

## An Unusual Cause of Nasal Obstruction in Adult

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**Abstract:** Rhabdomyosarcoma (RMS) is a malignant soft tissue tumor with phenotypical and biological features of skeletal muscle cells. This tumor is more common in children and is exceptional in adults. In fact, it accounts for less than 1% of all malignant tumors and 3% of soft tissue tumors in adults; as a result, there is a lack of information regarding the management and prognosis of the patients. In the differential diagnosis with RMS of the paranasal sinuses, the main tumors to be considered are in the category of the so-called 'small round blue cell tumors'. The treatments of those pathologies in this anatomic site are quite different from each other, requiring a definitive diagnosis before starting a therapeutic strategy. We report the case of a 49-year-old women diabetic patient, presented with a 3 months history of a right nasal obstruction. A nasosinusian computed tomography was performed; it objectified a right pansinusitis with complete filling of the ethmoidal cells and the right nasal fossa. The diagnosis of a nasal embryonal botryoid rhabdomyosarcoma was made after surgical excision of the lesion, histologic examination and immunohistochemistry studies.

**Index words:** Embryonal botryoid rhabdomyosarcoma, Adult, Nasal cavity

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### I. Introduction

Rhabdomyosarcoma is a malignant soft tissue tumor with phenotypical and biological features of skeletal muscle cells [1]. This tumor is more common in children and is exceptional in adults. In fact, it accounts for less than 1% of all malignant tumors and 3% of soft tissue tumors in adults [2]; as a result, there is a lack of information regarding the management and prognosis of the patients. RMS arises from mesenchymal tissue and can occur at various sites in the body. In adults, it has a predilection for the extremities, and only 15% of cases occur in the head and neck [3]. Rhabdomyosarcomas are subdivided into different subtypes with the most common being embryonal (ERMS) and alveolar (ARMS), and in adults, the disease carries a significantly worse prognosis.

The histological diagnosis is not always obvious given the small size of the biopsy material and several differential diagnoses are proposed. The treatments of those pathologies in this anatomic site are quite different from each other; therefore a radio-clinical and histological correlation is required to reach a definitive diagnosis.

We present a rare case of 49-years-old women presented with severe bilateral nasal obstruction due to embryonal botryoid rhabdomyosarcoma. We discuss the epidemiological, histopathological, therapeutic and prognostic aspects of this tumor in adult.

### II. Patient And Observation

68-year-old diabetic women presented with a 3 months history of right nasal obstruction.

On clinical examination, a reddish bleeding mass was seen in the right nasal cavity with bilateral jugulo-carotid nodes measuring 3 cm for the largest.

A nasosinusian computed tomography (CT) was performed. It showed a right pansinusitis with complete filling of the ethmoidal cells and the right nasal fossa, associated with jugulo-carotid and spinal nodes measuring 1 cm to 3 cm.

The patient underwent surgical excision of her lesion and histological examination showed a nasal mucosa infiltrated by a diffuse proliferation with a grape-like growth pattern. The cells are small and round, with fine chromatin, eccentric, small oval nuclei, atypical mitotic figures and eosinophilic cytoplasm (Figures 1-2).

Immuno-histochemical studies revealed a strong positivity for Desmine (Figure 3) and a focal staining for Myogenine (Figure 4) antibodies.

Immunostaining for lymphoid markers (CD20, CD3, CD5, CD7 and CD45), CD34, neuroendocrine markers (chromogranin and synaptophysin), melanocyt markers (Melan A, PS100 and HMB45), CD99, NSE and cytokeratin was negative.

The molecular study was not available in our service, but in the study of Lester D. R. Thompson and al (4) has shown that there was no difference in expression of epithelial markers between the cases that were confirmed by molecular evaluation versus those that were not.

The diagnosis of embryonal botryoid rhabdomyosarcoma was retained, and the patient received two cures of chemotherapy and died before the end of cures.

### **III. Discussion:**

Rhabdomyosarcoma (RMS) is the most frequent malignant mesenchymal tumor in children and occurs exceptionally in adults. In fact, it accounts for less than 1% of all malignant tumors and 3% of soft tissue tumors in adult [2]. As far as we know, it has been reported only few cases arising in the paranasal sinuses and orbit [5, 6].

Unlike pediatric RMS, this tumor shows no male predominance in adults [4].

RMS arises from mesenchymal tissue and can occur at various sites in the body, but most common affected sites are head and neck (35%), genitourinary tract (22%) and extremities (18%) [7]. In adults, It has a predilection for the extremities, and only 15% of cases occur in the head and neck, 13% of which is found in the nasosinus area, [3-8-9] in contrast to pediatric RMS which is mostly found in the head and neck [7].

