Case Report

Minimally Invasive Approach for Orbital and Parameningeal Rhabdomyosarcoma

Joel Caballero-García¹, Iosmill Morales-Pérez¹, Adolfo Michel Giol-Álvarez², Lázara Yaniet Martínez-Espinosa³, Edelberto Gamez-Pérez⁴ and Carlos Aparicio-García¹

¹Neurosurgeon, National Institute of Oncology and Radiobiology
²Neurosurgeon, Juan Manuel Márquez Pediatric Hospital
³Oncologist, National Institute of Oncology and Radiobiology
⁴Pathologist, National Institute of Oncology and Radiobiology

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ABSTRACT

Rhabdomyosarcoma (RMS) of head and neck region in adult is rare. Orbital and parameningeal subtypes constitute a treatment challenge due its relationship to the skull base and orbit. We report a 21-year-old male patient who presented with right ocular proptosis, bilateral nasal obstruction, frontal headache, right nasal discharge, epiphora and decrease of visual acuity in the right eye for the past 6 months. Computed Tomography (CT) and Resonance Magnetic Images (RMI) scans documented a large mass of the right nasal cavity, occupying totally the ethmoid and maxilar sinuses with markedly extension into the right orbit and anterior cranial base erosion with post contrast homogeneous enhancement. A previous biopsy of the neoplasm revealed an esthesioneuroblastoma. An endonasal endoscopic approach extended to the anterior cranial base and right orbit was performed with orbital preservation. Minimally invasive pericranial flap was used for the reconstruction. There were not transoperative or postoperative complications. 24 hours CT and three months postoperative RMI scans show a gross total resection. Definitive biopsy informs an embryonal RMS. He was immediately referred to chemoradiotherapy. Orbital parameningial RMS is a rare entity in adults but should be included in differential diagnosis. Endonasal endoscopic resection offers a faster recovery with minimal morbidity, providing a better life quality and immediately adjuvant treatment. Minimally invasive pericranial flap constitute a good alternative in cases with non-viable mucosal vascularized nasoseptal flap. Multimodality treatment, including chemoradiation and surgery, play an important role in the management.

Highlights

Rhabdomyosarcoma is a rare soft tissue malignancy derived from myogenic cells.
There is very little literature that provides substantial evidence regarding the outcomes of surgical treatment of parameningeal subtype.
Endonasal endoscopic resection offers a faster recovery with minimal morbidity, providing a better life quality and immediately adjuvant treatment.
Minimally invasive pericranial flap constitute a good alternative in cases with non-viable mucosal vascularized nasoseptal flap.
Multimodal treatment ae essential.

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Introduction

Rhabdomyosarcoma is a rare soft tissue malignancy derived from myogenic cells. It represents the most common soft tissue sarcoma of childhood and adolescence and the most common pediatric soft tissue neoplasm affecting the nasal cavity and paranasal sinuses [1]. On the other hand, it is unusual in adults, represented only 10% of soft tissue sarcomas and less than 1% of malignant nasosinus tumors [2]. These patients have more aggressive disease and a poorer prognosis [3].

We report a 21-year-old male patient with nasosinus embryonal Rhabdomyosarcoma with orbital and anterior cranial base invasion treated by endonasal endoscopic approach and quimioradiotherapy.

Case Report

A 21-year-old male patient presented with progressive right ocular proptosis, bilateral nasal obstruction, frontal headache, right nasal discharge, epiphora and decrease of visual acuity in the right eye for the past 6 months. Clinical exam showed severe right ocular proptosis with quemosis, inferior and lateral displacement of the eyeball and complete ophthalmoplegia. The right fundus showed optic nerve avulsion. Neurological exam was negative. There were not cervical lymph nodes associated.

Computed Tomography (CT) scan documented a large isodense mass of the right nasal cavity, occupying totally the ethmoid and maxilar sinuses with markedly extension into the right orbit and anterior cranial base erosion with post contrast homogeneous hyperdensity. Magnetic Resonance Image (RMI) showed this lesion isointense on T1 and hiperintense on T2 weighted images. In spite of the markedly orbital extension the lesion appeared well demarcated with a hipodense rim corresponding to the periorbita (figure 1, A). There were no intraparenchymal cerebral lesions (Figure: 1, A-C).

A biopsy of the neoplasm in the nasal cavity was performed and initially informed an esthesioneuroblastoma and the pretreatment classification was IV-A (TNM-UICC).

