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Ethmoidal alveolar rhabdomyosarcoma with orbital extension in a young adult: A case report and literature review

Ahmad Abbas
ahmadabbas20049@gmail.com
Arab American University
Ro'ya Khanfar
An-Najah National University
Razan Haj
Palestinian Ministry of Health
Hidaya Kmail
Arab American University
Kholoud Rehan
Palestinian Ministry of Health
Firas Jallad
Palestinian Ministry of Health
Razan Odeh
An-Najah National University

Case Report

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Abstract

Background

Rhabdomyosarcoma (RMS) is an uncommon malignant neoplasm of skeletal muscle differentiation that predominantly affects children. Sinonasal RMS is rare in adults, and the alveolar subtype tends toward aggressive local growth with regional and distant spread. Its clinical and radiologic presentation can overlap with inflammatory, infectious, and other malignant sinonasal processes, which makes diagnosis difficult.

Case presentation

A 28-year-old Middle Eastern man presented with a one-week history of right-sided proptosis and right eye pain, associated with diplopia and painful upward gaze but without prominent sinonasal symptoms. Computed tomography demonstrated a heterogeneous expansile mass centered in the right ethmoid and maxillary sinuses with extension through the lamina papyracea into the right orbit. Endoscopic sinus surgery with tumor debulking and biopsy demonstrated erosion of the lamina papyracea and extensive involvement of the ethmoid air cells. Histopathology showed an infiltrative malignant small round blue cell neoplasm arranged in nests and lobules separated by delicate fibrovascular stroma, with brisk mitotic activity, including seven mitotic figures per high-power field. Tumor cells showed diffuse strong nuclear expression of myogenin and MyoD1 and retained INI1. Pan-cytokeratin, CK5/6, chromogranin, synaptophysin, S100, CD99, FLI1, CD20, and CD3 were negative. Molecular testing was positive for FOXO1 rearrangement, supporting the diagnosis of alveolar rhabdomyosarcoma. Staging demonstrated no definite distant metastatic disease. Systemic chemotherapy with ifosfamide and etoposide was initiated, and the patient remains under treatment and follow-up.

Conclusion

Adult sinonasal alveolar rhabdomyosarcoma is rare and can be difficult to diagnose; it may present with orbital manifestations despite limited sinonasal symptoms. Its characteristic morphology, myogenic immunophenotype, and FOXO1 rearrangement help establish the diagnosis. Management requires a multidisciplinary team and careful evaluation of orbital and intracranial extension.

Introduction

Rhabdomyosarcoma (RMS) is an uncommon malignant tumor characterized by skeletal muscle differentiation and occurs predominantly in children and adolescents. In adults, sinonasal RMS is a particularly rare subset. In a population-based series of 186 adults with sinonasal RMS, the alveolar subtype accounted for 66.7% of cases, metastatic disease was present in 27.7%, and 5-year overall survival was 28.4% [1]. Another population-based analysis of 286 patients reported a median age of 24.5 years, with 5-, 10-, and 20-year disease-specific survival rates of 35.1%, 25.4%, and 12.0%, respectively; regional lymph-node involvement and distant metastases occurred in 54.3% and 32.2% of patients [2].

Adult sinonasal alveolar rhabdomyosarcoma (ARMS) can be difficult to distinguish from other small round blue cell neoplasms of the sinonasal tract. A clinicopathologic study of 52 adult sinonasal ARMS cases demonstrated frequent myogenic marker expression and pointed to molecular assessment, particularly FOXO1 rearrangement testing, as a key diagnostic tool [3]. Imaging may demonstrate aggressive local extension involving the orbit, skull base, and adjacent structures [4]. Given its rarity, aggressive behavior, and overlap with other sinonasal malignancies and inflammatory conditions, this tumor calls for early tissue diagnosis and multidisciplinary management.

We report a 28-year-old man with ethmoidal ARMS presenting predominantly with acute orbital symptoms and describe the clinical, radiologic, surgical, histopathologic, immunohistochemical, and molecular findings, together with a focused review of the relevant literature.

