Annals of Medicine and Surgery 75 (2022) 103424

Contents lists available at ScienceDirect

Annals of Medicine and Surgery

journal homepage: www.elsevier.com/locate/amsu

Case Report

Adult embryonal rhabdomyosarcoma in the nasal cavity; a case report with a review of the literature

Mariwan L. Fatah [a], Abdulwahid M. Salh [b,c], Ari M. Abdullah [b,d], Fahmi H. Kakamad [b,c,*], Shevan M. mustafa [e,f], Suhaib H. kakamad [g]

[a] Sulaymaniyah Teching Hospital _ENT Center, University of Sulaymaniyah Colloge of Medicine, Iraq
[b] Smart Health Tower, François Mitterrand Street, Sulaimani, Kurdistan, Iraq
[c] College of Medicine, University of Sulaimani, Sulaimani, Kurdistan, Iraq
[d] Sulaimani Teaching Hospital, Sulaimani, Kurdistan, Iraq
[e] Al Qalam University Colloge, MLT Department, Kirkuk, Iraq
[f] Tehran University of Medical Science, Colloge of Medicine, Hematology and Blood Transfusion Department, Tehran, Iran
[g] Kscien Organization, Hamdi Str., Azadi Mall, Sulaimani, Kurdistan, Iraq

ARTICLE INFO

Keywords:
Embryonal rhabdomyosarcoma
Nasal cavity
Adult
Good outcome

ABSTRACT

Introduction: Rhabdomyosarcoma (RMS) is a primitive malignant soft tissue tumor arising from premature mesenchymal cells. The current study presents a rare case of embryonal rhabdomyosarcoma in the nasal cavity of an adult patient.

Case presentation: An 18-year-old female presented with right nasal obstruction for five months. Examination showed a pale soft, painless mass in the right nasal cavity with attachment to the nasal septum at the osteo-cartilaginous junction. The patient was falsely suspected for bacterial infection, but later histological examination showed undifferentiated small round blue cell tumor with extensive necrosis. Immunohistochemistry confirmed the diagnosis of embryonal RMS. The patient was operated on for endoscopic sinus surgery to remove the mass with additional cleaning of surrounding sinuses.

Discussion: Embryonal RMS is a rare type of malignant tumor that mostly affects the head and neck area in children while usually occur in the extremities of adults. Encountering an adult ERMS of the nasal chambers represents a small portion of head and neck cases that lack specific presentations.

Conclusion: Adult embryonal RMS of the nasal cavity is exceedingly rare and presents a diagnostic and management challenge, with immunohistochemistry being the only definitive diagnostic method.

1. Introduction

Rhabdomyosarcoma (RMS) is a primitive malignant soft tissue tumor arising from premature mesenchymal cells with striated muscle differentiation. It is mostly found amongst children and adolescents, with a rare incidence in adults [1]. Histopathologically, RMS is categorized into embryonal, pleomorphic, alveolar, and mixed subtypes [2]. Embryonal Rhabdomyosarcoma (ERMS) in children is commonly reported, especially in the head and neck, which accounts for more than half of RMS incidences, while encountering of adult ERMS in those areas is very unlikely with only 9.3% of all adult ERMSs occurring in the head and neck regions [3,4]. An even rarer incidence of adult ERMS is the appearance of the disease inside the nasal chambers, with only a few reported cases in the literature.

This study aims to present a rare case of embryonal rhabdomyosarcoma in the nasal cavity of an adult patient, with a short review of the literature. The report has been written in line with SCARE 2020 guidelines [5].

Patient information: An 18-year-old female presented with right side nasal obstruction for 5 months. Past medical, surgical, family, and drug history was unremarkable.

Clinical findings: The patient was conscious and vitally stable. There was a pale soft, painless mass in the right nasal cavity. Probing was performed which showed a clear attachment to the nasal septum at the osteo-cartilaginous junction.

Diagnostic assessment: a swab from the mass showed no growth.