An endonasal endoscopic approach extended to the anterior cranial base and right orbit was performed. During nasal step a wide septostomy (two thirds of posterior nasal septum) was perform in order to facilitate a binarial approach using a four hands two surgeons technic as previously reported [4]. Then, centripetal removal of the lesion was carried out until both choanae and sphenoidal rostrum were visible. After that, a wide sphenoidotomy was performing in order to expose the posteroinfior margin of the dissection and to use both optocarotid recess as landmarks.

The frontal sinus was approached by Draf type III sinusotomy that represents the anterosuperior margin of the dissection. After that, a subperiosteal dissection of the naso-ethmoidal-sphenoidal complex was performed bilaterally to expose their lateral margins. Endoscopic medial maxillectomy type III was performed to remove the maxilar component of the tumor and obtain good control of the whole maxillary sinus. The anterior two thirds of orbital floor and right lamina papyracea were included in the dissection. The periorbita limited the tumor extension and it was removed in order to obtain better oncological margins. The partial removal of right nasal bone facilitated the resection of the most anterior component of the orbital extension. Nasolacrimal duct exposure and resection, just below the lacrimal sac was performed. The bilateral ethmoid roof exposure with removal of bony partitions was completed using a drill with a diamond burr. The anterior and posterior ethmoidal arteries were exposed, cauterized with bipolar electric forceps, and dissected. The crista galli was carefully detached from the dura mater and removed with blunt instruments. Then, the dura mater was incised and circumferentially cutting with a falciform scalpel at a safe distance from the suspected area of tumor spread. The falk cerebri was sectioned in a anterior posterior way at the level of the sphen-ethmoidal planum. Fortunately, there was not brain invasion. The surgical margins were checked by frozen section.

The resulting skull base defect was reconstructed by the endoscopic endonasal multilayer technique using autologous underlay fat, inlay fascia and pericranial flap minimally invasive harvested as described previously by Zanation and cols [5]. At the end of the surgery ocular proptosis and quemosis were reversed completely (figure 1, B). A Foley catheter balloon maintained the pericranial flap until the fifth day of surgery when it was removed. The flap was observed well attached, viable and there were not postoperative fistulae.

Surgical bleeding of 680 ml and surgical time was 310 minutes. There were not transoperative or postoperative complications. The patient was extubated in the operating room and stayed in ICU only the first 24 hours after surgery. 24 hours postoperative CT scan showed minor frontal pneumoencephalus without hematoma or other complications. The hospital stay was 5 days. Postoperative MRI – three months later – confirm total removal of the lesion.

Definitive biopsy showed a mass composed of polygonal to round blue cells. Immunohistochemical studies showed tumor cells positive for Myo D1, highly specific and sensitive for Rhabdomyosarcoma and negative to vimentin, desmin, cytokeratins and S-100 protein. (figure 4). Definitive diagnosis was Embrional Rhabdomyosarcoma. The extension study was negative. The patient was immediately discussed in our staff and referred to chemo- therapy and Intensive Modulated Radiation Therapy to the right middle face.

Figures
Figure 1: Preoperative and postoperative images. A: preoperative coronal T1-weighted image shows the extensive lesion extended into the orbit. Hypointense margins apparently well definite correspond to the medial orbital wall and periorbita. B: preoperative axial T1-weighted image shows the extension into the orbital compartment. C: preoperative sagittal T2-weighted image shows the anterior cranial base infiltration (black arrow). D: 24 hours postoperative sagittal reconstruction of CT scan shows the extent of the resection and the absence of pneumoencephalus or other complications. E: 24 hours postoperative axial CT scan shows the removal of the tumor and ocular globes in their normal position. F: 3 months postoperative sagittal T2-weighted image shows the local control and the reconstruction of anterior cranial base (periocranium).

Figure 2: Different surgical steps. A: endoscopic view revealed an irregular mass occupying the whole right nasal cavity, which easily bleeds on touch. B: after the whole sphenoidotomy both lateral optic-carotid recesses were visible and useful as a landmark. C: removing the orbital component of the tumor. A portion of periorbita was resected as an oncological margin. D: opening the dura of anterior cranial base with a falciform scalp. E: after the partial dural removal, the exposed frontal lobes and draft III procedure that limit the most anterior part of the cranial base resection. F: The minimally invasive pericranial flaps harvested widely cover the anterior cranial base defect and the sellar floor. cr=clival recesses; fr=frontal recesses communicating after the Draft III procedure; p=periorbita; t=tumor

Figure 3: Preoperative (A) and immediately postoperative (B) photographs of the patient. Completely and immediately resolution of proptosis and chemosis may be observed.

