OPEN

PRS GLOBAL OPEN

ORIGINAL ARTICLE
Pediatric/Craniofacial

Nasal Embryonal Rhabdomyosarcoma in the Pediatric Population: Literature Review and Report of Midline Presentation

Jordan H. Larson, BS*
Rachel Rutledge, PA-C†
Laura Hunnell, PA-C‡
Daniel K. Choi, MD, MS§
Robert G. Kellogg, MD¶
Sanjay Naran, MD†‖**

Background: Congenital midline nasal masses are rare anomalies and are typically benign nasal dermoid sinus cysts (NDSCs). Rhabdomyosarcomas (RMSs) are even less common, and only a fraction affect sites like the external nose, nasal cavity, nasopharynx, and paranasal sinuses. We review the clinical presentation and treatment of nasal, nasopharyngeal, and paranasal RMSs and report the first documented midline presentation.

Methods: We queried PubMed for articles with titles containing the terms rhabdomyosarcoma or sarcoma botryoides and nose, nasal, paranasal, sinonasal, nasopharynx, or nasopharyngeal. We then searched the references of each included article using the same parameters and continued this process iteratively until no new articles were found.

Results: The paranasal sinuses were the most commonly affected site, followed by the nasopharynx, nasal cavity, and external nose. Two patients presented with involvement of the external nose, but each presented with involvement of the right ala rather than a midline mass. The rates of intracranial extension and/or skull base involvement were comparable to those of NDSCs. The alveolar subtype was most common, followed by the embryonal subtype.

Conclusions: Most midline nasal masses are benign; however, we report the first documented presentation of an RMS as a midline nasal mass. Accordingly, RMS should be included in the differential diagnosis of midline nasal masses in the pediatric population. Surgery for midline nasal masses is sometimes delayed due to the risks of interfering with developing structures and early anesthesia. However, early surgical treatment should be considered given this new differential and its predilection for early metastasis. *(Plast Reconstr Surg Glob Open 2021;9:e3534; doi: 10.1097/GOX.0000000000003534; Published online 20 April 2021.)*

INTRODUCTION

Congenital midline nasal masses are rare anomalies in children, with an incidence of 1 per 20,000–40,000 live births.[1–7] The differential diagnosis of such masses has traditionally included nasal dermoid sinus cyst (NDSC), encephalocele or meningoencephalocele, epidermoid cyst, sinus pericranii, hemangioma, and aberrant ethmoid sinus as well as acquired conditions like polyp, sebaceous cyst, mucocele, lipoma, fibroma, neurofibroma, adenoma, teratoma, osteoma, chondroma, ganglioneuroma, carcinoma, and Pott's puffy tumor.[1–6] Of these, NDSCs are the most common and account for as many as 61% of midline nasal masses in children.[2,4,5]

NDSCs typically present as a midline mass, pit, sinus tract, or fistula located anywhere from the glabella to the columella, most commonly along the dorsum.[1–7] They are generally firm, noncompressible, and nonpulsatile; do not transilluminate; and do not enlarge with crying or compression of the internal jugular vein (ie, they are negative for the Furstenberg sign).[1–4] Sinus openings with intermittent discharge of a "cheesy material" and hairs protruding through a cutaneous punctum are characteristic of an NDSC.[2–5,7] Infection is common and may lead to intracranial complications such as meningitis and

*From the *Chicago Medical School, Rosalind Franklin University of Medicine and Science, North Chicago, Ill.; †Advocate Children's Hospital, Division of Pediatric Plastic and Craniofacial Surgery, Park Ridge, Ill.; ‡Advocate Children's Hospital, Division of Pediatric Neurosurgery, Park Ridge, Ill.; §Advocate Children's Hospital, Division of Pediatric Hematology/Oncology, Park Ridge, Ill.; ¶Department of Neurological Surgery, University of Pittsburgh School of Medicine, Pittsburgh, Pa.; ‖Department of Plastic Surgery, University of Pittsburgh School of Medicine, Pittsburgh, Pa.; **Section of Plastic and Reconstructive Surgery, University of Chicago Medicine and Biological Sciences, Chicago, Ill.*

Received for publication January 7, 2021; accepted February 22, 2021.

The study was IRB exempt and conformed to the Declaration of Helsinki.

DOI: 10.1097/GOX.0000000000003534

Disclosure: *The authors have no financial interest to declare in relation to the content of this article.*

cerebral abscess in the presence of intracranial extension,[2–4,7] thereby warranting radiological evaluation and surgical excision of these masses. The frequency of intracranial extension varies, with reported rates as low as 4% and as high as 68%.[2,3,5–7]

Rhabdomyosarcomas (RMSs) are less common than NDSCs, with an estimated incidence of 4.4–4.5 per 1 million children.[8,9] However, they are the most common soft-tissue sarcomas among the pediatric population,[8,10–15] accounting for 5%–8% of pediatric solid tumors and 2.9% of all pediatric cancers in the United States.[9,11,13] Thirty to forty percent of RMSs affect the head and neck,[8–14] and 40%–50% of head and neck RMSs affect parameningeal sites such as the nasal cavity, nasopharynx, paranasal sinuses, middle ear, mastoid region, pterygopalatine fossa, and infratemporal fossa.[10,11,13] The involvement of parameningeal sites is clinically significant because, like NDSCs, tumors in these areas carry risk of involvement of the skull base and/or intracranial extension, which confers parameningeal RMSs a poorer prognosis.[8,9,12,14,15] Although RMSs are rare, and only a fraction affect parameningeal sites, RMSs are still the most common pediatric malignancies affecting the nose, nasopharynx, and paranasal sinuses.[9,15]