* Corresponding author. Doctor City, Building 11, Apartment 50, Sulaimani, Zip code: 0064, Iraq.
E-mail address: fahmi.hussein@univsul.edu.iq (F.H. Kakamad).

https://doi.org/10.1016/j.amsu.2022.103424
Received 3 January 2022; Received in revised form 19 February 2022; Accepted 27 February 2022
Available online 2 March 2022

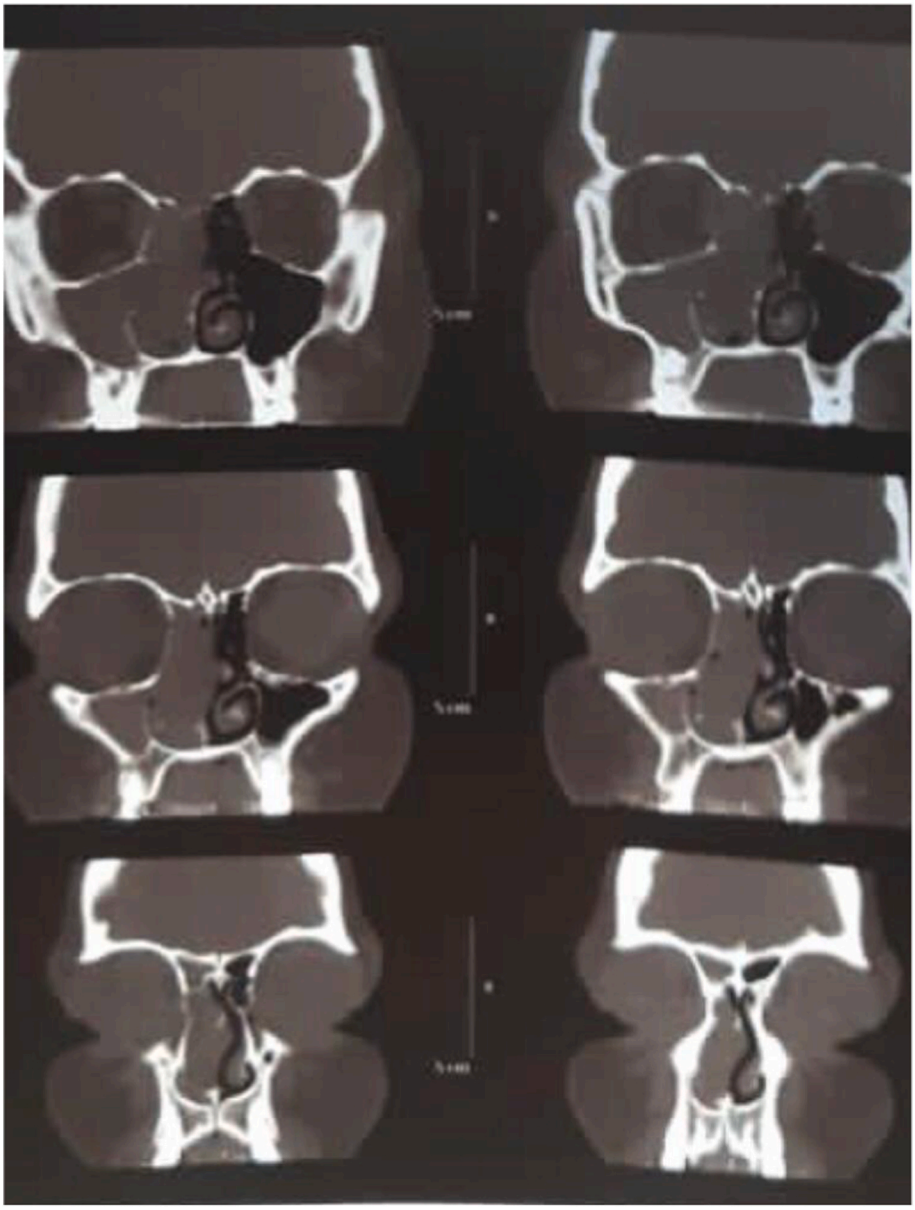

Fig. 1. Computed tomography (CT) scans of the paranasal sinuses (coronal view) showing diffuse mucosal hypertrophy of the right sinuses including frontal, maxillary, ethmoidal and sphenoidal sinuses with obstruction and dilatation of Osteomeatal Complex (OMC).