Case presentation

A 28-year-old Middle Eastern man presented with a one-week history of right-sided proptosis and right eye pain. He reported diplopia and pain on upward gaze. There were no major sinonasal symptoms at presentation. The patient's medical history was

unremarkable, with no significant comorbidities or previous surgical interventions. The patient was not taking any regular medications at the time of presentation.

Computed tomography of the paranasal sinuses demonstrated a heterogeneous expansile mass centered in the right ethmoid and maxillary sinuses. The lesion extended through the lamina papyracea into the right orbit, involving the regions of the medial and superior rectus muscles. The initial differential diagnosis included an invasive fungal process as well as neoplastic and other infectious causes (Fig. 1).

The patient underwent functional endoscopic sinus surgery under general anesthesia. Following nasal decongestion, a 0° endoscope was introduced. The middle turbinate was medialized, and the uncinate process was removed to gain access to the ethmoid sinus. An anterior and posterior ethmoidectomy was subsequently performed. A mass was identified within the ethmoid air cells and was debulked and excised for histopathologic examination.

The lamina papyracea was found to be eroded by the lesion. Dissection was performed carefully to avoid injury to the medial rectus muscle. Involved ethmoid partitions were removed to achieve orbital decompression, followed by hemostasis and nasal packing.

Histopathologic examination demonstrated sinonasal mucosa lined by respiratory epithelium with underlying connective tissue/periosteum and seromucinous glands. Infiltrating the tissue was a malignant small round blue cell neoplasm arranged predominantly in nests and lobules separated by delicate fibrovascular stroma.

The tumor cells had scant to moderate cytoplasm, round to oval nuclei, finely granular chromatin, and variably conspicuous nucleoli. Mitotic activity was brisk, with seven mitotic figures per high-power field. No necrosis, keratinization, gland formation, or overt squamous differentiation was identified (Fig. 2).

Immunohistochemical staining demonstrated diffuse strong nuclear expression of myogenin and MyoD1, supporting skeletal muscle differentiation. INI1 expression was retained. The tumor was negative for Pan-CK, CK5/6, chromogranin, synaptophysin, S100, CD99, FLI1, CD20, and CD3 (Fig. 3).

The absence of epithelial markers argued against sinonasal carcinoma, while negative neuroendocrine and S100 staining made neuroendocrine and neural-lineage neoplasms less likely. The lack of CD99 and FLI1 expression argued against Ewing sarcoma. Retained INI1 expression also argued against an INI1-deficient sinonasal malignancy. Together, the characteristic morphology and diffuse myogenic marker expression pointed to rhabdomyosarcoma, and molecular testing demonstrating a FOXO1 rearrangement confirmed the diagnosis of alveolar rhabdomyosarcoma.

Computed tomography of the chest, abdomen, and pelvis identified a tiny approximately 3-mm pulmonary nodule and an approximately 8-mm subpleural ground-glass opacity in the lingula, for which follow-up imaging was recommended. No definite metastatic disease was identified.

Postoperative magnetic resonance imaging demonstrated heterogeneous diffusion-restricting abnormal tissue in the right ethmoid region with extension toward the sphenoid and maxillary sinuses. The margins adjacent to the medial and superior rectus muscles were indistinct, although there was no definite intraconal extension. An enhancing diffusion-restricting lesion measuring approximately 1.8 × 1.6 cm in the right maxillary sinus was suspicious for possible residual tumoral involvement. Enhancement was also noted along the cribriform plate, without evidence of meningeal or intracranial extension. The optic nerves were unremarkable (Fig. 4).

Subsequent FDG PET/CT demonstrated expected postsurgical changes without abnormal focal uptake suggestive of metabolically active residual tumor. Bilateral symmetric upper jugular lymph nodes measuring up to 1.3 cm demonstrated an SUVmax of 4.5 and were favored to be reactive. A rounded hyperdense lesion in the right maxillary sinus demonstrated an SUVmax of 4.4 and was favored to represent an inflammatory retention cyst. No suspicious distant FDG-avid disease was identified.