Four major histological subtypes of RMS have been identified: embryonal, alveolar, mixed, and pleomorphic.[11–13,15] There exist botryoid and spindle cell variants of the embryonal subtype,[11,13] and pleomorphic RMS may also be called undifferentiated or anaplastic.[11–13] Embryonal RMS is the most common histological subtype, particularly in younger children and in the head and neck.[10–15] Alveolar RMS is the second most common histological subtype in children but the most common subtype in adults, often associated with the t(1;13) and t(2;13) PAX-FOX01 translocations, and it may be the most common histological subtype in the sinonasal tract, specifically.[8,9,12,15,16] Like tumor location, histological subtype carries clinical significance, as the embryonal subtype is generally associated with a better prognosis than the alveolar subtype.[8,12,14,15]

We set out to review the current literature regarding the clinical presentation of RMSs of the nose, nasopharynx, and paranasal sinuses and establish their distribution, their histological subtypes, and the timing of their surgical treatment. In doing so, we demonstrate that an RMS has not previously been reported as a midline nasal mass, and in light of our report of the first documented midline presentation, we discuss the ideal timing for resection of a midline nasal mass in the pediatric patient.

METHODS

We conducted a review of the literature regarding RMS of the nose, nasopharynx, and paranasal sinuses. We began by querying PubMed for articles with titles containing the following search terms: rhabdomyosarcoma or sarcoma botryoides and nose, nasal, paranasal, sinonasal, nasopharynx, or nasopharyngeal. We included all English-language articles with full-text versions available online. We excluded articles that did not include at least 1 patient under 10 years old in order to focus on RMSs in young children and to avoid the data being confounded by RMSs in older children and adults, which may present differently. We chose 10 years of age as the upper limit because 0–10 years old is the most common age range in which RMSs present in the pediatric population. We then searched the reference list of each of the included articles using the same search terms, inclusion criteria, and exclusion criteria detailed above and included all articles that met these criteria. We continued this process iteratively until no new articles could be found.

RESULTS

Our literature review generated a list of 40 articles with dates of publication ranging from 1955 to 2020.[8–47] The list of articles and their patients' characteristics are included in Table 1.

These 40 articles included 877 patients who presented with RMS of the nose, nasopharynx, and/or paranasal sinuses. Because 3 of these articles did not present their own patients but rather queried the US National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) Incidence Database or the American College of Surgeons and American Cancer Society's National Cancer Database (NCDB),[9,12,15] some of the patients included in this article's dataset may have been counted more than once.

The included articles were heterogeneous with respect to their study types, which included case reports, case series, and database searches, as well as the types of data they reported. Many articles did not report certain categories of data listed in Table 1, such as the number of patients less than 10 years old and/or the involvement of certain sites. In particular, the database searches and larger case series, which did not include a full case write-up for each patient, were missing some of the types of data of interest. Because these types of articles accounted for the bulk of the 877 patients included in this article, some of the categories of data listed in Table 1 included information from only a small proportion of the total cohort of patients. Accordingly, for each of the categories of data listed in Table 1, we performed analyses only on the subset of patients for whom the relevant data were available rather than the total cohort of 877 patients.

Age data were available for 503 of the 877 total patients. Of these 503 patients, 225 (44.7%) were below the age of 10 years. Two of 175 patients, or 1.1% of the patients for whom the relevant data were available, presented with involvement of the external nose. However, each presented with involvement of the right nasal ala rather than a midline nasal mass.[18,24] Data regarding nasopharyngeal involvement were available for 397 patients, 166 (41.8%) of whom presented with such involvement. Data regarding involvement of the nasal cavity and/or paranasal sinuses were available for 799 patients. Of these patients, 244 (30.5%) presented with involvement of the nasal cavity and 516 (64.6 %) with involvement of the paranasal sinuses. Data were available regarding intracranial extension, skull base involvement, and both

intracranial extension and skull base involvement for 146, 138, and 69 patients, respectively. Twenty-three (15.6%) of 146 patients presented with intracranial extension, 38 of 138 (27.5%) with skull base involvement, and 7 of 69 (10.1%) with both intracranial extension and skull base involvement. These rates of intracranial extension and/or skull base involvement are comparable to those of NDSCs.

Finally, histological subtype was available for 843 patients. In accordance with the previous literature, the embryonal and alveolar subtypes predominated, accounting for 301 (35.7%) and 367 (43.5%) of cases, respectively. Although embryonal RMS is the most common histological subtype as reported in the literature, particularly in the head and neck and in younger children,[10–15] a number of studies have reported that alveolar RMS may be more common in the sinonasal tract specifically,[9,12,16] which is supported by these data.