Computed tomography (CT) scans showed diffuse mucosal hypertrophy of the right sinuses including frontal, maxillary, ethmoidal and sphenoidal sinuses with obstruction and dilatation of Osteomeatal Complex (OMC) (Fig. 1). Histopathological examination of a punch biopsy showed an undifferentiated small round blue cell tumor with extensive necrosis (Fig. 2). Immunohistochemistry staining (CK, Chromogranin, Synaptophysin, S100, CD99, CD34, and Desmin) confirmed the diagnosis of ERMS. CT scans of the chest and abdomen were normal.

Therapeutic intervention: Under general anesthesia, the patient was operated on for endoscopic sinus surgery to remove the mass. The mass was eroding the posterior part of the septum. Hence, posterior septectomy, middle meatal antrostomy, and posterior ethmoidectomy were done. The right frontal sinus ostium and right sphenoid sinus were opened and cleaned.

Follow-up and outcomes: The operation was uneventful, and the mass was fully removed. The patient was healthy four months after the surgery.

2. Discussion

RMS is the most common malignant neoplasm of the soft tissues, which is predominantly found in children with extreme rarity in adults. It is believed to recapitulate the early developmental form of skeletal muscle cells [6]. The first description of RMS was by Weber et al., in 1854, from which in 1958 it was categorized by Horn and colleagues into embryonal, alveolar, and pleomorphic types [7,8]. It has been observed that ERMS mainly affects the head and neck area of the pediatric population while occurring most frequently in the extremities of adults. Adult ERMS of the nasal chambers represent only 10%–15% of head and neck cases [9].

Even though RMS is thought to be derived from primitive mesenchymal cells that tend to differentiate into striated skeletal muscles, however, they can be found in other parts of the body where the skeletal muscle is lacking, such as; gall bladder, prostate, and urinary bladder, creating confusion regarding their origins [10].

The range of presentations regarding RMS might change drastically

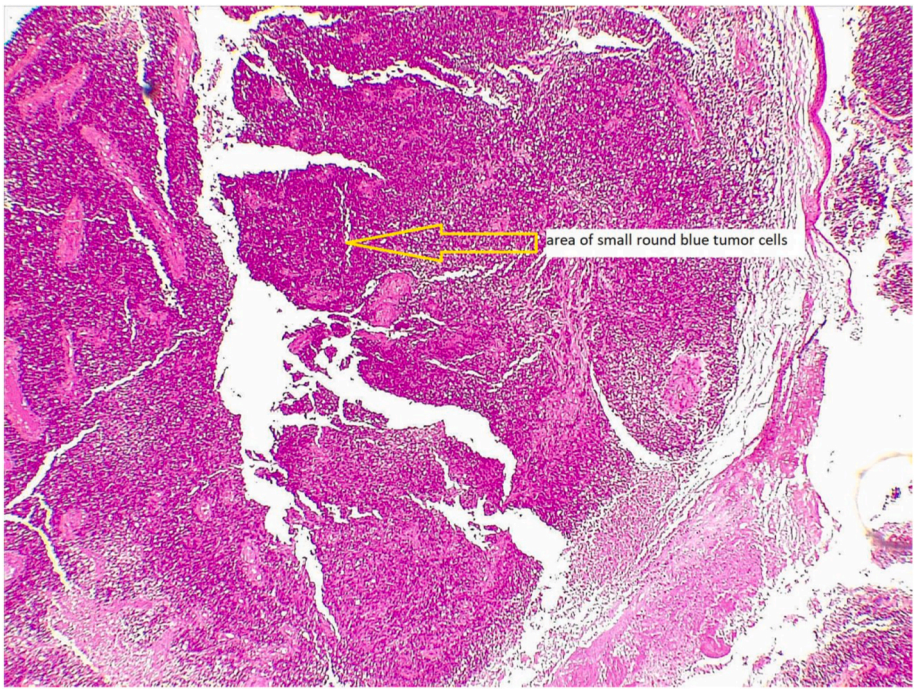

Fig. 2. Microscopic examination of the specimen showing small blue round cells with malignant proliferation. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