Given the diagnosis of adult sinonasal ARMS, systemic chemotherapy with ifosfamide and etoposide (IE) was initiated. The patient experienced mild nausea during treatment. He also had one episode of shortness of breath during a previous chemotherapy cycle, which did not recur. During treatment, folliculitis involving the chest and back improved with doxycycline and topical benzoyl peroxide. The patient remains clinically stable under ongoing treatment and follow-up.

The major events in the patient's clinical course, from initial presentation and diagnosis through surgical management, systemic staging, and ongoing oncological treatment, are summarized in (Fig. 5).

Discussion

Adult sinonasal ARMS is rare, and its presentation can differ substantially from the more familiar pediatric form of RMS. Population-based studies have shown poor long-term outcomes in adults with sinonasal RMS, with a substantial proportion presenting with regional or distant disease [1,2]. Because the tumor is rare, diagnosis is often uncertain at first, particularly when the presenting symptoms are dominated by orbital findings rather than nasal obstruction, epistaxis, or other sinonasal complaints.

Our patient presented with acute unilateral proptosis, orbital pain, diplopia, and painful upward gaze. Imaging demonstrated a mass centered in the ethmoid and maxillary sinuses with extension through the lamina papyracea into the orbit. Orbital involvement is common among adult sinonasal RMS cases, with one imaging series reporting orbital involvement in 81.5% of patients and skull involvement in 55.6% [4]. The ethmoid sinuses sit directly against the orbit, so tumors here can reach the orbit early even when sinonasal symptoms are still minimal.

The principal published adult series and imaging studies relevant to sinonasal RMS are summarized in Table 1.

Table 1
Clinicopathologic and imaging characteristics of adult sinonasal rhabdomyosarcoma in published series

Study	Year	Study design / patients	Histology	Main anatomical sites / imaging findings	Treatment / outcome
Stepan et al. [1]	2017	Population-based analysis; 186 adults with sinonasal RMS	ARMS most common (66.7%)	Sinonasal RMS; metastatic disease in 27.7%	Chemotherapy + radiotherapy most common; 5-year OS 28.4%; metastatic disease associated with worse survival
Thompson et al. [3]	2018	Clinicopathologic series; 52 adults with sinonasal ARMS	All 52 ARMS	Large destructive nasal/sinonasal masses; multiple contiguous paranasal sites in 46/52; cervical adenopathy in 41/52	Surgery 16; radiotherapy 41; chemotherapy 45; high-stage disease common; **mean OS 36.1 months**
Liu et al. [4]*	2023	Retrospective MRI study; 27 adults with sinonasal RMS	ERMS 14; ARMS 7; mixed 6	Maxillary mucosal involvement 51.9%; botryoid enhancement 55.6%; skull involvement 55.6%; orbital involvement 81.5%; lymph-node metastasis 66.7%	MRI demonstrated characteristic infiltrative sinonasal disease and frequent orbital/skull involvement
Durzynska et al. [5]	2023	Retrospective cohort; 13 adults	Sinonasal RMS	Maxillary sinus 7; ethmoid sinus 5; sphenoid sinus 1	Multimodal management; survival and prognostic factors evaluated
Wang et al. [16]	2017	MRI comparison study; 12 sinonasal RMS vs 52 sinonasal carcinomas	RMS subtypes not limited to ARMS	Skull involvement in 8/12 RMS cases; heterogeneous enhancement in 10/12	Multiparametric MRI findings, including ADC, may help distinguish RMS from sinonasal carcinoma
Hahn et al. [17]	2022	Adult head and neck RMS cohort	Embryonal/alveolar and pleomorphic RMS	Head and neck primary sites	Multimodality treatment; evaluated management, outcomes, and effect of IMRT on locoregional control

*The paper is an MRI comparison study but is included because it provides relevant imaging data for adult sinonasal RMS, although it was not restricted to alveolar histology.

The principal diagnostic challenge in this case was the small round blue cell morphology. The sinonasal tract contains a broad differential for such tumors, including Ewing sarcoma, sinonasal carcinoma, neuroendocrine malignancies, lymphoid neoplasms, and other poorly differentiated tumors. The morphology alone is therefore insufficient. In this case, diffuse nuclear myogenin and MyoD1 expression established skeletal muscle differentiation, while the negative epithelial, neuroendocrine, neural, hematolymphoid, and Ewing-associated markers helped narrow the differential [3].