CASE

Our patient was a 9-month-old boy referred to the Division of Pediatric Plastic and Craniofacial Surgery at Advocate Children's Hospital in July 2019 with a 2-month history of a slowly developing mass over the right dorsum of the nose. The parents denied any red spot at this location at birth and any subsequent drainage, rhinorrhea, fever, or chills. Physical examination revealed a 1-cm diameter subcutaneous, firm, slightly mobile, and nontender nodular mass over the right nasal dorsum that did not enlarge when the patient cried (Fig. 1). On suspicion of a nasal dermoid, the patient underwent an MRI with and without contrast 1 week later to evaluate for intracranial extension, which revealed a right para-midline extranasal mass with an attached fibrous stalk that extended intracranially through the foramen cecum. Restricted diffusion of the lesion suggested a nasal dermoid or neuroglial heterotopia (Fig. 2).

The patient was taken to the OR shortly after radiologic confirmation of intracranial extension (exactly 30 days following presentation), where a combined approach was performed by Pediatric Plastic Surgery and Pediatric Neurosurgery. An incision was made along the right nasal sidewall, and the extranasal mass was freed from the surrounding muscle and cartilage. Dissection was carried around the entire mass, down to the connected stalk. A coronal incision was then made, and a pedicled pericranial flap was harvested before a bifrontal craniotomy was performed. Following elevation of the bifrontal bone flap and retraction of the brain at the skull base, the intracranial stalk was identified. Dissection was carried around the stalk, and the stalk was tied off, cauterized, and cut, thus allowing for delivery of the nasal mass, with the stalk attached, through the nasal incision. The lining of the tract was debrided, and the tract was interrogated from both the skull base and the nasal approach until the instruments were found to meet. The tract was then packed with bone shavings, obliterating the connection between the brain and the nose, and a pericranial flap was inset over it, creating a seal between the tract and the intracranial space.

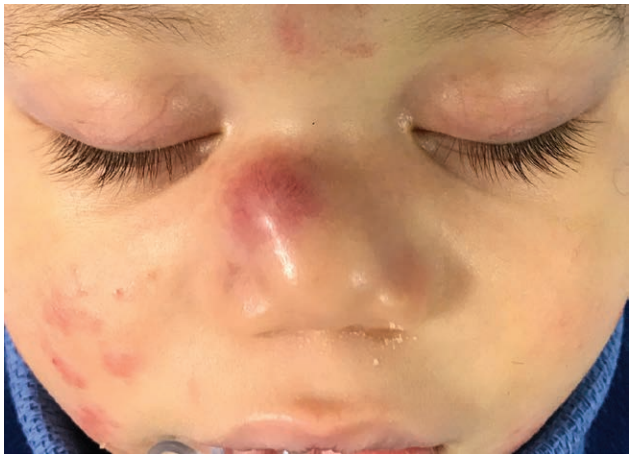

Fig. 1. Frontal view of a 1-cm diameter subcutaneous, erythematous, nodular mass over the right nasal dorsum.

The procedure was performed without complication, and the specimen was sent to pathology for evaluation. Immunohistochemical and genetic analyses determined the tumor to be an embryonal RMS. Following the discussion of the diagnosis with the parents, the patient was referred to Pediatric Oncology for further workup. Bilateral bone marrow aspirate and biopsy; lumbar puncture; CT with contrast of the head and neck and chest, abdomen, and pelvis; and whole-body PET/CT demonstrated no evidence of metastatic disease, and the patient was diagnosed with Stage II, Group IIA RMS, which was classified as low risk per the Children's Oncology Group (COG) Risk Stratification Criteria. Standard treatment for low-risk RMS would involve a combination of chemotherapy, surgical resection, and radiation. However, current COG guidelines for local therapy (ie, surgery and/or radiation) in children less than 24 months old allow for individualization of treatment to permit careful balancing of long-term morbidity against the increased risk of local failure and death.[48] Given the risk of late effects associated with radiation therapy to the nose and developing brain in such a young patient, the decision was made to treat the patient with only surgical resection and intensified chemotherapy per the intermediate-risk D9803 chemotherapy protocol, omitting radiation. Follow-up MRIs of the face with and without contrast at 3 and 6 months postsurgery revealed residual subtly enhancing soft tissue at the surgical site, which was biopsied to rule out residual tumor. Histopathologic analysis determined the tissue to be scar with no evidence of malignancy. At most recent follow-up, 11 months following initial diagnosis, the patient is doing well with no evidence of recurrence at the conclusion of chemotherapy.

DISCUSSION

Midline nasal masses are uncommon congenital malformations. When they do occur, they are typically benign NDSCs. Although head and neck masses in children may occasionally be RMSs, they are significantly less common than NDSCs. Moreover, pediatric head and neck RMSs most commonly affect parameningeal sites, including the nasal cavity, nasopharynx, and paranasal sinuses. They rarely present as masses involving the external nose. Our

