

# Orbito-Ethmoidal Rhabdomyosarcoma in an Adult Patient: A Case Report and Review of the Literature

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## Key Words

Computed tomography · Magnetic resonance imaging · Ethmoidal region · Rhabdomyosarcoma · Radiochemotherapy

## Abstract

We report a patient who presented to the ENT service complaining of nasal obstruction, exophthalmos, edema and ipsilateral facial congestion. Imaging studies revealed an aggressive noncalcified solid mass centered in the left nasoethmoidal region and heterogeneous avid enhancement following contrast media injection. Subsequently, a biopsy confirmed the presence of solid alveolar rhabdomyosarcoma. The patient was treated with chemoradiation therapy for 7 weeks. Due to the advanced stage of the disease, the patient was enrolled in a palliative care and pain control program.

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## Introduction

Rhabdomyosarcoma (RMS) is a malignant tumor of striated muscle origin and derives from primitive mesenchyme that retains its capacity for skeletal muscle differentiation. It is one of the most common sarcomas in newborns and children. Approximately 35% of all RMS occur in the head and neck. The combined use of chemoradiotherapy and surgery improves the survival rate significantly, namely up to 5 years.

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## Case Presentation

We report on a 42-year-old male patient who, without a significant clinical history, presented to the ENT service complaining of nasal obstruction, exophthalmos, edema and ipsilateral facial congestion. CT and MRI were performed. CT showed an aggressive, noncalcified, solid mass centered in the left nasoethmoidal region invading the left side of the frontal sinus, left maxillary sinus and the left orbit. There was no evidence of intracranial invasion (fig. 1, fig. 2). T2-weighted MRI showed a mildly hypointense, solid mass centered in the left nasoethmoidal region invading the left side of the frontal sinus. The mass was abutting the left. The tumor was also invading the left orbit with lateral displacement of the medial rectus muscle and globe (fig. 3). Intracranial invasion and heterogeneous avid enhancement of the mass was seen in the postcontrast fat-saturated images (fig. 4, fig. 5).

A biopsy was performed under local anesthetic. Immunohistochemistry tests were positive for desmin (fig. 6), myogenin (fig. 7), vimentin (fig. 8), actin (fig. 9), S-100, chromogonin and synaptophysin and negative for NSE (neuron-specific enolase). Final pathology reported a solid, alveolar RMS. The case was presented and discussed at the Head and Neck Tumor Board and a decision was made to initiate chemoradiation therapy consisting of 4 cycles of ifosfamide, doxorubicine, vincristin and mesna. Follow-up imaging of the patient demonstrated disease progression with intracranial invasion. Conventional radiotherapy treatment was started with a total dose of 63 Gy, administered in 1.8-Gy doses during 7 weeks (fig. 10). Disease progression was seen in spite of chemoradiotherapy. The patient was referred to the pain control and palliative care program.

## Discussion

RMS is a malignant tumor with striated muscle differentiation deriving from primitive mesenchyme [1] that retains its capacity for skeletal muscle differentiation [2]. RMS was first described in the English literature in 1937 and in 1992 in children as a tumor mainly composed of bundles of cells with myogenic differentiation by immunohistochemical and ultrastructural analysis. Rubin et al. [3] 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 [4].

This sarcoma is one of the most common soft tissue sarcomas in newborns, children, and young adults [5]. 20–25% of the cardiac neoplasms in adults are sarcomas [6]. The annual incidence of RMS in the USA is 4.6 per million in subjects 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 [7], representing approximately 4–8% of all pediatric cancers [8]. Although head and neck tumors are rare in children [9], approximately 60% of all pediatric RMS cases occur in the head and neck [9–11].

RMS has different grades of striated muscle cell differentiation and it may occur in any part of the body [10]. Four different histopathological types have been described: embryonal, alveolar, pleomorphic and undifferentiated [7], with embryonal and alveolar being the two most common histopathological types described in childhood [12]. The embryonal type represents 70% of all cases, is mainly seen in children under the age of 12 and carries the best prognosis. The alveolar type is more frequent 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 [13]. The pleomorphic variety is less frequent and occurs more often in an older population [7].

Anatomically, RMS are classified as parameningeal, orbital, nonparameningeal and nonorbital. Approximately 40% of all newly diagnosed RMS arise in 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 nonorbit, nonparameningeal sites (10% of all cases). The parameningeal tumors carry the worst prognosis [7, 14].

The parameningeal sites include the 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 [15]. RMS of the temporal bone carries a poor prognosis due to its proximity to the brain and vital structures. Jaffe et al. found a 2-year survival rate of 0% in a review of 20 cases from the literature [16].

The nonorbital and nonparameningeal forms include the scalp, parotid gland, oropharynx, larynx and oral cavity. The tongue, palate and cheeks are the most common oral sites [7]. Of the 35% of all RMS that occur in the head and neck, 10–12% present in the oral cavity. RMS rarely occur in the salivary glands [17].

