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Case Report

Unmasking rhabdomyosarcoma: A rare nasal cavity tumor in adults

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Abstract

Rhabdomyosarcoma (RMS) is a rare mesenchymal malignant tumor that primarily affects children, with adult cases being exceptionally uncommon. Embryonal RMS of the nasal cavity is an even rarer occurrence. We present a case of a 37-year-old male with progressive nasal obstruction, rhinorrhea, anosmia, and swelling near the medial canthus of the eye.

Imaging revealed an aggressive mass in the left nasal cavity with bony destruction. Histopathological and immunohistochemical analyses confirmed the diagnosis of embryonal RMS. Due to its aggressive nature, RMS requires prompt diagnosis and a multidisciplinary treatment approach, including chemotherapy and radiotherapy. This case underscores the importance of considering RMS in adult sinonasal tumors for timely intervention and improved prognosis.

Keywords: Immunohistochemica, Embryonal RMS, Histopathological, Rhabdomyosarcoma (RMS)

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1. Introduction

Rhabdomyosarcoma (RMS) is a mesenchymal malignant tumor and belongs to soft tissue sarcomas with striated muscle differentiation. This tumor primarily affects adolescents. It accounts for fewer than 1% of all solid malignant tumors in adults, making it a rare diagnosis in this age group.[1] Rhabdomyosarcoma is the most common soft-tissue sarcoma that is most typically found in children under 15 years of age.[2] The rhabdomyosarcoma sites of predilection in adults and children are different.[3] Sultan et al.[4] reported that only 9.3% of all adult rhabdomyosarcomas occurred in the head and neck region.

Only a small number of cases of adult Embryonal RMS have been documented in the literature, making the disease's emergence inside the nasal chambers an even rarer event.

Here, we are describing a similar rare of embryonal rhabdomyosarcoma of nasal cavity in a middle aged male patient.

2. Case Report

Our patient is a 37-year-old gentleman who presented to the ENT department with complaints of rhinorrhea and anosmia since 2 months along with swelling near the medial canthus of eye since 3 weeks. Nasal discharge was insidious in onset, progressively increasing, discharge was mucoid to watery without blood streaks or foul smelling. Patient also had headache more on frontal region, persistent throughout the day for last 2-3 weeks. There was no history trauma/bleeding from nose/ facial pain or blurred vision.

On physical examination, anterior rhinoscopy revealed high deviated nasal septum to the right side and grade 2 adenoid. Clinical examination revealed a mass in the left nasal cavity. Computed tomography (CT) and magnetic resonance imaging (MRI) of the nasal cavity and paranasal sinuses were performed. Axial CT scan with bone window setting shows a poorly defined, heterogeneous, relatively isodense mass of the left nasal cavity. The mass destroys adjacent bony structures and extends to the ipsilateral

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maxillary sinus. MR imaging also revealed the similar findings.

Patient underwent endoscopic nasal biopsy and tissue was sent for histopathological study.

We received multiple grey white soft tissue bits measuring 1.5 x 1 x 0.6 cm. Routine tissue processing was done and hematoxylin & eosin (H&E) stained slides were examined. Histomorphology shows a poorly differentiated malignancy with round cell morphology (**Figure 1**A). Tumor is arranged in sheets, cords and clusters (**Figure 1**B). Individual tumor cells are round to ovoid in shape with hyperchromatic nucleus and scant to moderate pale cytoplasm (**Figure 1**C). Mitotic activity and apoptosis is significant (**Figure 1**D). Surrounding stroma is hyalinised and desmoplastic. Based on histology, the differentials of RMS, Large cell lymphoma, Desmoplastic round cell tumor, NUT carcinoma and Undifferentiated/poorly differentiated carcinoma were considered and extensive immunohistochemistry (IHC) pannels were applied.

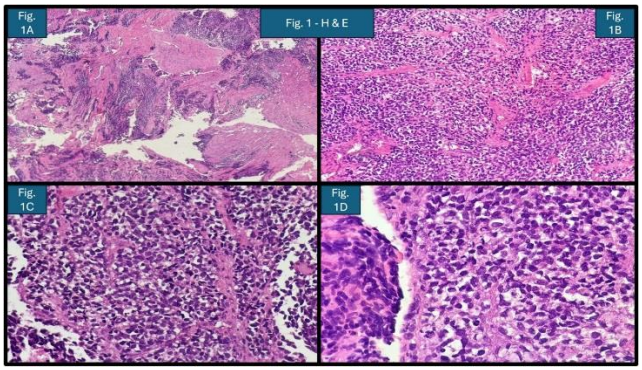

Figure 1: H&E pictomicrographs; **A:** Scanner image (4x) shows a poorly differentiated round blue cell tumor; **B:** Low power (10x) shows a tumor cells arranged in diffuse sheets infiltrating the muscle and connective tissue; **C** & **D:** High power (40x) shows highly pleomorphic tumor cells with large hyperchromatic nuclei and scanty pale cytoplasm exhibiting striking nuclear atypia and increased mitotic activity.