based on their primary sites. ERMS might usually appear as a soft brown or dark yellow mass in association with areas of hemorrhage and necrosis [11]. Even though several studies reported the presence of nasal obstruction and swelling of the affected area in ERMS patients, specific presentation and symptoms regarding ERMS of the nasal cavity is not well described [4,12]. The case in the current study presented with nasal obstruction due to a soft pale mass in the nasal cavity with no other significant symptoms.

The diagnosis of ERMS presents a challenge to physicians as it is sometimes mistaken for a microbial infection or abscess, but eventually, the failure to acquire isolates and the increase in mass size will lead to achieving a biopsy from the mass and assigning of the mass as possible neoplasm [13]. Imaging techniques such as ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI) are not reliable for the definite diagnosis of such tumors and produce similar findings in both adult and pediatric ERMS, but they can be used to show tumor location, margin, extension, size, and heterogeneity of the tumor to muscles [14,15]. Histopathological findings of EMRS will show undifferentiated small round blue cells and spindled cells with no patterns [16]. Definitive diagnosis of the mass can only be achieved via immunohistochemistry, with myogenin and desmin being specific markers [14]. In the current study, similar findings were observed that lead to the identification of adult ERMS.

The recommended management approach in children is surgical excision in combination with radiotherapy and chemotherapy [17]. However, due to the tumor location and proximity to vital systems, complete radical excision is not always possible, and due to the aggressive and invasive nature of these tumors in adults, other adapted managements are required [9]. The management of the current case also went through according to the same protocol by completely excising the tumor and cleaning relevant surrounding areas, followed by chemoradiotherapy.

While the prognosis of ERMS might depend on age, the primary site, tumor size, and sub-type, clinical outcome and prognosis in adult patients are more flawed than that of the pediatric population [9,11]. It has been reported that distant metastasis of ERMS can occur in adults, and the 5-year survival rate is said to be nearly 60% [18]. Thus, the current study is associated with an excellent clinical outcome.

In conclusion, ERMS of the nasal cavity is an exceedingly rare type of malignant tumor in adults that presents a diagnostic and management challenge, with immunohistochemistry being the only definitive diagnostic tool. Furthermore, due to their poor prognosis and clinical outcome compared to the pediatric population, adult ERMS might require particular treatment adaptations.

Ethical approval

The manuscript approved by ethical committee of Kscien organization.

Sources of funding

No source to be stated.

Registration of research studies

Not applicable
.

Guarantor

Fahmi Hussein Kakamad is Guarantor of this submission.

Consent

Written informed consent was obtained from the patient's family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Abdulwahid M. Salh: major contribution of the idea, literature review, final approval of the manuscript.

Mariwan L. Fatah: Surgeons performing the operation, final approval of the manuscript.

Ari M. Abdullah, Shevan M. Mustafa, Suhaib H. kakamad and Fahmi H. Kakamad: literature review, final approval of the manuscript.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

None to be declared.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.103424.

