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Case Report

Sinonasal Alveolar Rhabdomyosarcoma in an Adult Patient: A Case Report and Review of the Literature

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Abstract: We report on a patient who presented to the ENT service with right side nasal obstruction. Imaging studies revealed an aggressive non-calcified solid heterogeneous mass centered in the right naso ethmoidal region. The mass was hyper enhanced following contrast media administration. The patient underwent partial tumor resection and a biopsy was performed confirming the presence of Solid Alveolar Rhabdomyosarcoma. The patient was treated with chemo-radiation therapy.

Keywords: CT, MRI, Ethmoidal, Rhabdomyosarcoma, Radio-Chemo Therapy

1. Introduction

Rhabdomyosarcoma (RMS) is a malignant tumor of striated muscle origin. It is derived from primitive mesenchyme that retained its capacity for skeletal muscle differentiation. It is one of most common sarcomas in newborns and childhood. Approximately 35% of RMS occur in the head and neck region. The combined use of chemo-radiotherapy and surgery improves survival rate significantly for up to 5 years.

2. Clinical Case

We report on a 22 years old insulin resistant female patient who presented to the Ear Nose and Throat (ENT) clinic with right side nasal obstruction. No history of epistaxis was reported but proptosis of the right eye was observed and loss of eyeball adduction. In the ipsilateral gonial region, hypoesthesia was present for two months duration. Lymphadenopathy of 4 cm in size was observed in the II A ipsilateral cervical ganglionic level. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) were performed.

The CT showed an aggressive non-calcified heterogeneous solid mass with a maximum diameter of 4.5 x 4.5 x 6.2 cm, centered in the right nasal fossa and hyper enhanced following intravenous contrast media administration. The mass invades the right side of the frontal, maxillary,

ethmoidal and sphenoid sinuses, hard palate, right orbit and alveolar bone. There was lymphadenopathy of the ipsilateral IB, II and V ganglionic levels. Evidence of intracranial invasion was no demonstrated. (Fig. 1)

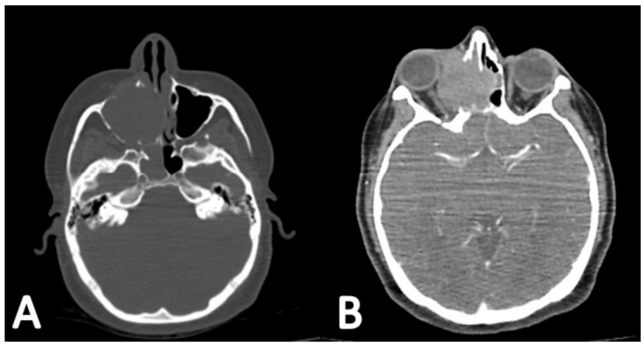

Fig. 1. *CT: A bone window axial plane, expansive process in ethmoidal and orbital region. B soft tissue window after contrast intravenous administration axial plane, showing high enhancement and invasion of the right orbital cavity.*

The MRI showed an isointense T2 solid mass centered in the right nasoethmoidal region invading the right side of the frontal sinus. (Fig. 3) The tumor is seen invading the right orbit with slight lateral displacement of the medial rectus muscle and globe (Fig. 4). Intracranial invasion and hyper enhancement of the mass is seen in the post contrast fat saturated images. (Fig. 2)

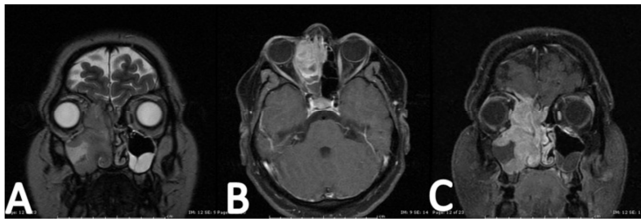

Fig. 2. *Magnetic Resonance (MRI) A. T2 sequence coronal plane showing right isointense mass and mucous retention in left maxillary sinus. B T1 FAT SAT and Gadolinium axial plane, showing lateral displacement of the globe. C T1 FAT SAT and Gadolinium coronal plane, showing a heterogenous process highly enhanced with intracranial invasion.*

Osseous Scintigraphy failed to reveal distant metastases and a bone marrow study of the right iliac crest did not show malignant cells.