Table 1. Literature Review Patient Characteristics

Study			Patients		Site (Nonexclusive)				
Title	Author	Year	n	Children < 10	External Nose	Nasal Cavity	Nasopharynx	Paranasal Sinuses	Nose NOS
Rhabdomyosarcoma of the nasopharynx	St. John EG, Woo Z-P	1955	1	1	0	1	1	0	0
Unusual nasal tumors in children; glioma and rhabdomyosarcoma	Crosby JF	1957	1	1	1	0	0	0	0
Sarcoma botryoides of the nasopharynx	Prior JT, Stoner LR	1957	1	1	0	1	1	0	0
Embryonic sarcoma (rhabdomyosarcoma) of the nasopharynx presenting with facial palsy	Holborow CA, White LL	1958	1	1	0	0	1	1	0
Rhabdomyosarcoma of the nasopharynx	Perkins HN, Stewart PD	1959	1	1	0	1	1	1	0
Radiosensitivity of malignant round-cell rhabdomyosarcoma in the nasal fossa of a child	Vaeth JM, Piatt TH	1961	1	1	0	1	1	1	0
Rhabdomyosarcoma of the nasopharynx	White A, Verma PL, Bullimore J	1974	1	1	0	1	1	1	0
Cure of an embryonal rhabdomyosarcoma of the nose of an infant by interstitial yttrium-90 microspheres: a case report	Ariel IM	1978	1	1	1	1	0	0	0
Nasopharyngeal rhabdomyosarcoma: a clinical perspective*	Canalis RF, Jenkins HA, Hemenway WG, Lincoln C	1978	21	18	0	0	21	3	0
Nasopharyngeal rhabdomyosarcoma: a clinical perspective*	Canalis RF, Jenkins HA, Hemenway WG, Lincoln C	1978	34	US	US	US	34	US	US
Rubinstein-Taybi syndrome and nasopharyngeal rhabdomyosarcoma	Sobel RA, Woerner S	1981	1	1	US	US	1	US	US
Rhabdomyosarcoma of the nasopharynx	Eavey R, Weber AL, Healy G	1982	1	1	0	1	1	0	0
Case report: nasopharyngeal rhabdomyosarcoma and Gorlin's naevoid basal cell carcinoma syndrome	Beddis IR, Mott MG, Bullimore J	1983	1	1	0	0	1	0	0
Alveolar rhabdomyosarcoma arising in the nasal cavity of a 3-year-old child	Pardo RJ, Acosta RE, Espaillat J, Penneys NS	1988	1	1	0	1	0	1	0
Rhabdomyosarcoma of the nasopharynx a case with recurrence of tumour after 20 years	Wight RG, Harris SC, Shortland JR, Shaw JD	1988	1	1	0	0	1	0	0
Nasopharyngeal rhabdomyosarcoma and multiple lentigines syndrome: a case report	Heney D, Lockwood L, Allibone EB, Bailey CC	1992	1	1	0	0	1	1	0
Rhabdomyosarcoma of the nose and paranasal sinuses in adults and children	Callender TA, Weber RS, Janjan N, et al.	1995	37	US	US	US	US	US	37
Rhabdomyosarcoma of nasopharynx	Das SK, Bhowmick A, Mukherjee S, Ghosh LM, Banerjee S	1999	1	1	0	1	1	1	0
Embryonal rhabdomyosarcoma of nasopharynx	Tuli BS, Parmar TL	1999	1	1	0	1	1	0	0
Nasopharyngeal rhabdomyosarcoma	Desarda KK, Gill SJ, Bora MP	2000	1	1	0	0	1	0	0
Botryoid rhabdomyosarcoma of the nose: pitfalls of pathology	Caversaccio M, Stauffer E	2001	1	1	0	1	0	1	0
Nasopharyngeal rhabdomyosarcoma in a patient with hypohidrotic ectodermal dysplasia syndrome	Cankaya H, Kösem M, Kiris M, Uner A, Metin A	2002	1	1	0	1	1	0	0
Pediatric sinonasal rhabdomyosarcoma: three cases and a review of the literature	Herrmann BW, Sotelo-Avila C, Eisenbeis JF	2003	3	2	0	2	1	1	0

(*Continued*)

Table 1. (*Continued*)

Cranial Involvement			Histological Subtype						
Intracranial Extension Alone	Skull Base Invasion Alone	Both IC and SB	Embryonal	Embryonal (Botryoid)	Embryonal (Spindle Cell)	Alveolar	Mixed	Pleomorphic	Unspecified
0	0	0	1	0	0	0	0	0	0
0	0	0	0	0	0	0	0	0	1
0	1	0	0	1	0	0	0	0	0
0	0	1	0	0	0	0	0	0	1
0	0	1	0	0	0	0	0	0	1
0	0	0	0	0	0	1	0	0	0
0	0	0	0	0	0	0	0	0	1
0	0	0	1	0	0	0	0	0	0
7	1	0	15	6	0	0	0	0	0
US	US	US	US	US	US	US	US	US	US
US	US	US	0	0	0	0	0	0	1
0	1	0	0	0	0	0	0	0	1
0	1	0	0	0	0	0	0	0	1
0	0	0	0	0	0	1	0	0	0
0	0	0	0	1	0	0	0	0	0
1	0	0	0	0	0	0	0	0	1
5	US	US	16	0	0	15	1	0	5
0	0	0	1	0	0	0	0	0	0
0	0	0	1	0	0	0	0	0	0
0	0	0	0	0	0	0	0	0	1
0	0	0	0	1	0	0	0	0	0
0	0	0	1	0	0	0	0	0	0
0	0	1	0	2	0	0	0	0	1

(*Continued*)

Table 1. (*Continued*)