Figure 4: Photomicrographs of the surgical specimens. A: Low magnification reveals diffuse small round cells (hematoxylin and eosin, original magnification ×20). B: Immunohistochemistry staining was diffusely nuclear-positive for myogenin (original magnification ×40).

Discussion
Soft tissue sarcoma of the head and neck in an adult patient is very rare with only several cases reported in literature [1, 2, 6-10]. RMS is a rare and aggressive malignancy first described in 1854 by Weber and colleagues possibly originating from primitive mesenchymal cells that arise anywhere in the body, including sites where striate muscle is not found like nasal cavity and paranasal sinuses which represent only 10% of soft tissue sarcomas and less than 1% of malignant nasosinusal tumors in adults [11, 12].

According to this fact, only a very small portion of Rhabdomyosarcoma cases has been reported to develop in the adult population older than 20 years [2]. This data corresponding with the patient who was a young adult. There is not sex predominance in general, but some studies show a female preponderance [12].

There are four histological types: embryonal, alveolar, pleomorphic, and spindle cell/sclerosing [13]. Among them, the embryonal type is the most common in head and neck region accounting for about 75% while the alveolar and pleomorphic are rare [14]. However, a study in children and young adults showed the commonest RMS arising from the paranasal sinus is the alveolar-type [15]. The diagnosis of embryonal-type rhabdomyosarcoma is confirmed by microscopic visualization of small round embryoblasts arranged in nests or cards separated by connective tissue trebaculae and nuclear-positive for myogenin. The International Classification of Pediatric Sarcomas further classifies these histologic subtypes based on prognosis, in poor prognosis category including the alveolar subtype, intermediate risk category including the embryonal subtype, and the superior prognosis group including the botryoid and spindle cell types [16]. Botryoid subtype is considered a special form of embryonal RMS [17].

Due to the fact that most of the tumor consists of solid nest cells of small rounded cells as in our case, tumors of the Ewing family, neuroblastoma, malignant rhabdoid tumor, leukemia, lymphoma, poorly differentiated carcinoma, melanoma and neuroendocrine carcinoma should be considered in the differential diagnosis [12].

It can occur in any anatomic location, and when occurring in the head and neck region three different primary sites have been recognized: parameningeal (involving the nasal cavity and paranasal sinuses, nasopharynx, middle ear, infratemporal fossa and pterigopalatine space), orbital and nonorbital/nonparameningeal [12]. Parameningeal sites
account for 41%–51.2% of head and neck RMS. It has been reported only few cases arising in the paranasal sinuses and orbit like our patient [10, 19]. Torres-Peña y cols presented two male adults (24 and 26 years respectively) with anterior cranial base alveolar RMS with orbital extension [20]. Those patients were treated with a combination of chemotherapy and radiation therapy following excisional biopsy by means craniofacial resection. The survival was 5 months and 2 years respectively due to systemic complications associated with the invasion.

Clinical manifestations result from tumor expansion into surrounding structures. Usually early symptoms of paranasal sinus Rhabdomyosarcoma are atypical due to its relatively concealed location and it commonly manifests with nasal obstruction, swelling of the maxillary region, purulent nasal discharge, epistaxis or headache which are consistent with presentation of the patient [6]. Many authors report ophthalmic presentations such as proptosis, epiphora, diplopia and retro-orbital pain like the patient was present [21].

Due to the absence of typically clinical manifestation and its low incidence, this tumor is frequently confused with other types of rapidly progressive malignant tumors of the head and neck, including nasopharyngeal carcinoma, lymphoma, primitive neuroectodermal tumors, Langerhans cell histiocytosis, olfactory neuroblastoma, osteosarcoma and metastasis [12].

CT and MRI are complementary studies indicated in these patients. CT scan in useful to evaluate any bone erosion. Lee et al. reported that ten cases of head and neck RMS appearing as isodense (100%) on pre-contrast CT and homogeneously enhanced (60%) on post-contrast CT scans similar to the patient CT scan findings [22]. On the other hand, MRI are helpful to distinguish the localization and limits of the lesion. Hagiwara et al. presented eight head and neck RMS with isointensity (37.5%) on T1, and heterogeneous hyperintensity (87.5%) on T2 [23]. Those founds are consistent with our patient and other authors [12]. It has been reported that tumors more than 5 cm in diameter have poorer prognosis > like this patient. Other evaluation includes bone scan and abdominal ultrasound in other to determine the staging. In this patient we did not found any signs of metastasis disease.