References

[1] A.E. Chang, X. Chai, S.M. Pollack, E. Loggers, E. Rodler, J. Dillon, et al., Analysis of clinical prognostic factors for adult patients with head and neck sarcomas, Otolaryngology-Head Neck Surg. (Tokyo) 151 (6) (2014) 976–983.
[2] D.M. Parham, Pathologic classification of rhabdomyosarcomas and correlations with molecular studies, Mod. Pathol. 14 (5) (2001) 506–514.
[3] R. Taseer, T.T. Ahmed, Embryonal rhabdomyosarcoma presenting as lung metastasis in an adult: an uncommon presentation, Cureus 13 (2) (2021) 1–8.
[4] M.S. Kang, H.J. Kim, Embryonal rhabdomyosarcoma of the nasal cavity in an adult: a case report, Investig. Magn. Reson. Imag. 13 (1) (2009) 97–100.
[5] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.
[6] G. Patil, S. Halawar, S. Sagari, R. Babannavar, S. Purohit, Embryonal rhabdomyosarcoma occurring on mandibular gingiva in an adult, J. Clin. Diagn. Res.: J. Clin. Diagn. Res. 7 (9) (2013) 2088.
[7] O. Miloglu, S.S. Altas, M.C. Buyukkurt, B. Erdemci, O. Altun, Rhabdomyosarcoma of the oral cavity: a case report, Eur. J. Dermatol. 5 (3) (2011 Jul) 340.
[8] R.C. Horn Jr., H.T. Enterline, Rhabdomyosarcoma: a clinicopathological study and classification of 39 cases, Cancer 11 (1) (1958) 181–199.
[9] M. Hamou, Y. El Amrani, A. Lachkar, A. Eabdenbitsen, F. Elayoubi, R. Ghailan, Adult embryonal rhabdomyosarcoma of the nose and paranasal sinuses misdiagnosed as an orbital cellulitis: a case report, PAMJ-Clinic. Med. 5 (3) (2021) 1–7.
[10] D.M. Parham, D.A. Ellison, Rhabdomyosarcomas in adults and children: an update, Arch. Pathol. Lab Med. 130 (10) (2006) 1454–1465.
[11] C.D.M. Fletcher, J.A. Bridge, P.C.W. Hogendoorn, F. Mertens (Eds.), WHO Classification of Tumors of Soft Tissue and Bone, fourth ed., International Agency for Research on Cancer (IARC), Lyon, 2013, pp. 123–136.
[12] E. Palacios, A. Quiroz-Casian, L.G. Garcia, P.J. Daroca, H.R. Neitzschman, Adult case of large sinonasal embryonal rhabdomyosarcoma with intracranial extension, Ear Nose Throat J. 92 (5) (2013) 177–178.
[13] M. Duval, R. Faingold, A.S. Carret, S.J. Daniel, Nasal ala rhabdomyosarcoma misdiagnosed as a facial infection, Int. J. Pediatr. Otorhinolaryngol. Extra 3 (1) (2008) 44–47.
[14] A. El Sanharawi, B. Coulibaly, J.P. Bessede, K. Aubry, Paranasal sinus rhabdomyosarcoma: a rare tumor of poor prognosis, Eur. Ann. Otorhinolaryngol. Head Neck Dis. 130 (1) (2013) 26–29.
[15] J.H. Lee, M.S. Lee, B.H. Lee, D.H. Choe, Y.S. Do, K.H. Kim, et al., Rhabdomyosarcoma of the head and neck in adults: MR and CT findings, Am. J. Neuroradiol. 17 (10) (1996) 1923–1928.
[16] C.A. Hardaway, B.S. Graham, D.J. Barnette, B.D. Feldman, Embryonal rhabdomyosarcoma presenting in an adult: a case report and discussion of immunohistochemical staining, Am. J. Dermatopathol. 25 (1) (2003) 45–52.
[17] G. Moretti, R. Guimarães, K.M. Oliveira, F. Sanjar, R.L. Voegels, Rhabdomyosarcoma of the head and neck: 24 cases and literature review, Braz. J. Otorhinolaryngol. 76 (4) (2010) 533–537.
[18] W.M. Crist, J.R. Anderson, J.L. Meza, C. Fryer, R.B. Raney, F.B. Ruymann, et al., Intergroup rhabdomyosarcoma study-IV: results for patients with nonmetastatic disease, J. Clin. Oncol. 19 (12) (2001) 3091–3102.