Study			Patients		Site (Nonexclusive)				
Title	Author	Year	n	Children < 10	External Nose	Nasal Cavity	Nasopharynx	Paranasal Sinuses	Nose NOS
Long-term follow-up and prognosis of orbital apex syndrome resulting from nasopharyngeal rhabdomyosarcoma	Shindler KS, Liu GT, Womer RB	2005	3	3	0	0	3	0	0
Rhabdomyosarcomas of the nose and paranasal sinuses: treatment results in 15 cases	Wurm J, Constantinidis J, Grabenbauer GG, Iro H	2005	15	6	0	9	3	10	0
Rhabdomyosarcoma of the nasal vestibule in a child	Tanyous GH	2006	1	1	0	1	0	0	0
Sinonasal rhabdomyosarcoma in children and young adults	Ahmed AA, Tsokos M	2007	14	1	0	0	0	14	0
Management of paediatric sinonasal rhabdomyosarcoma	Fyrmpas G, Wurm J, Athanassiadou F, et al.	2009	14	8	0	7	5	11	0
Rhabdomyosarcoma of nose, nasopharynx and paranasal sinuses	Mondal PK, Pal I, Misra S, Biswas S, Bera SP	2009	6	2	0	4	1	6	0
Paediatric nasopharyngeal rhabdomyosarcoma: a case series and literature review	Healy JN, Borg MF	2010	5	4	US	US	5	US	US
Ophthalmic complications following treatment of paranasal sinus rhabdomyosarcoma in comparison to orbital disease	Gandhi PD, Fleming JC, Haik BG, Wilson MW	2011	17	US	0	7	7	10	0
Spinal cord glioblastoma induced by radiation therapy of nasopharyngeal rhabdomyosarcoma with MRI findings: case report	Ahn SJ, Kim I-O	2012	1	1	US	US	1	US	US
Incidence trends and long-term survival analysis of sinonasal rhabdomyosarcoma**	Sanghvi S, Misra P, Patel NR, Kalyoussef E, Baredes S, Eloy JA	2013	181	67	US	28	59	94	0
Sinonasal rhabdomyosarcoma: prognostic factors and treatment outcomes	Thompson CF, Kim BJ, Lai C, et al	2013	16	6	0	3	3	15	0
Pediatric sinonasal rhabdomyosarcoma: a case report	Bostanci A, Asik M, Turhan M	2015	1	1	0	1	1	1	0
Spontaneous internal jugular vein thrombosis in rhabdomyosarcoma of the nasopharynx	Walsh M, Meghji S	2016	1	1	0	0	1	0	0
A population-based analysis of survival for sinonasal rhabdomyosarcoma†	Unsal AA, Chung SY, Unsal AB, Baredes S, Eloy JA	2017	286	US	US	86	US	200	0
Therapeutic outcome and prognostic factors in sinonasal rhabdomyosarcoma: a single-institution case series‡	Li W, Lu H, Wang D	2019	40	5	0	30	2	33	0
Clinicopathologic traits and prognostic factors associated with pediatric sinonasal rhabdomyosarcoma**	Siddiqui SH, Siddiqui E, Bavier RD, et al	2019	157	75	US	50	US	107	0
Pediatric sinonasal rhabdomyosarcoma: clinical characteristics and surgical role	Al Momen A, Alshammari SM, Al Shakhs A, et al.	2020	4	4	0	4	3	3	0
Case report of nasopharyngeal rhabdomyosarcoma causing obstructive sleep apnoea	Love RL, MacKay SG	2020	1	1	0	0	1	0	0

(*Continued*)

Table 1. (*Continued*)

Cranial Involvement			Histological Subtype						
Intracranial Extension Alone	Skull Base Invasion Alone	Both IC and SB	Embryonal	Embryonal (Botryoid)	Embryonal (Spindle Cell)	Alveolar	Mixed	Pleomorphic	Unspecified
1	0	2	2	0	0	1	0	0	0
US	7	US	9	0	0	6	0	0	0
0	0	0	0	0	0	1	0	0	0
US	US	US	1	0	0	13	0	0	0
US	7	US	10	0	0	4	0	0	0
US	US	US	5	0	0	1	0	0	0
US	US	US	3	1	0	0	0	1	0
US	US	US	4	0	0	13	0	0	0
US	US	US	1	0	0	0	0	0	0
US	US	US	81	0	0	65	0	0	35
3	1	0	8	2	0	5	0	0	1
0	0	0	1	0	0	0	0	0	0
0	0	0	0	0	0	0	0	0	1
US	US	US	68	0	2	157	2	5	52
4	18	US	18	0	0	6	1	2	13
US	US	US	51	0	3	78	1	0	24
2	0	2	3	0	0	0	0	0	1
0	1	0	0	1	0	0	0	0	0

*Some articles included in this review were already populated within our query. As such, only the new patients included in this article were counted. The article contained 2 tables, 1 with detailed patient information and 1 without (ie, simply a list of patients with nasopharyngeal rhabdomyosarcoma). Due to the different natures of these 2 tables, each was counted on a separate row.
†These articles only included information on primary tumor location and not sites of tumor extension. As such, the values included in this table most likely underestimate the true counts of involved sites.
‡This article did not include detailed information on individual patients but rather aggregate counts of patients with maxillary sinus, ethmoid sinus, sphenoid sinus, and frontal sinus involvement (20, 26, 10, and 8, respectively), all of which were counted in this table as paranasal sinus involvement. Therefore, the true number of patients with paranasal sinus involvement could be anywhere from 26 to 40, so we chose the mean.
NOS, not otherwise specified; US, unspecified.