The Intergroup Rhabdomyosarcoma Study Group trials do not consider sinonasal disease an independent disease group, and there is otherwise very little literature that provides substantial evidence regarding the outcomes of surgical treatment of parameningeal [25]. Surgical modality was first described by Simon and Enneking in 1976 for soft tissue sarcomas and remains as the basis of sarcoma management [26]. A complete resection is achieved by resecting the tumor along with 0.5 cm margin of normal tissue around it [24].

Orbital and parameningeal RMS constitute a treatment challenge due its relationship to the skull base and orbit. A combination of radiation and multiagent chemotherapy has largely become the mainstay of treatment [17]. The functional morbidity and potential cosmetic associated with craniofacial resection has raised caution among authors, especially in children [6]. The Intergroup Rhabdomyosarcoma Study group proposed multimodal therapy, which involves surgery, chemotherapy, and radiotherapy [25]. Surgery is considered in localized, very early-stage disease or as salvage therapy in those who fail chemoradiation. Nevertheless, in the last two decades enormous advancements have been made in skull base and reconstructive surgery, permitting for more minimally invasive approaches to these difficult-to-access areas and more sophisticated reconstructions.

Endonasal endoscopic surgery, specifically, have been a good alternative to classical craniofacial surgery in non-advanced malignant anterior cranial base lesions and there are some reports for its in more advanced lesions [27, 28]. In spite of that, there are only few reports of parameningial and none orbital RMS treated by means endonasal endoscopic surgery, maybe due the fact of its low frequency [8].

The advantages of vascularized pedicle nasoseptal flaps during cranial base reconstruction after minimally invasive approach have been demonstrated, minimizing the risk of cerebrospinal fistulae [26, 28]. However, in patients with advanced malignant nasosinusal tumors the nasoseptal flap described for Haddad and Basabasteugey in 2006 isn’t viable due the septal invasion of the tumor that makes an ontologically safe surgery impossible [29]. A number of heterologous materials and tissue sealants have been used during reconstructions although there are expensive for development countries. In 2009 Zanation et al. described a minimally invasive harvested pericranial flap for anterior cranial base reconstruction [5]. Authors believe that this approach combines three important characteristics: (1) the effectiveness of pediculate and vascularized flaps, (2) their minimal costs and (3) their minimally cosmetic consequences. The patient of this report presented a good evolution without cerebrospinal fistulae or other complications demonstrating the effectiveness of this technique.

Radiotherapy is reserved for patients who develop recurrence following completion of initial treatment [30]. However, it should be considerate when residual tumor is suspected or in parameningial localizations due the relation with vital structures that limit the oncological safe removal. Combinations of cyclophosphamide, vincristine and actinomycin-D (VAC) are the accepted chemotherapeutic agents [7].

Poor prognostic factors include age > 10 years, meningeal involvement/intracranial extension, large tumor size, gross residual disease after treatment, and distant metastasis. A number of studies have showed that both age and alveolar subtype portend a worse prognosis for RMS overall the alveolar subtype, parameningeal involvement, and advanced stage of disease [3, 14-17]. Anyway, prognosis is generally poor due to the lack of early clinical symptoms as well as the proximity and extension into the meninges and skull base [17]. Therefore, earlier diagnosis is essential for the treatment of rhabdomyosarcoma and specific multimodal treatment should be motivated as early as possible. However, the embryonal subtype seems to have a better prognosis [7].

There are two points of interest to kept in mind in this case: first, discrepancies of a previous biopsy and definitive biopsy can be present and could conduce to a misdiagnosis and subsequent strategy error. Second, in such cases is preferable to make a most radical surgery as soon as possible, in order to assay the most oncological safe surgery possible.

Conclusion

Orbital parameningial RMS is a rare entity in adults but should be included in differential diagnosis. Endonasal endoscopic resection...
offers a faster recovery with minimal morbidity, providing a better life quality and immediately adjuvant treatment. Minimally invasive pericranial flap constitute a good alternative in cases with non-viable mucosal vascularized nasoseptal flap. Multimodality treatment, including chemoradiation and surgery, plays an important role in the management.

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