A biopsy was performed under general anesthesia showing a solid neoplasm that grew forming lobes, cells with large cytoplasm, oval nucleus with fine chromatin and small nucleolus. Immunohistochemistry tests were positive for Actin and Myogenin and confirmed muscle differentiation (Fig. 3) negative for OCTV4, CD99, NSE, S-100, HMB45, EMA, Keratin AE1/3, Chromogranin and synaptophysin. Final pathology reported a "Solid alveolar rhabdomyosarcoma". Lymphovascular penetration was not observed.

The Head and Neck Tumor Board reviewed the patient's findings and the final diagnosis was determined to be Parameningeal Alveolar Rhabdomyosarcoma, (PAR) T3bN1M0. Chemo-radiation therapy was recommended.

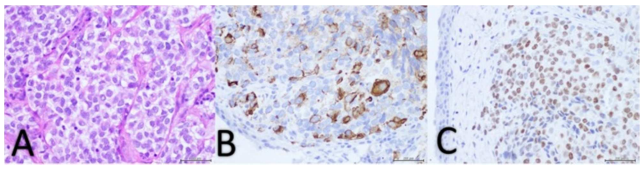

Fig. 3. *Inmunohistochemistry study (20x). A Hematoxylin and eosin stain. B Positive Actin stain. C Positive Myogenin stain.*

Conventional radiotherapy consisted of a total dose of 54 Gy given over 7 weeks in 1.8 Gy increments (Fig. 4). Following radiation therapy the patient was treated with chemotherapy consisting of a VAC regimen for 6 months: Vincristine each week and Dactinomicyn and cyclophosphamide + Mesna every 21 days.

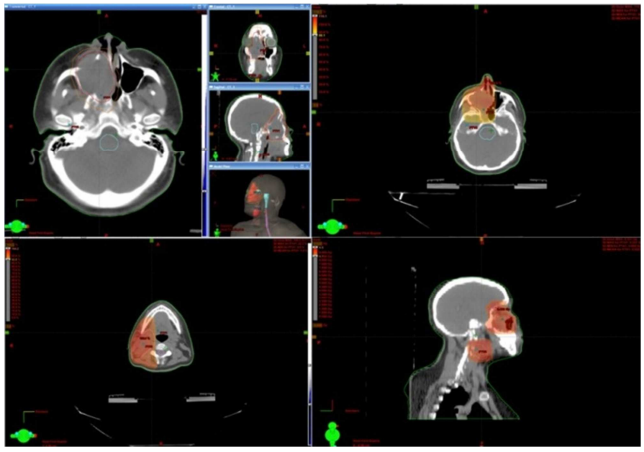

Fig. 4. *Three-dimensional conformal radiation plan.*

Consequently, the patient developed Neovascular Glaucoma in the right eyeball from radiotherapy complication. This was treated with intracameral injection of Avastin, with no complications noted after procedure. A 2 year follow up MRI failed to reveal any ethmoid, maxillary or right nostril expansions. 3 years after treatment was completed, a follow up MRI exam showed recurrence of disease in right nasal nostril with intracranial invasion and lymphoadenopathy in IIA left ganglionic level (Fig. 5). Following additional reviewed by the HNTB, palliative chemotherapy was recommended. He received 4 cycles of Ifosfamide and Etoposide with Mesna, showing regular tolerance and stabilization of nasal tumor. Upon finishing this article, 4 years after diagnosis, the patient remains in palliative care.

