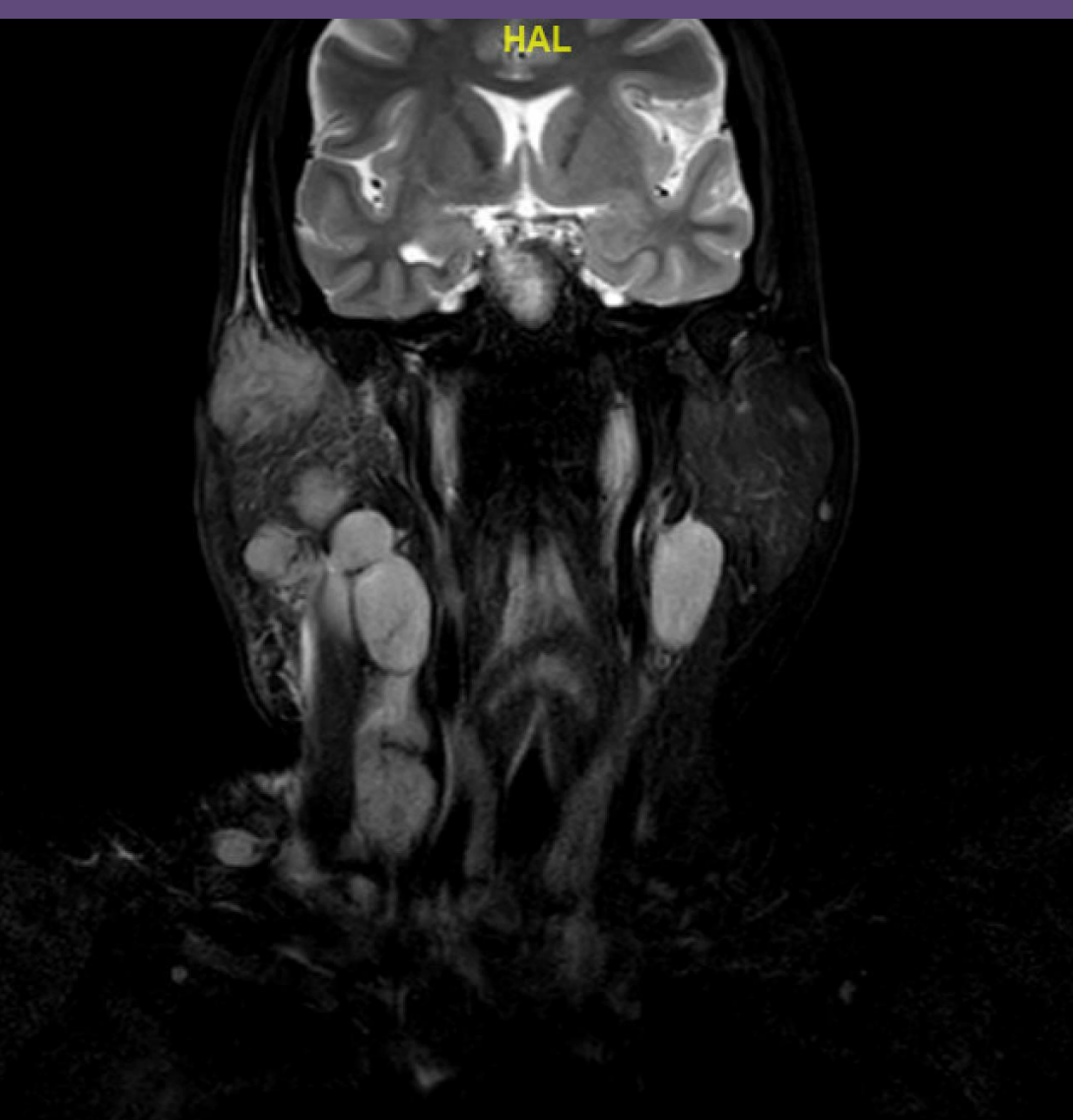
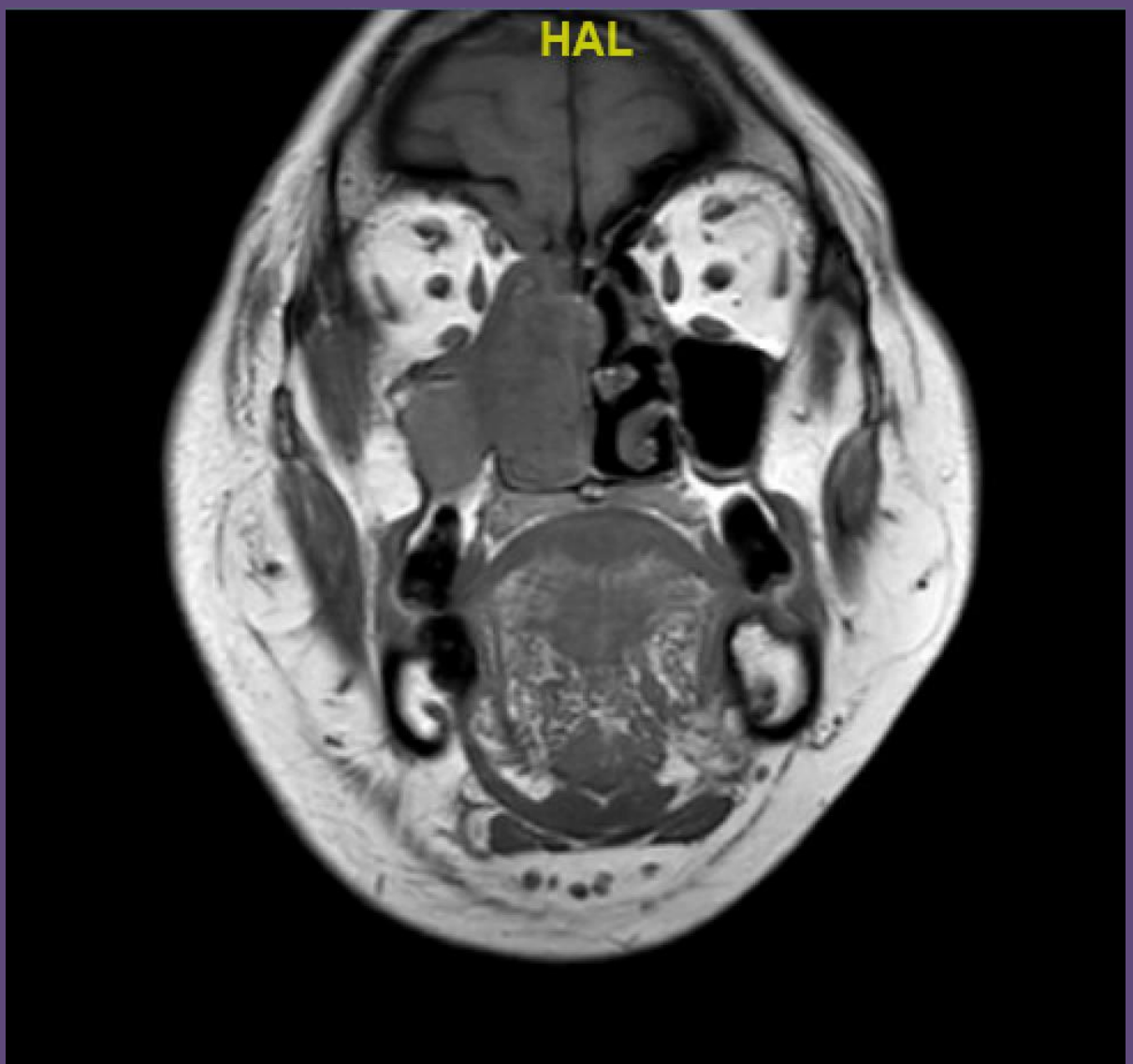
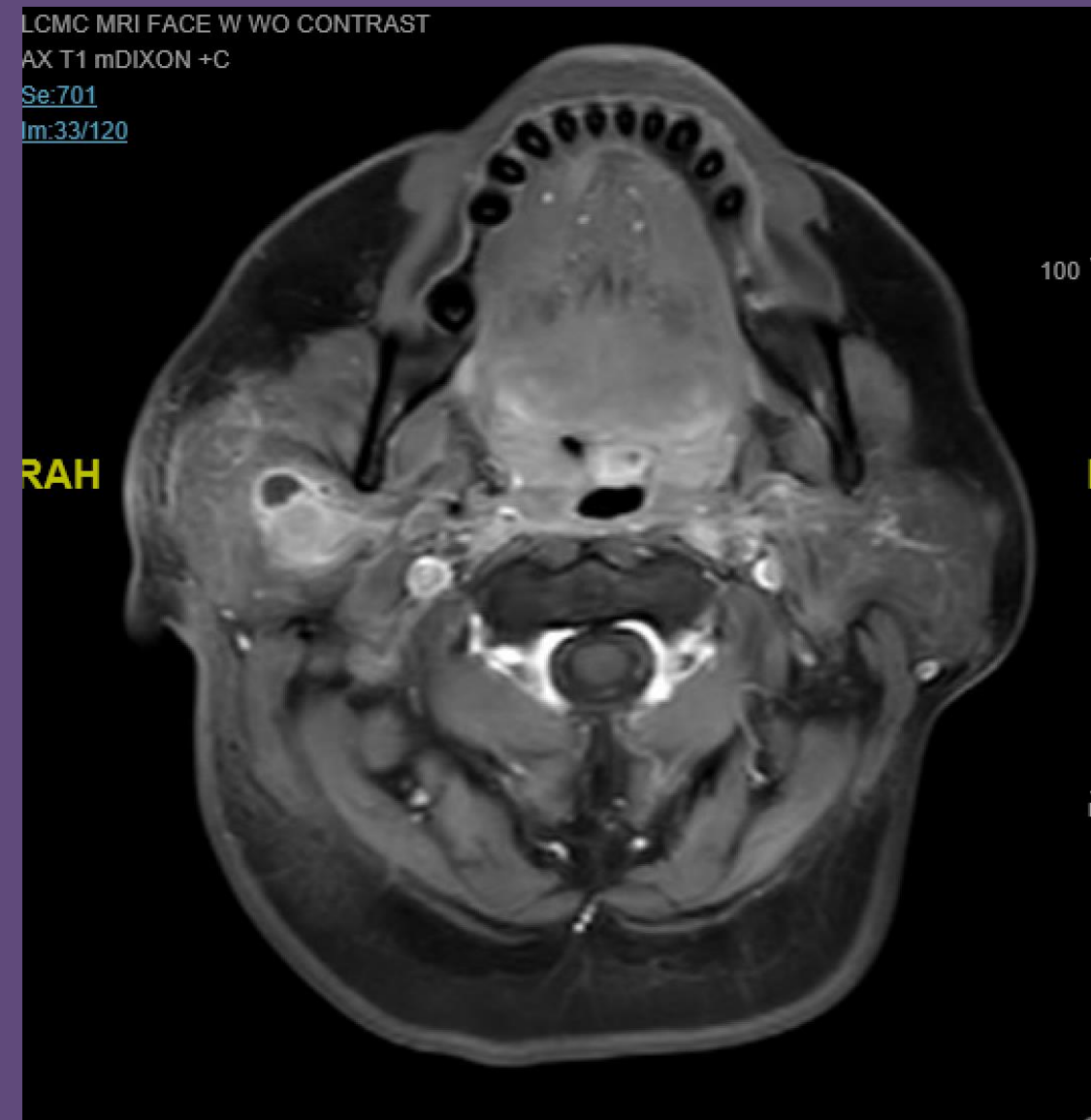
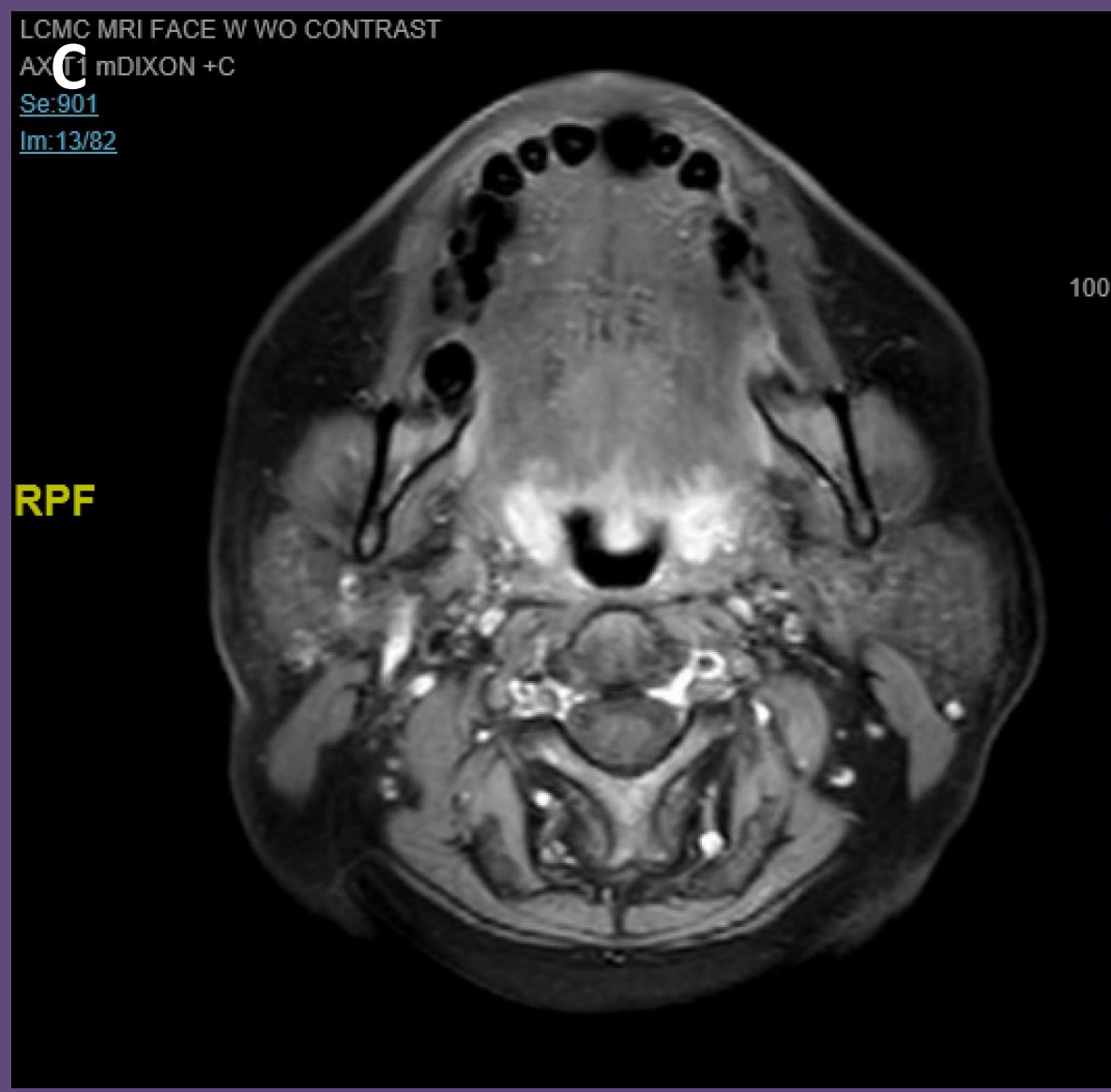
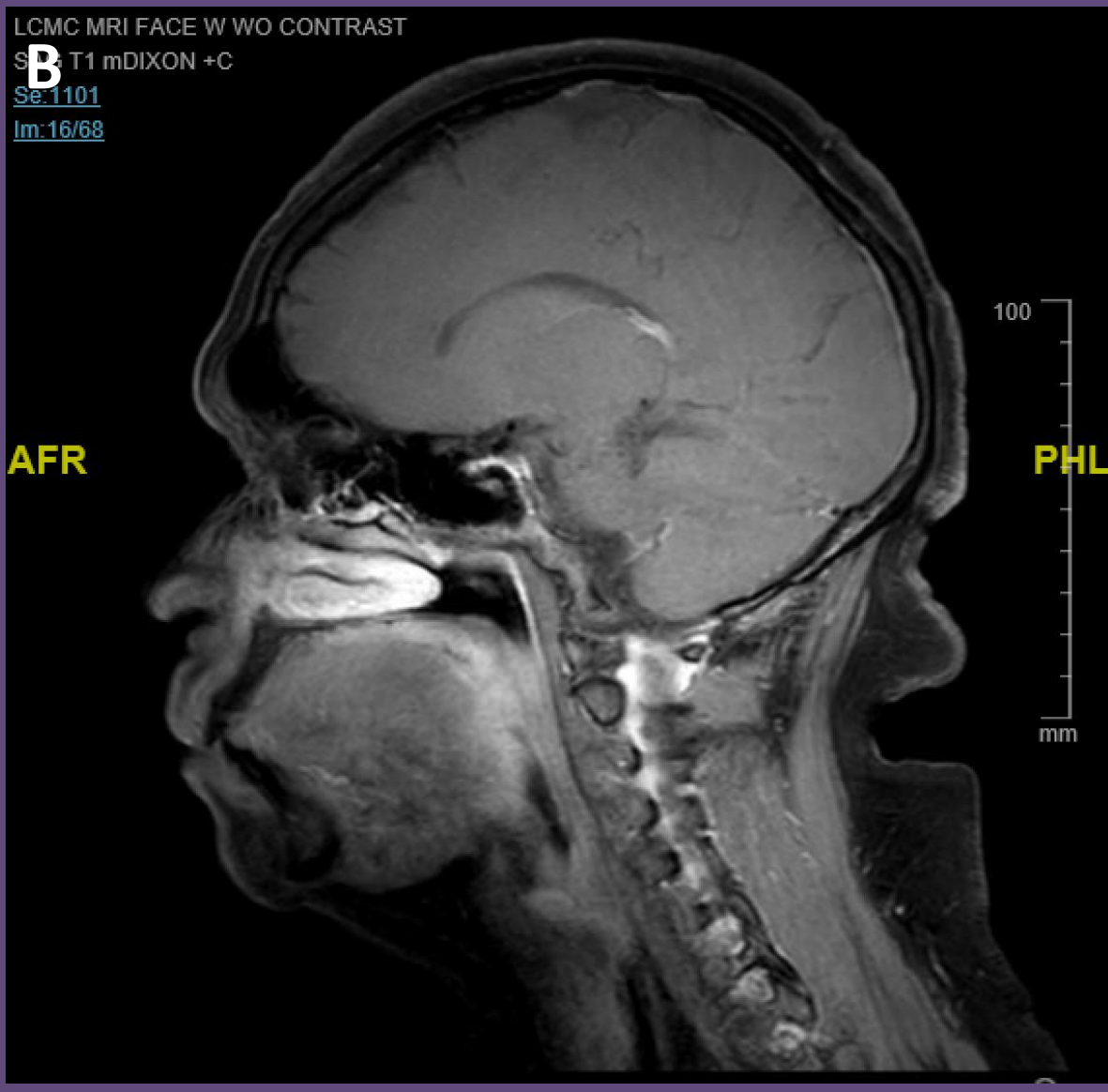


Figure 3. Coronal T2 Neck MRI. An intraparotid lymph node demonstrates central areas of nonenhancement consistent with necrosis.



Repeat Imaging

Figure 4. MRI Face with and without contrast 1/4/2023. A. Substantially decreased size of the mass in the right inferior concha B. Sagittal view of right inferior concha mass. Also, shows interval resolution of the lymphadenopathy of the neck C. Coronal T2 Neck MRI showing interval resolution of right parotid gland mass as compared to figure 3.



Results

- MRI results show substantially decreased size of the mass in the right inferior concha, resolved right parotid gland mass and resolved cervical lymphadenopathy
- Due to interval decrease in size of the right inferior concha mass, there is no plan for surgical intervention at this time
- Will proceed with chemoRT according to

Discussion and Conclusions

- Although ARMS remains a rare diagnosis in adults, it should be considered in adult patients presenting with head and neck masses as it is aggressive with high rates of metastasis
- Prompt diagnosis and treatment could result in better patient outcomes

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