The FOXO1 rearrangement provided an additional molecular feature supporting ARMS. FOXO1 rearrangements are strongly associated with alveolar RMS, and molecular confirmation is particularly useful when morphology and immunophenotype overlap with other small round blue cell neoplasms. A systematic review and meta-analysis of head and neck RMS reported PAX-FOXO1 fusion in a substantial proportion of molecularly characterized alveolar tumors [12]. In the present case, the positive FOXO1 rearrangement, together with the characteristic nested morphology and diffuse myogenic marker expression, supported the final diagnosis.

Imaging serves two roles here: identifying the primary lesion and mapping its orbital, skull-base, and intracranial relationships. MRI in this patient raised concern for possible residual disease in the right maxillary sinus and demonstrated postoperative abnormality near the orbital muscles and cribriform plate. However, the subsequent PET/CT showed no convincing metabolically active residual disease. These findings illustrate the limitations of postoperative imaging: inflammatory and postsurgical changes may mimic tumor, while the absence of FDG avidity cannot definitively exclude microscopic residual disease.

Surgical management in sinonasal ARMS must balance tissue diagnosis and local control against the risk of injury to critical orbital and skull-base structures. In this patient, endoscopic surgery allowed direct visualization and biopsy of the ethmoid lesion, while careful dissection around the lamina papyracea and medial rectus enabled orbital decompression without documented injury to the extraocular muscles.

Recent literature on head and neck sarcomas supports an individualized, multidisciplinary approach. A retrospective study of adult head and neck sarcomas described a wide range of histologic subtypes, surgical approaches, and reconstructive requirements [19]. For extensive orbital resections, a series of 78 exenterations from Gustave Roussy proposed planning the reconstruction around the eventual prosthesis, with surgeons and prosthetists working together [20], although exenteration is not needed in every case of orbital involvement. Imaging remains essential for defining tumor extent and narrowing the differential diagnosis. A retrospective CT series of 163 patients with osteosarcoma, chondrosarcoma, or Ewing sarcoma of the maxillofacial bones found that CT features helped separate these subtypes, but its results should not be extrapolated directly to sinonasal soft-tissue tumors [21]. A recent case report described a 37-year-old woman with sinonasal ARMS who presented with acute, nonspecific ocular and nasal symptoms, which overlaps with the ocular presentation in our patient [22]. For sinonasal ARMS with orbital extension, the extent of surgery should be decided for each patient from imaging and pathology, with input from surgery, oncology, and radiology.

Because adult sinonasal ARMS is uncommon, treatment strategies are largely extrapolated from pediatric RMS protocols and individualized according to disease extent and multidisciplinary assessment. Published adult head and neck ARMS cases have described combinations of chemotherapy, surgery, and radiotherapy, with regimens including VAC, VAI, and VIE [18]. In the present case, systemic chemotherapy with ifosfamide and etoposide was initiated after surgical diagnosis and staging.

Several published adult cases describe sinonasal or paranasal ARMS extending into the orbit and initially mimicking more common inflammatory or ophthalmologic conditions [6–11,13–15]. The reported cases, including their sites of origin, patterns of orbital involvement, treatment, and outcomes, are summarized in Table 2. Taken together with the present case, these reports argue for keeping ARMS on the differential whenever an adult presents with an orbital mass and an adjacent sinonasal lesion.

Table 2
Published adult cases of sinonasal/alveolar rhabdomyosarcoma with orbital involvement.