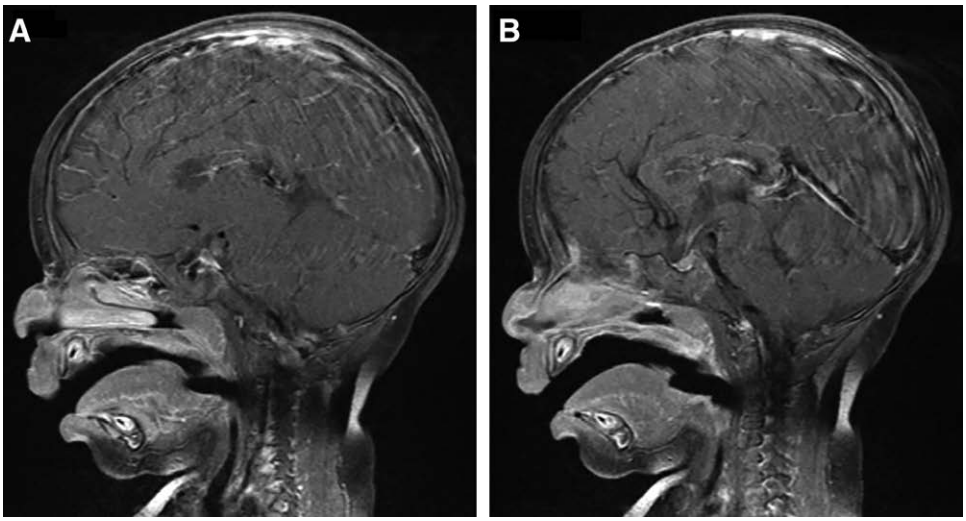

Fig. 2. Sagittal T1-weighted MRI; A, Demonstrating an 11-mm diameter mildly enhancing right paramidline mass at the base of the nose. The mass appears to be extranasal in location, superficial to the nasal bones, and there is restricted diffusion of the lesion; B, Demonstrating an enhancing fibrous stalk that extends from the mass superiorly and posteriorly through the foramen cecum into the intracranial compartment.

review of the literature revealed only 2 cases of RMS involving the external nose. Neither presented as a midline nasal mass. As such, the differential diagnosis of a midline nasal mass historically has not included RMS; however, we report the first documented presentation of an RMS as a midline nasal mass in a child. Given RMS's predilection for early metastasis,[11–13,16] its omission from the list of differential diagnoses for a midline nasal mass may no longer be appropriate and may have negative clinical implications.

There is no definitive consensus regarding the timing of surgery for midline nasal masses. Surgery for masses such as NDSCs is often recommended to be performed early to prevent deformity of the growing nose and reduce the risk of infection given the possibility of meningitis.[2,4,5,7,49] Others argue that the timing of surgery must be a compromise between excising the mass before the onset of complications and the risk of interfering with developing structures, therefore recommending that surgery be delayed until 2 years of age.[50] Still others recommend that surgery for midline nasal masses such as NDSCs is not necessary if they are asymptomatic and intracranial extension and malignancy have been ruled out.[1,6] Furthermore, beyond the risk of interfering with developing structures, the desire for early surgical intervention must also be balanced against the risks of pediatric anesthesia. As with any neonatal or infant procedure, it is important to appreciate that exposure to general anesthetics for lengthy periods of time or over multiple surgeries or procedures may negatively affect brain development in children younger than 3 years of age.[51] Infants less than 1 month old are also at significantly increased risk of perioperative adverse events, and infants between 1 month and 1 year of age are at increased risk of major postoperative events, particularly respiratory compromise.[52]

Given these considerations, we typically wait until after 1 year of age to resect suspected NDSCs without evidence of intracranial extension at our institution, performing surgery at 12–13 months of age. That being said, in reviewing the literature, we found that surgery for suspected NDSCs often takes place substantially later than 1 year of age. This is often the result of a delayed referral pattern. In a number of studies, the average or median age at the time of surgery for suspected NDSCs ranged from 19 months to 5.5 years.[3,5,49,50,53] The average reported delay from diagnosis to surgery is 0.9 years.[3] However, surgery for suspected NDSCs may be expedited if intracranial extension is apparent on imaging.[2] In retrospect, it is fortuitous that our patient's midline nasal mass presented with intracranial extension because had it not, we likely would have waited an additional 2 months (ie, until he were over 1 year of age) before resecting it. Such a delay may have put him at increased risk for metastasis before we had the opportunity to obtain a pathological diagnosis and refer him for definitive treatment with chemotherapy. Metastasis is a very poor prognostic factor in RMS, decreasing the survival rate from 39%–62% to 3%–24% in 2 of the larger, more recent database studies,[9,12] so it is of the utmost importance to catch and treat these diagnoses as early as possible. This experience has changed the management of nasal masses in patients under 1 year of age at our institution. Although the differential diagnosis of a midline nasal mass historically has not included RMS, given the case presented here, we now include RMS and recommend that RMS be included in the differential diagnosis of a midline nasal mass. We further argue that midline nasal masses even without intracranial extension be treated as aggressively and excised as promptly as those with intracranial extension. The benefit of definitively diagnosing a potentially aggressive RMS outweighs the risk of interfering with developing structures or earlier anesthesia.

Sanjay Naran, MD, FAAP
Division of Pediatric Plastic and Craniofacial Surgery
Advocate Children's Hospital
1675 Dempster Street, 3rd Floor
Park Ridge, IL 60068
E-mail: sanjaynaran@hotmail.com

PATIENT CONSENT

Parents or guardians provided written consent for the use of the patient's image.