RMS can have a syndromic presentation such as their association with Beckwith-Wiedemann syndrome (10% of all cases) [18]. Metastasis occurs by hematogenous or lymphatic spread, most commonly to the lungs, bones and brain [7]. The 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 [19].

A multidisciplinary treatment approach is most effective. The combined use of chemoradiotherapy and surgery has significantly improved the survival rate of head and neck RMS to 5 years in the last 30 years [7]. A study indicates that approximately 65% of the children diagnosed with RMS survive with combined therapy [9]. 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 [20]. Improved and innovative operative techniques of craniofacial surgical reconstruction have resulted in satisfactory functional and cosmetic results [9].

In conclusion, RMS is a rare head and neck tumor that occurs in the adult population and has a poor prognosis despite aggressive therapy.

## Disclosure Statement

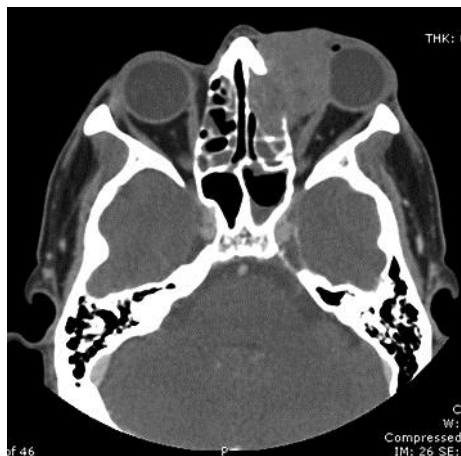
The authors have no conflicts of interest to declare.

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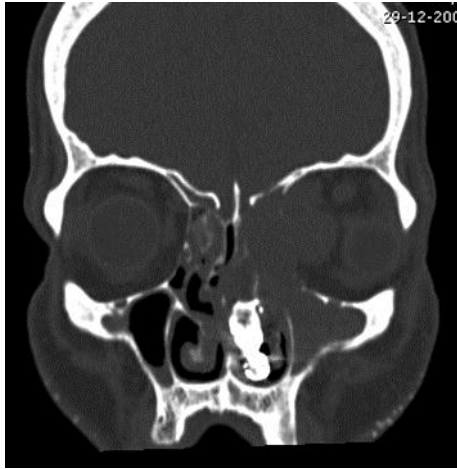
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**Fig. 1.** Expansive process in the ethmoidal and orbital region.



**Fig. 2.** Invasion of the left frontal and maxillary sinuses.



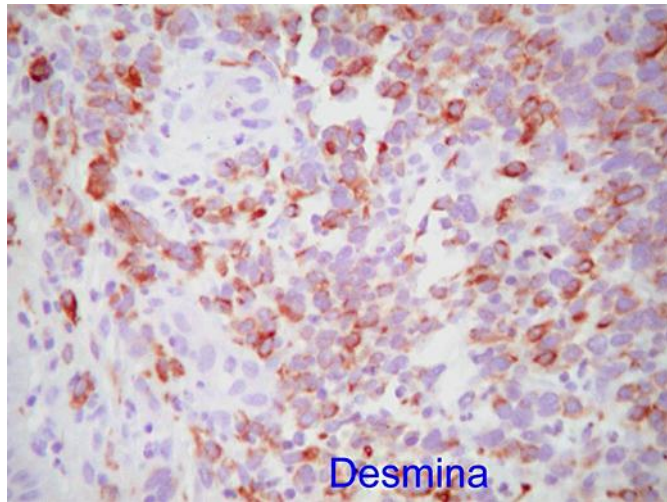
**Fig. 3.** MRI, T2 sequence. Mildly hypointense mass without intracranial invasion.



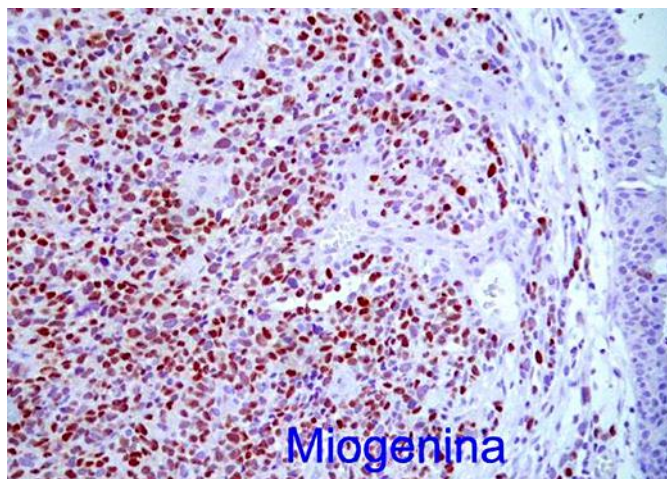
**Fig. 4.** T1-weighted MRI FSGD (fat sat and gadolinium). Heterogenous avid enhancement.