Author	Year	Age / Sex	Primary site	Orbital involvement	Clinical presentation	Treatment	Outcome
Moon et al. [6]	2006	48 / M	Ethmoid sinus	Orbit; medial rectus compression	Proptosis, orbital symptoms	Debulking + orbital decompression + systemic therapy	Cervical/spinal metastases; died after 7 months
Bagdonaite et al. [8]	2013	52 / M	Paranasal sinuses	Orbital extension	Orbital inflammation, diplopia	Chemoradiotherapy	In remission at 2-year follow-up
Sanz-Marco et al. [9]	2014	25 / F	Ethmoid sinus	Orbit	Orbital discomfort; initially mimicked sinusitis	Chemotherapy + radiotherapy	Complete remission and resolution of orbital symptoms
Parikh et al. [10]	2014	40 / M	Maxillary sinus	Left orbit	Facial pain, nasal congestion, epistaxis, epiphora	Chemotherapy ± radiotherapy	Cervical lymph-node metastases; undergoing treatment
Torres-Peña et al. [11]	2015	24 / M	Ethmoid sinus	Right orbit	Orbital involvement	Chemotherapy + radiotherapy	Recurrence/systemic disease; died after 24 months
Torres-Peña et al. [11]	2015	26 / M	Nasal cavity	Orbit; ethmoid/frontal extension	Orbital/nasal presentation	Chemotherapy + radiotherapy	Systemic disease; died after 5 months
Eshraghi et al. [7]	2016	23 / M	Ethmoid sinus	Orbit; later recurrent medial orbital mass	Anosmia, proptosis; later orbital recurrence	Chemotherapy + radiotherapy	Orbital recurrence; subsequently treated; no further recurrence reported at 2 years
Liu et al. [14]	2016	Adult / M	Sphenoid sinus	Orbital apex/cavernous sinus	Visual symptoms mimicking optic neuritis	Multimodal therapy	Aggressive sinonasal ARMS
Joo et al. [13]	2018	21 / F	Maxillary sinus	Orbit	Proptosis, nasal obstruction	Surgery + VAC chemotherapy + radiotherapy	Complete regression; ocular function preserved at 1 year
Ahmad Fahmi et al. [15]	2023	Adults; 2 cases	Nasal cavity / ethmoid region	Orbital extension in reported cases	Nasal/orbital symptoms	Multimodal management	Illustrates aggressive adult sinonasal ARMS
Current case	2026	28 / M	Ethmoid sinus	Orbit; medial/superior rectus relationship; no definite intraconal extension	Unilateral proptosis, ocular pain, diplopia, painful upward gaze	Endoscopic surgery/debulking + systemic therapy	No convincing distant metastatic disease on staging; ongoing treatment/follow-up

This case combines an uncommon adult sinonasal ARMS arising in the ethmoid region, early orbital presentation, detailed endoscopic surgical findings, a characteristic myogenic immunophenotype, and molecular confirmation by FOXO1 rearrangement testing. It also shows, in practice, how imaging, pathology, and molecular findings fit together when working up an aggressive sinonasal small round blue cell tumor.

Limitations

This report has the limitations inherent to a single-case study. Long-term oncologic outcome is not yet available because the patient remains under active treatment and follow-up. Complete details regarding chemotherapy dosing and the total number of cycles

administered were not available at the time of manuscript preparation. In addition, postoperative MRI findings were suspicious for possible residual tumor, whereas PET/CT did not demonstrate metabolically active residual disease; therefore, microscopic residual disease cannot be definitively excluded on imaging alone.

Conclusion

Adult sinonasal alveolar rhabdomyosarcoma is a rare, aggressive malignancy that may present predominantly with orbital symptoms despite limited sinonasal complaints. In patients with an aggressive sinonasal small round blue cell tumor, the characteristic morphology and myogenic immunohistochemistry point to the diagnosis, and molecular demonstration of FOXO1 rearrangement can confirm it. Management depends on accurately assessing orbital and skull-base extension and coordinating care across surgery, oncology, and radiology.

Declarations

Declarations

Ethics approval and consent to participate :

Ethics approval was not required for this case report.

Consent for Publication:

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

Competing interests

The authors declare that they have no competing interests.