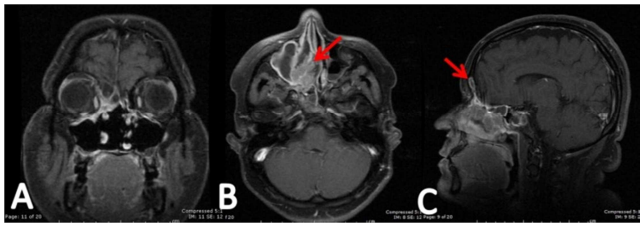

Fig. 5. *Magnetic Resonance (MRI) T1 sequence FAT SAT and Gadolinium. A Follow up 2 years do not showing evidence of residual or recurrent tumor. B and C follow up 3 years showing nasal recurrence and intracranial invasion (red arrows).*

3. Discussion

RMS is a malignant tumor with striated muscle differentiation. It is derived from primitive mesenchyme that retained its capacity for skeletal muscle differentiation. [1, 2] RMS was first described in the English literature in 1937 and in children in 1992. The tumor is mainly composed of bundles of cells with myogenic differentiation by immunohistochemical and ultra structural analysis. Rubin et al. described the first two examples of RMS with spindle cells in adults. Since then and until 2007, 21 cases have been described in the English literature. [3]

This sarcoma is one of the most common soft tissues sarcomas in newborns, children, and young adults. [4] 20 to 25% of the cardiac neoplasms in adults are sarcomas. [5]

The annual incidence of RMS in the USA is 4.6 per million in people under 20 years of age. RMS may occur in all age groups but is more prevalent in the first and second decades of life with a peak between 2 and 6 years of age, [6] It represents approximately 4 - 8% of all pediatric cancers. [7] Although tumors of head and neck are rare in children [8], approximately 60% of pediatric RMS cases occur in the head and neck. [9, 8, 10]

RMS has different grades of striated muscle cell differentiation and it may occur in any part of the body. [9] Four different histopathological types have been described: embryonic, alveolar, pleomorphic and undifferentiated. [6] The two most common histopathological types described in childhood are embryonic and alveolar. [11] The embryonic type represents 70% of cases and it is mainly seen in children under the age of 12 and carries the best prognosis. The alveolar type occurs more frequently in the extremities affecting an older age group. It generally shows the chromosomal translocation (t2: 13; p35-14), carrying a more ominous prognosis than the other types of RMS. [12] The pleomorphic variety is less frequent and occurs more often in an older population. [6]

Anatomically, RMS is classified as parameningeal, orbital and non-parameningeal non-orbital. Approximately 40% of newly diagnosed RMS arises in the head and neck structures including parameningeal sites (16% of all cases, and almost half of all head and neck cases), the orbit or eyelid (10% of all cases), and other non-orbit, non-parameningeal sites (10% of all cases). The parameningeal tumors carry the worst prognosis. [1, 6]

The parameningeal sites include nose, nasopharynx, paranasal sinuses, middle ear, mastoid, infratemporal fossa and pterygopalatine fossa. Soderberg described the first case of an aggressive RMS in the middle ear. RMS of the temporal bone carries a poor prognosis due to its proximity to the brain and vital structures. A review of 20 cases from the literature by Jaffe et al. found a 0% two-year survival rate in [13]

The non-orbital and non-parameningeal forms include scalp, parotid gland, oropharynx, larynx and oral cavity. The tongue, palate and cheeks are the most common oral sites. [6] Of the 35% of RMS that occur in the head and neck, 10-12% present in the oral cavity. RMS rarely occurs in the salivary glands. [14]

RMS can have a syndromic presentation such as their association with Beckwith-Wiedemann syndrome (10% of cases). [13]

Metastasis occurs by hematogenous or lymphatic spread, most commonly to the lungs, bones and brain. [4] Prognosis is influenced by the anatomic location at the time of presentation, patient's age, completeness of resection, extent of metastatic disease and tumor histology. [15]

A multidisciplinary treatment approach is most effective. In the last 30 years the combined use of chemoradiotherapy and surgery has significantly improved the survival rate of head and neck RMS rates to 5 years [6] A study indicates that approximately 65% of children diagnosed with RMS will survive with combined therapy. [8] In the pediatric parameningeal RMS cases, the treatment of choice is chemoradiotherapy with surgery having a limited role due to the relative inaccessibility of the lesions and associated surgical morbidity. [16]

Improved and innovative operative techniques of craniofacial surgical reconstruction have resulted in satisfactory functional and cosmetic results [8]

4. Conclusion

Rhabdomyosarcoma is a rare head and neck tumor in the adult population with poor prognosis despite aggressive therapy. Imaging studies play an important role in providing valuable information related to the involvement of critical anatomical organs.

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