REFERENCES

1. Hughes GB, Sharpino G, Hunt W, et al. Management of the congenital midline nasal mass: a review. *Head Neck Surg.* 1980;2:222–233.
2. Moses MA, Green BC, Cugno S, et al. The management of midline frontonasal dermoids: a review of 55 cases at a tertiary referral center and a protocol for treatment. *Plast Reconstr Surg.* 2015;135:187–196.
3. Winterton RI, Wilks DJ, Chumas PD, et al. Surgical correction of midline nasal dermoid sinus cysts. *J Craniofac Surg.* 2010;21:295–300.
4. Van Wyhe RD, Chamata ES, Hollier LH. Midline craniofacial masses in children. *Semin Plast Surg.* 2016;30:176–180.
5. Ni K, Li X, Zhao L, et al. Diagnosis and treatment of congenital nasal dermoid and sinus cysts in 11 infants: a consort compliant study. *Medicine (Baltimore).* 2020;99:e19435.
6. Volck AC, Suárez GA, Tasman AJ. Management of congenital midline nasofrontal masses: case report and review of literature. *Case Rep Otolaryngol.* 2015;2015:159647.
7. Rahbar R, Shah P, Mulliken JB, et al. The presentation and management of nasal dermoid: a 30-year experience. *Arch Otolaryngol Head Neck Surg.* 2003;129:464–471.
8. Thompson CF, Kim BJ, Lai C, et al. Sinonasal rhabdomyosarcoma: prognostic factors and treatment outcomes. *Int Forum Allergy Rhinol.* 2013;3:678–683.
9. Siddiqui SH, Siddiqui E, Bavier RD, et al. Clinicopathologic traits and prognostic factors associated with pediatric sinonasal rhabdomyosarcoma. *Int Forum Allergy Rhinol.* 2019;9:363–369.
10. Li W, Lu H, Wang D. Therapeutic outcome and prognostic factors in sinonasal rhabdomyosarcoma: a single-institution case series. *J Cancer Res Clin Oncol.* 2019;145:2793–2802.
11. Al Momen A, Alshammari SM, Al Shakhs A, et al. Pediatric sinonasal rhabdomyosarcoma: clinical characteristics and surgical role. *Int J Surg Case Rep.* 2020;2020:1–6.
12. Unsal AA, Chung SY, Unsal AB, et al. A population-based analysis of survival for sinonasal rhabdomyosarcoma. *Otolaryngol Head Neck Surg.* 2017;157:142–149.
13. Healy JN, Borg MF. Paediatric nasopharyngeal rhabdomyosarcoma: a case series and literature review. *J Med Imaging Radiat Oncol.* 2010;54:388–394.
14. Bostanci A, Asik M, Turhan M. Pediatric sinonasal rhabdomyosarcoma: a case report. *Exp Ther Med.* 2015;10:2444–2446.
15. Sanghvi S, Misra P, Patel NR, et al. Incidence trends and long-term survival analysis of sinonasal rhabdomyosarcoma. *Am J Otolaryngol.* 2013;34:682–689.
16. Ahmed AA, Tsokos M. Sinonasal rhabdomyosarcoma in children and young adults. *Int J Surg Pathol.* 2007;15:160–165.
17. St. John EG, Woo ZP. Rhabdomyosarcoma of the nasopharynx. *Radiology.* 1955;65:218–222.
18. Crosby JF. Unusual nasal tumors in children; glioma and rhabdomyosarcoma. *Plast Reconstr Surg 1946.* 1957;19:143–149.
19. Prior JT, Stoner LR. Sarcoma botryoides of the nasopharynx. *Cancer.* 1957;10:957–963.
20. Holborow CA, White LL. Embryonic sarcoma (rhabdomyosarcoma) of the nasopharynx presenting with facial palsy. *J Laryngol Otol.* 1958;72:157–165.
21. Perkins HN, Stewart PD. Rhabdomyosarcoma of the nasopharynx. *J Laryngol Otol.* 1959;73:612–618.
22. Vaeth JM, Piatt TH. Radiosensitivity of malignant roundcell rhabdomyosarcoma in the nasal fossa of a child. *Radiology.* 1961;77:94–96.
23. White A, Verma PL, Bullimore J. Rhabdomyosarcoma of the nasopharynx. *J Laryngol Otol.* 1974;88:271–276.
24. Ariel IM. Cure of an embryonal rhabdomyosarcoma of the nose of an infant by interstitial 90yttrium microspheres: a case report. *Int J Nucl Med Biol.* 1978;5:37–40.
25. Canalis RF, Jenkins HA, Hemenway WG, et al. Nasopharyngeal rhabdomyosarcoma: a clinical perspective. *Arch Otolaryngol.* 1978;104:122–126.
26. Sobel RA, Woerner S. Rubinstein-Taybi syndrome and nasopharyngeal rhabdomyosarcoma. *J Pediatr.* 1981;99:1000–1001.
27. Eavey R, Weber AL, Healy G. Rhabdomyosarcoma of the nasopharynx. *Ann Otol Rhinol Laryngol.* 1982;91(2 pt 1):230–231.
28. Beddis IR, Mott MG, Bullimore J. Case report: nasopharyngeal rhabdomyosarcoma and Gorlin's naevoid basal cell carcinoma syndrome. *Med Pediatr Oncol.* 1983;11:178–179.
29. Pardo RJ, Acosta RE, Espaillat J, et al. Alveolar rhabdomyosarcoma arising in the nasal cavity of a 3-year-old child. *Pediatr Dermatol.* 1988;5:254–256.
30. Wight RG, Harris SC, Shortland JR, et al. Rhabdomyosarcoma of the nasopharynx a case with recurrence of tumour after 20 years. *J Laryngol Otol.* 1988;102:1182–1184.
31. Heney D, Lockwood L, Allibone EB, et al. Nasopharyngeal rhabdomyosarcoma and multiple lentigines syndrome: a case report. *Med Pediatr Oncol.* 1992;20:227–228.
32. Callender TA, Weber RS, Janjan N, et al. Rhabdomyosarcoma of the nose and paranasal sinuses in adults and children. *Otolaryngol Head Neck Surg.* 1995;112:252–257.
33. Das SK, Bhowmick A, Mukherjee S, et al. Rhabdomyosarcoma of nasopharynx. *Indian J Otolaryngol Head Neck Surg.* 1999;51:90–93.
34. Tuli BS, Parmar TL. Embryonal rhabdomyosarcoma of nasopharynx. *Indian J Otolaryngol Head Neck Surg.* 1999;51:55–56.
35. Desarda KK, Gill SJ, Bora MP. Nasopharyngeal rhabdomyosarcoma. *Indian J Otolaryngol Head Neck Surg.* 2000;52:173–174.
36. Caversaccio M, Stauffer E. Botryoid rhabdomyosarcoma of the nose: pitfalls of pathology. *ORL J Otorhinolaryngol Relat Spec.* 2001;63:178–182.
37. Cankaya H, Kösem M, Kiris M, et al. Nasopharyngeal rhabdomyosarcoma in a patient with hypohidrotic ectodermal dysplasia syndrome. *Auris Nasus Larynx.* 2002;29:313–316.
38. Herrmann BW, Sotelo-Avila C, Eisenbeis JF. Pediatric sinonasal rhabdomyosarcoma: three cases and a review of the literature. *Am J Otolaryngol.* 2003;24:174–180.
39. Shindler KS, Liu GT, Womer RB. Long-term follow-up and prognosis of orbital apex syndrome resulting from nasopharyngeal rhabdomyosarcoma. *Am J Ophthalmol.* 2005;140:236–241.
40. Wurm J, Constantinidis J, Grabenbauer GG, et al. Rhabdomyosarcomas of the nose and paranasal sinuses: treatment results in 15 cases. *Otolaryngol Head Neck Surg.* 2005;133:42–50.
41. Tanyous GH. Rhabdomyosarcoma of the nasal vestibule in a child. *Sultan Qaboos Univ Med J.* 2006;6:87–89.
42. Fyrmpas G, Wurm J, Athanassiadou F, et al. Management of paediatric sinonasal rhabdomyosarcoma. *J Laryngol Otol.* 2009;123:990–996.
43. Mondal PK, Pal I, Misra S, et al. Rhabdomyosarcoma of nose, nasopharynx and paranasal sinuses. *Indian J Otolaryngol Head Neck Surg.* 2009;61:317–319.
44. Gandhi PD, Fleming JC, Haik BG, et al. Ophthalmic complications following treatment of paranasal sinus rhabdomyosarcoma in comparison to orbital disease. *Ophthalmic Plast Reconstr Surg.* 2011;27:241–246.
45. Ahn SJ, Kim IO. Spinal cord glioblastoma induced by radiation therapy of nasopharyngeal rhabdomyosarcoma with MRI findings: case report. *Korean J Radiol.* 2012;13:652–657.