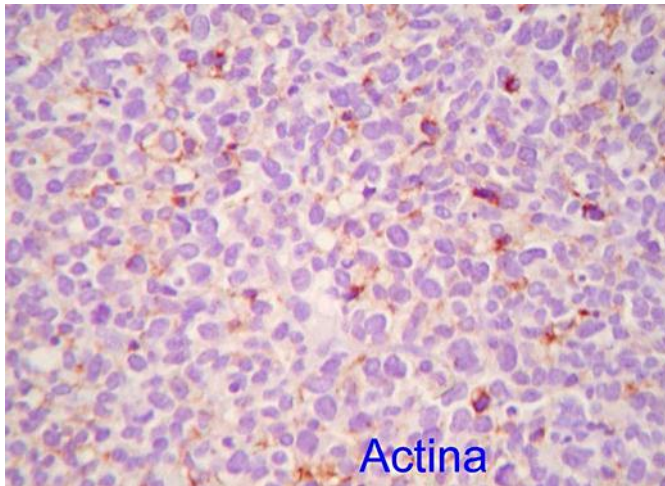
**Fig. 5.** T1-weighted MRI FSGD (fat sat and gadolinium). Lateral displacement of the globe. Mucous retention in the left maxillary sinus.



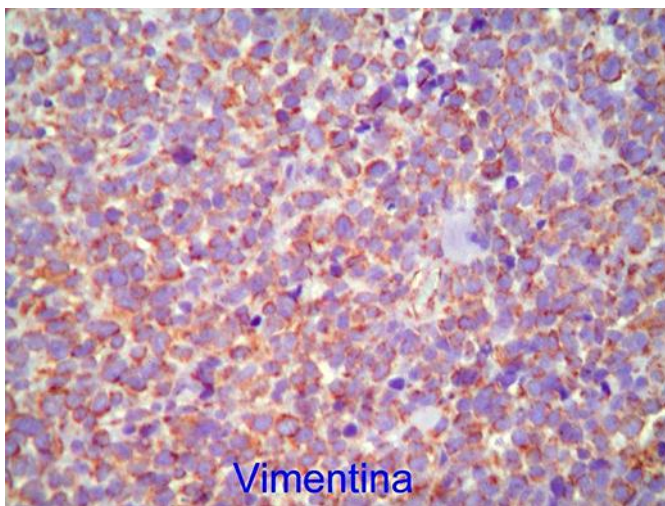
**Fig. 6.** Desmin-positive staining.



**Fig. 7.** Myogenin-positive staining.



**Fig. 8.** Actin-positive staining.



**Fig. 9.** Vimentin-positive staining.



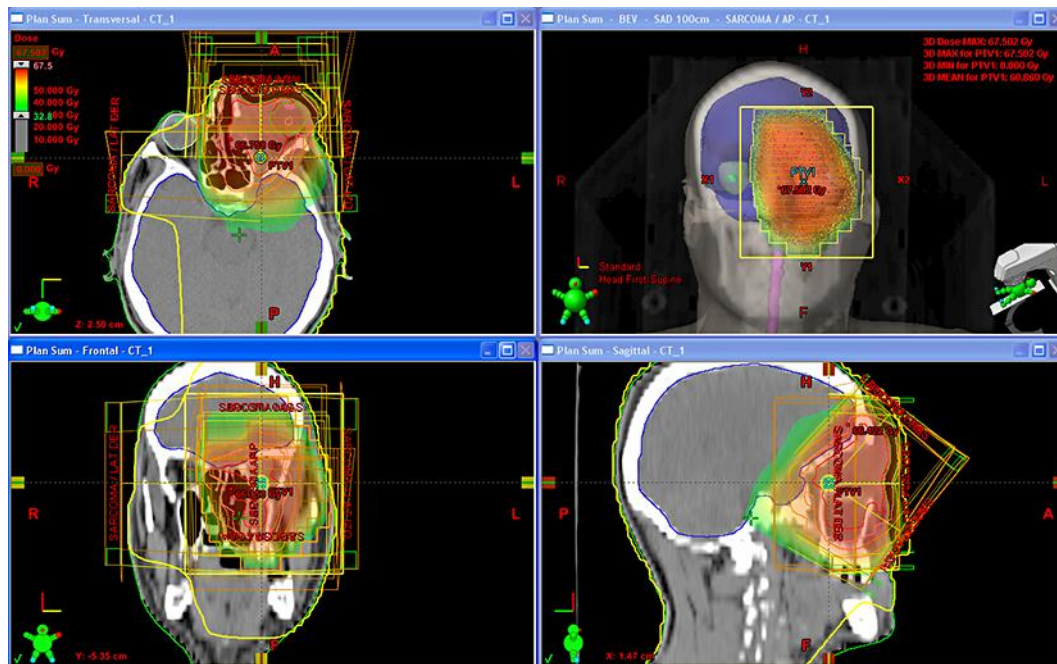


Fig. 10. Tridimensional conformal radiation plan.