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Author Contribution

AA contributed to data collection, validation, writing of the original draft, writing—review and editing, and final approval of the version to be published. RK contributed to data collection, writing of the original draft, and writing—review and editing. RH contributed to data collection, writing of the histopathological and pathology descriptions, and supervision. HK contributed to writing—review and editing and final approval of the version to be published. KR provided clinical supervision of the case and contributed to writing—review and editing. FJ provided supervision of the histopathological evaluation and pathology-related aspects of the case. RO contributed to supervision and final approval of the version to be published. All authors reviewed and approved the final manuscript and agreed to be accountable for all aspects of the work.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

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Figures

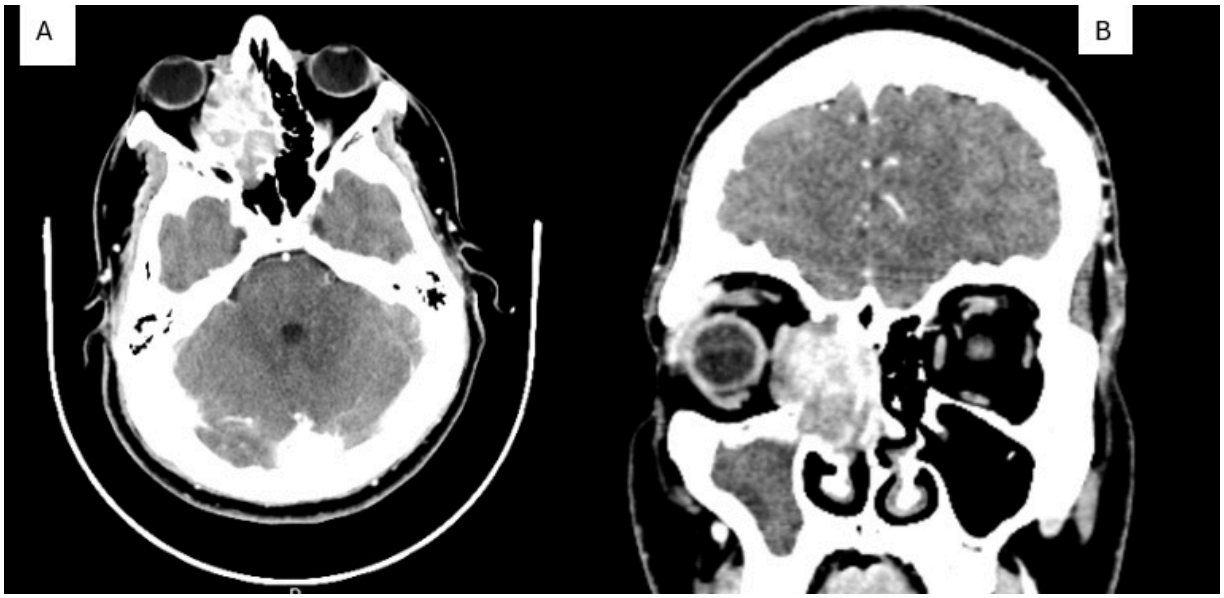


Figure 1

Initial contrast enhanced computed tomography of the paranasal sinuses. (A) Axial and (B) coronal CT images demonstrating a heterogeneous soft-tissue mass centered in the right ethmoid and maxillary sinuses, with extension through the lamina papyracea into the right orbit and involvement of the medial and superior orbital regions.