46. Walsh M, Meghji S. Spontaneous internal jugular vein thrombosis in rhabdomyosarcoma of the nasopharynx. *BMJ Case Rep.* 2016;2016:bcr2015213455.
47. Love RL, MacKay SG. Case report of nasopharyngeal rhabdomyosarcoma causing obstructive sleep apnoea. *ANZ J Surg.* 2020;90:904–905.
48. Malempati S, Rodeberg DA, Donaldson SS, et al. Rhabdomyosarcoma in infants younger than 1 year: a report from the Children's Oncology Group. *Cancer.* 2011;117:3493–3501.
49. Bradley PJ. The complex nasal dermoid. *Head Neck Surg.* 1983;5:469–473.
50. Morgan DW, Evans JN. Developmental nasal anomalies. *J Laryngol Otol.* 1990;104:394–403.
51. U.S. Food & Drug Administration. FDA Drug Safety Communication: FDA approves label changes for use of general anesthetic and sedation drugs in young children. FDA. Available at https://www.fda.gov/drugs/drug-safety-and-availability/fda-drug-safety-communication-fda-approves-label-changes-use-general-anesthetic-and-sedation-drugs. 2017. Accessed September 8, 2020.
52. Cohen MM, Cameron CB, Duncan PG. Pediatric anesthesia morbidity and mortality in the perioperative period. *Anesth Analg.* 1990;70:160–167.
53. Cheng J, Kazahaya K. Management of pediatric nasal dermoids. *J Neurol Surg Part B Skull Base.* 2012;73:A156.