Clinically, as in our case, the main symptoms are nasal obstruction, bleeding and rhinorrhea [3].

Although, the macroscopic aspect is not pathognomonic of RMS, it is assumed that the RMS developed in cavities (bladder, nasopharynx, and sinus) is generally rather limited, multinodular, cystic or polypoid, grayish white mass with a glossy, gelatinous surface. The deep RMS of extremities is less limited, infiltrating, firm or elastic, grayish with a smooth or granular surface [10]. In our case, a reddish bleeding mass was seen in the right nasal cavity but that did not allow us to make any diagnoses based only on the macroscopic aspect of the lesion.

Histologically, three histopathological patterns are seen: embryonal (70%), alveolar (20%) and pleomorphic (10%). In the sinonasal tract, RMS is predominantly of alveolar type with a solid alveolar subtype, mostly comprising undifferentiated tumor cells arranged in a solid alveolar pattern surrounded by dense hyalinized stroma [11]. The embryonal type is rarely seen in this location and it is composed of primitive mesenchymal cells that show variable degrees of skeletal muscle differentiation. They are moderately cellular with alternating hypercellular areas. Sheets of small, stellate, spindled or round cells with scant or deeply eosinophilic cytoplasm and eccentric, small oval nuclei with a light chromatin pattern and inconspicuous nucleoli [10].

The embryonal type is subdivided into three categories: the botryoid, spindle cell, and not-otherwise-specified (NOS).

The botryoid variant, by definition, arises beneath epithelial mucosal surfaces and has a cambium layer, pleomorphism and myxoid stroma. The spindled variant occurs most commonly in scrotal soft tissues.

In our study, based on the histological aspect, we proposed several diagnoses eliminated subsequently by a large immunohistochemical panel. Among this diagnoses, we eliminated a lymphoma by the negativity of lymphoid markers, malignant solitary fibrous tumor eliminated by the negativity of CD34. The neuroendocrine tumors were eliminated by negativity of the neuroendocrine markers. The negativity of melanocyt markers allowed us to eliminate a small cell malignant melanoma, and by the negativity of the Cytokeratin, we have eliminated a Merkel cell carcinoma, thus showing that the diagnosis was not obvious.

The immuno-histochemical study of RMS classically shows an expression of myogenin, myoD [12] and desmin and negativity for cytokeratin, epithelial membrane antigen, CD45, CD 99 and S-100 protein. At the genetic level, alveolar RMS (ARMS) predominantly shows t(2;13) (q35;q14) translocation leading to formation of PAX3/FKHR fusion transcript or t(1;13)(p36;q14) translocation forming PAX7/FKHR fusion transcript.

Concerning adult RMS, Montone and al [10] reported 13 cases of adult parameningeal RMS with 3 cases of ERMS, 9 of ARMS and 1 unclassified. Darren J. Little and al [13] reported 82 cases of adult RMS with embryonal subtype in 28 patients (34%), alveolar in 19 (23%), and pleomorphic in 35 (43%). Emmanuelle Bompas [14] and al reported 157 cases of adult RMS with 33 (21%) cases of ERMS, 55 (35%) of ARMS and 69 (44%) of pleomorphic RMS.

Differentiating embryonal from alveolar RMS can be difficult on histology alone, particularly in a biopsy. The presence of solid alveolar tumor cells beneath the sinonasal mucosa may give a false impression of a cambium layer, posing diagnostic problems in differentiating it from embryonal RMS. In such cases, myogenin positivity is helpful, with ERMS showing <50% positivity vs diffuse nuclear reactivity in ARMS.

In our case, immuno-histochemical studie revealed a strong positivity for Desmine and a focal staining for Myogenine, which allowed us to retain the diagnosis of an ERMS.

In the differential diagnosis with RMS, the main tumors to be considered are in the category of the so-called 'small round blue cell tumors', which in adults should include small cell carcinoma, Merkel cell carcinoma, small cell malignant melanoma, lymphoma and olfactory neuroblastoma. Less commonly in this population, desmoplastic small round cell tumor (DSRCT), and Ewing sarcoma may also be diagnosed [15]. Like in our case, these tumors are eliminated by an immunohistochemical study.