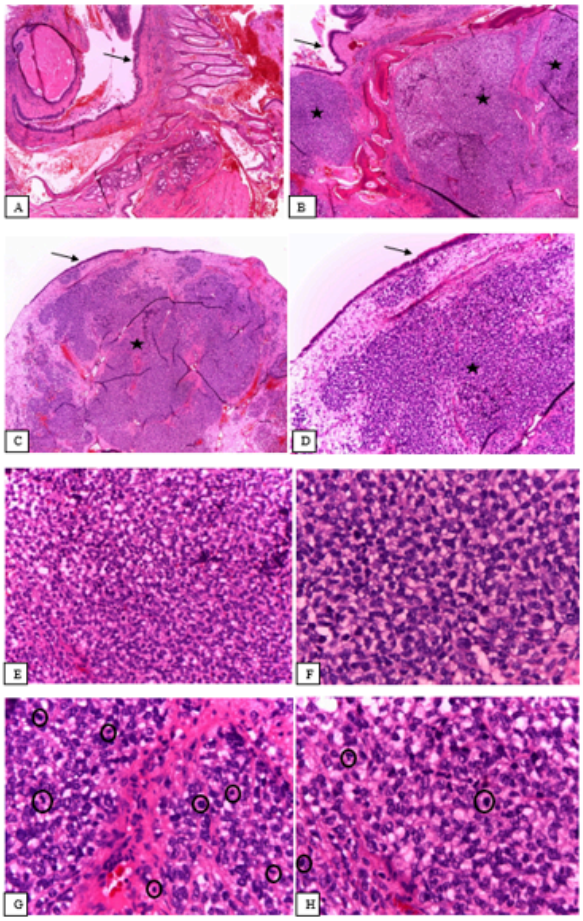


Figure 2

Photomicrographs of ethmoid sinus sections show sinonasal mucosa lined by respiratory-type epithelium (black arrow) with an underlying thin layer of connective tissue tightly bound to the underlying periosteum of the ethmoid bone and seromucinous glands. An infiltrative malignant neoplasm is seen beneath the epithelial lining (black star) (A, B, C: H&Ex4). and composed of nests and lobules of small- to intermediate-sized round blue cells separated by fibrovascular stroma (D: H&E x10, E: H&E x20). The tumor cells exhibited scant-to-moderate cytoplasm and round-to-oval nuclei with finely granular chromatin and variably prominent nucleoli (F: H&E x40). Brisk mitotic activity is identified, 7 mitotic figures per one high power field are identified- black circles (G, H: H&E x40).

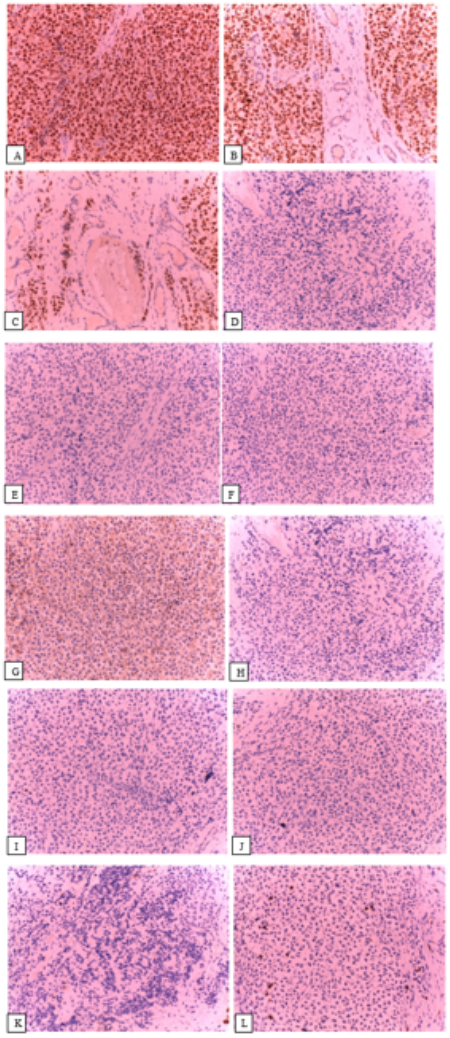


Figure 3

Photomicrographs of immunohistochemical panel shows as following; strong diffuse positive staining for myogenin and MyoD1 (A,B: H&Ex20, respectively), retained nuclear expression for INI1 in tumor cells (C: H&E x20), and negative staining for Pan-CK, CK5/6, S100, CD99, chromogranin, synaptophysin, FLI1, CD20 and CD3 in tumor cells (D-L: H&E x20, respectively).



Figure 4

Postoperative axial magnetic resonance imaging of the brain and paranasal sinuses. Axial contrast-enhanced MRI demonstrating heterogeneous, diffusion-restricting abnormal tissue involving the right ethmoid air cells with extension into the right sphenoid and maxillary sinuses and adjacent medial orbital region. There is no definite intraconal or intracranial extension.