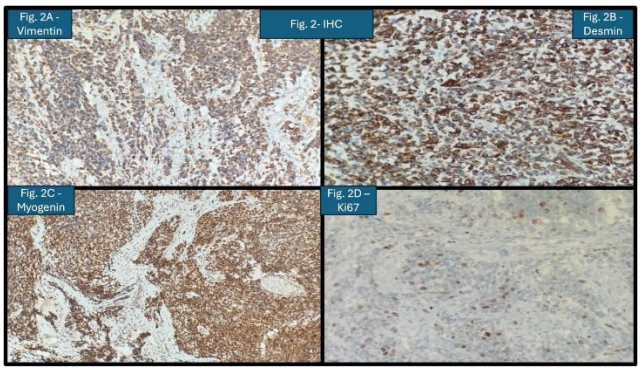

Figure 2: IHC analysis; **A:** Vimentin positive; **B:** Desmin positive; **C:** Myogenin positive; **D:** Ki67 stains 15-20% of nuclei

On IHC (**Figure 2**A to D), tumor cells are diffusely positive for vimentin, desmin & myogenin while negative for pan-CK, LCA, p40, synaptophysin, NKX2.2, EMA & NUT1. Ki-67 index was 15-20%. After IHC analysis, a diagnosis of Embryonal RMS was rendered.

3. Discussion

Weber et al. described RMS for the first time in 1854, and Horn and associates classified it into embryonal, alveolar, and pleomorphic varieties in 1958.[5,6] RMS is the most prevalent soft tissue malignant tumor, primarily affecting youngsters and extremely uncommon in adults. It is thought to replicate the physical characteristics of skeletal muscle cells in their early developing stage.[7] First, the incidence of adult rhabdomyosarcoma is very low, especially in the embryonal type.[8,9] Out of 51 adult cases of rhabdomyosarcoma during a 5-year period, Lee et al. only reported 1 case (2%) of nasal cavity rhabdomyosarcoma.[9] Based on anatomical region, head & neck RMS can be classified further into orbital, parameningeal, and non-orbital non-parameningeal forms. Parameningeal RMS include sites such as middle ear cleft, paranasal sinuses, nasal cavity, pterygopalatine, and infratemporal fossa region.[10] Given its propensity to erode the base of the skull, spread into the cerebral region, and invade the perineural space, it is said to have the worst prognosis. Moreover, poor response to treatment has been noted among the patients.[11,12]

Histopathology and immunochemistry are the gold standards for diagnosing this entity. Tumor cells appear small, round with large hyperchromatic nuclei and scanty pale cytoplasm surrounded by hyalinized stroma.[13] Histopathological findings of EMRS will show un-differentiated small round blue cells and spindled cells with no patterns.[14] Immunohistochemistry is the only way to make a definitive diagnosis of the mass, and myogenin and desmin are unique markers for this diagnosis.[15] In immunohistochemistry, RMS shows strong and diffuse reaction with vimentin and muscle-specific antigens, such as, desmin, muscle actins (including smooth muscle isoforms), myogenin, and MyoD1.[16]

In our case, strong positivity for vimentin, myogenin and desmin were observed that lead to the identification of adult Embryonal RMS.

Treatment for rhabdomyosarcoma should be multidisciplinary and involve radiation, chemotherapy, and surgery.[10,12] Because of the relative inaccessibility of the lesions and the associated surgical morbidity, chemoradiotherapy is the preferred treatment for parameningeal RMS, with surgery playing a limited role.[10] The delay in diagnosis and treatment worsens the already low 5-year survival rates in adult.[17] Adults have a far worse prognosis than children, regardless of treatment, and the outcome is also dependent on the tumor size, histological subtype, and location.[18] Distant metastasis occurs, in adults,

at the time of diagnosis in 60% of cases.[19] Recent clinical trials targeting rhabdomyosarcoma have predominantly focused on pediatric and adolescent populations.[20,21]

4. Conclusion

Despite being an extremely uncommon adult tumor, RMS should not be ruled out due to its aggressiveness and invasiveness, which necessitate both immediate identification and customized therapy. Due to the possibility of metastasis and recurrence, it need ongoing radiological and clinical surveillance. Timely diagnosis and aggressive treatment are essential for improving outcomes.

5. Source of Funding

None.

6. Conflict of Interest

None.

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