Case description of head and neck alveolar rhabdomyosarcoma in an adult patient

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Background:

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 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.

Methods:

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

Results/Case Summary:

44-year-old female presented for two weeks of rapidly progressive right sided cervical lymphadenopathy, weight loss and night sweats. The lymphadenopathy was fixed and tender to palpation. 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 cyclophosphamide (VAC) were completed. Today MRI results show substantially decreased size of the mass in the right inferior concha, resolved right parotid gland mass and resolved cervical lymphadenopathy. Patient will likely undergo surgery soon with radiation therapy to follow.

Conclusion:

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.