The prognosis of RMS depends on the primary site, histological subtype and tumor size. A favorable prognosis is seen in tumors less than 5 cm, patient age < 20 years, lack of regional or distant metastasis and negative surgical margins. Adult RMS has a poor prognosis as it is usually advanced at presentation, with extensive local involvement making complete surgical excision difficult. The 5-year survival rate is only 8 % in head and neck RMS. RMS in a parameningeal location, such as the nasal cavity poses a significant risk of subarachnoid dissemination.

Most cases of RMS are treated aggressively with a multidisciplinary approach based on surgery, radiation, and chemotherapy [16] with the primary aim of cure, maintaining quality of life and function preservation. First-line surgical resection is considered for tumors of small size, easily accessible, as some limbs or paratesticular tumors [17]. Adjuvant or neoadjuvant chemotherapy depends on the tumor characteristics (location, accessibility, size and operability). Irradiation is only used when surgical removal is impossible, incomplete or in case of local recurrence [17]. Chemotherapy protocols used in adults with RMS are usually derived from pediatric RMS clinical trials conducted by specialists from international cooperative groups [18].

#### **IV. Conclusion:**

Embryonal rhabdomyosarcoma of the nasal cavity is very rare in adults. It should be considered in the differential diagnosis of poorly differentiated soft tissue mass of the nasal cavity not only in children, but also in adults.

#### **Competing interests**

The authors declare that they have no competing of interest.

#### **Authors' contributions**

AMAL DOUIDA drafted the manuscript. All authors read and approved the final manuscript.

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#### **Ethics approval and consent to participate**

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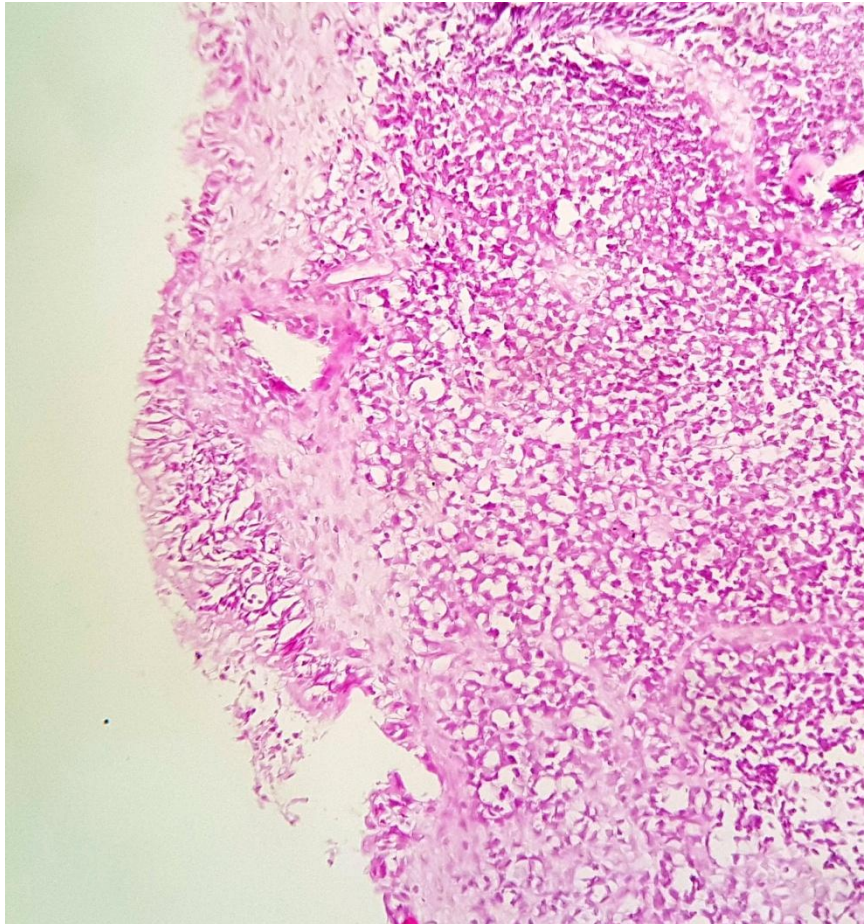
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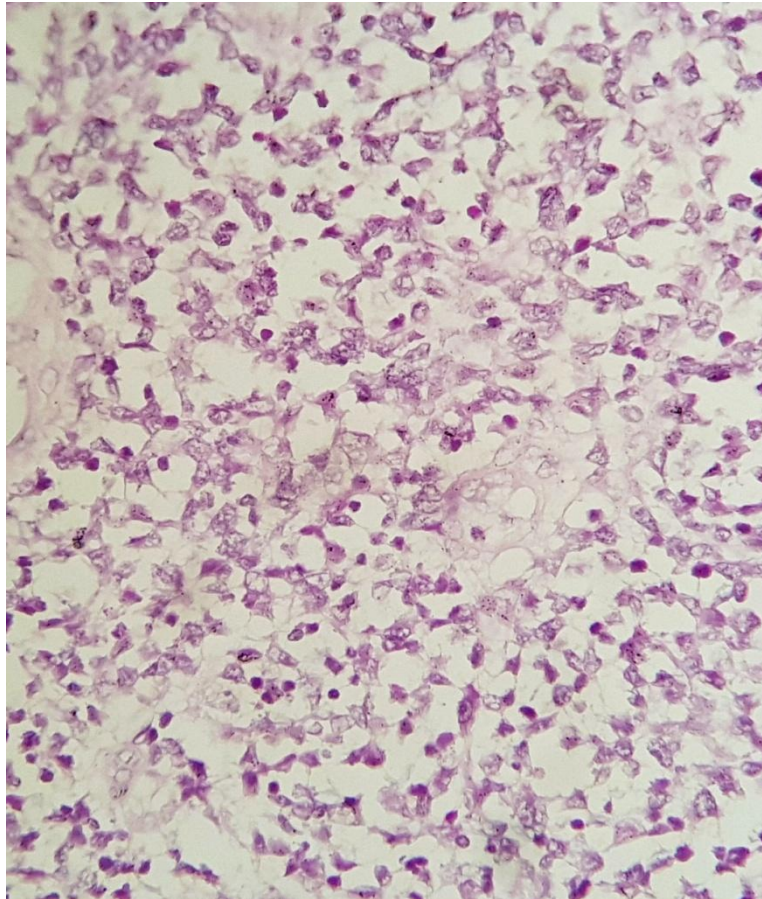
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**LEGENDS**