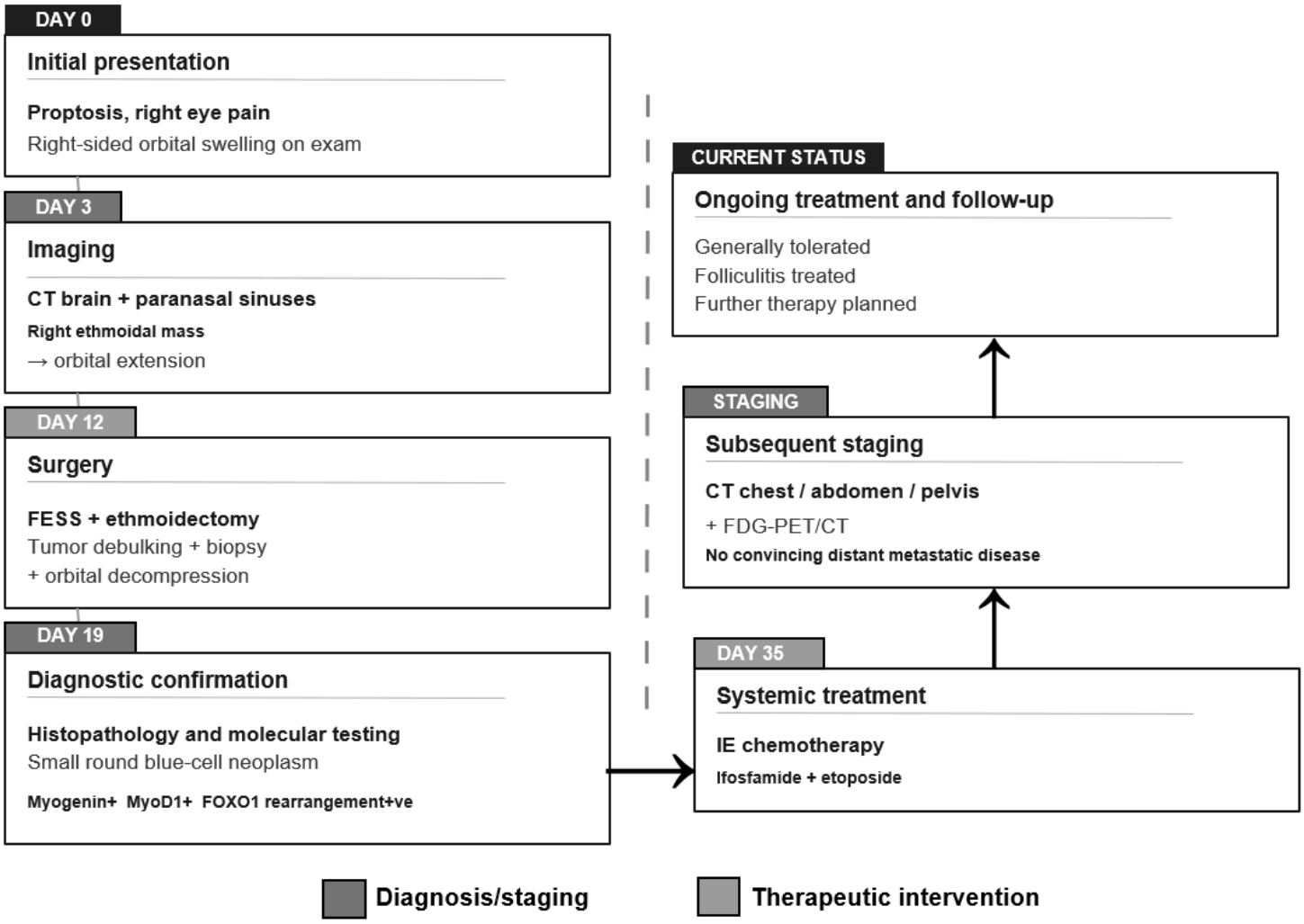


Figure 5

Timeline of the patient's clinical course. The timeline summarizes the major clinical events, including initial presentation with right-sided proptosis and orbital symptoms, diagnostic imaging, endoscopic surgical management, histopathological diagnosis of rhabdomyosarcoma, postoperative imaging and systemic staging, initiation of ifosfamide and etoposide chemotherapy, treatment-related adverse events, and ongoing oncological follow-up.

Supplementary Files

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