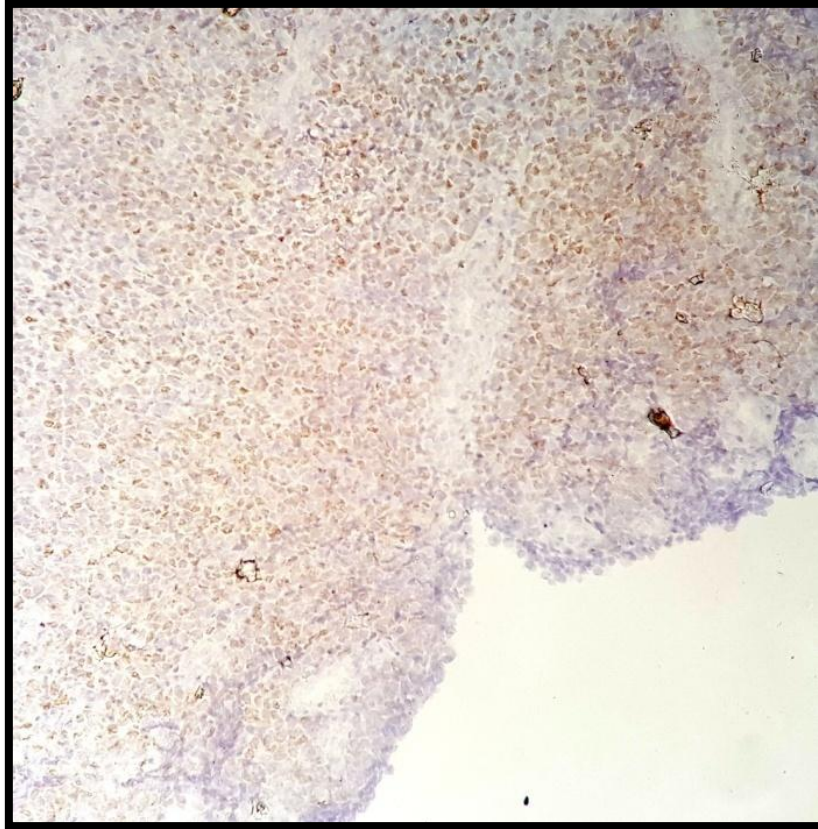
**Figure 1 :** (HESx100) nasal mucosa infiltrated by a diffuse proliferation



**Figure 2 :** (HESx200) tumor proliferation made of small round cells with fine chromatin and eosinophilic cytoplasm



**Figure 3:** Immunohistochemistry showing diffuse positivity for Desmine



**Figure 4:** Immunohistochemistry showing focal positivity for Myogenine

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