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CASE REPORT

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Ectopic rhabdoid meningioma of the orbit in a child: case report and review of the literature

Yağmur Seda Yeşiltaş, MD,[1] Kaan Gündüz, MD,[1] Aylin Okçu Heper, MD,[2] and Esra Erden, MD[2]

Departments of [1]Ophthalmology and [2]Pathology, Ankara University Faculty of Medicine, Ankara, Turkey

In August 2016, an 11-year-old boy presented to the authors' institution with a right orbital tumor that was located superotemporally (superolaterally) and adherent to the sclera. The patient's past medical history revealed that he had undergone 2 previous craniotomies elsewhere in June 2008 and July 2010 for a superomedially located orbital lesion that had been histopathologically diagnosed as a neurothekeoma. After the second craniotomy, the patient underwent adjuvant intensity modulated radiotherapy (IMRT) to the right medial orbit. At the authors' institution, total excision of the orbital tumor was performed via an anterior conjunctival orbitotomy. Histopathological examination revealed a rhabdoid meningioma. Review of the histopathology obtained at the time of previous tumor excisions showed that the lesion was misdiagnosed as neurothekeoma and instead represented a meningioma from the beginning. The patient was started on a regimen of oral sunitinib and remained free of recurrence at 1.5 years of follow-up. Ectopic meningioma of the orbit is a rare entity. Rhabdoid meningioma is a rarely seen subtype of meningioma, accounting for 1%–3% of all intracranial meningiomas. To the best of the authors' knowledge, this is the first case of an ectopic orbital rhabdoid meningioma reported in the literature. They suspect that tumor seeding during the previous surgeries might have played a role in the occurrence of the tumor in an orbital location not targeted by IMRT.

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KEYWORDS ectopic meningioma; immunohistochemistry; irradiation; magnetic resonance imaging; orbit; orbitotomy; pathology; pediatric; rhabdoid meningioma; sunitinib; surgery; oncology

The distribution and frequency of pediatric orbital tumors differ substantially from those seen in adults. About 80% of pediatric orbital tumors are benign lesions. The most common pediatric orbital tumor is dermoid cyst, accounting for 17%–46% of all orbital lesions, followed by capillary hemangioma (4%–12%) and inflammatory lesions (5%–8%).[3,26] The most common malignant orbital tumor is rhabdomyosarcoma, constituting 4%–7% of all orbital tumors in children.[26,42] In 2 series, orbital meningiomas accounted for 0.4%–1.4% of all pediatric orbital tumors.[26,42]

Meningiomas arise from arachnoid cells of the meninges and can occur either intracranially or extracranially. Extracranial or extradural meningiomas have been reported to occur in the orbit, scalp, nasal sinuses, nasopharynx, neck, and in distant sites, such as the skin, finger, lung, mediastinum, and adrenal gland. Extracranial meningiomas may arise from ectopic meningocytes, arachnoid cap cells, multipotential mesenchymal cells, or metaplasia of various mesenchymal cell types, including fibroblasts and Schwann cells.[27]

In the orbit, approximately two-thirds of orbital meningiomas extend secondarily from the meninges through bone of the skull base (secondary orbital meningioma); less than one-third arise primarily in the orbit from the optic nerve sheath (primary orbital meningioma). A rare subset of primary orbital meningiomas is the ectopic variety, which can arise in the orbit far from the optic nerve sheath or bone.[11]

Rhabdoid meningioma (RM) is an uncommon variant of meningioma. Initially described in 1998 by Kepes et al. and Perry et al., RM was classified as a separate entity for the first time in the 2000 WHO classification of tumors of the central nervous system.[22,38] Because it often exhibits malignant histological features and follows an aggressive clinical course, it has been classified as a grade III (anaplastic/malignant) neoplasm.[53]

Here, we report a case of ectopic orbital RM occurring

ABBREVIATIONS EMA = epithelial membrane antigen; IMRT = intensity modulated radiotherapy; RM = rhabdoid meningioma.

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in an 11-year-old boy and review previously published cases of intracranial RM and ectopic orbital meningioma.

Case Report

History and Examination

In August 2016, an 11-year-old boy presented to our institution with a 2-month history of proptosis of the right eye. He had a history of an orbital tumor managed elsewhere and had undergone craniotomy twice (June 2008 and July 2010), followed by intensity modulated radiotherapy (IMRT [5040 cGy]) to the right orbit 1 month after the second craniotomy. Review of the MRI studies from 2008 and 2010 revealed that the tumor was located superomedially (Fig. 1A and B). The tumor was diagnosed histopathologically as a neurothekeoma after both surgeries (Fig. 1C and D).

At presentation to our institution, the patient's visual acuity in the right eye was hand motions. There was a relative afferent pupillary defect in the right eye. The eyeball was displaced medially and inferiorly. The motility of the right eye was restricted in all positions of gaze, notably superiorly and laterally. There was 5 mm of proptosis in the right eye as determined on Hertel exophthalmometry. The intraocular pressure was 17 mm Hg. External examination and slit-lamp biomicroscopy showed that the tumor was located superotemporally (superolaterally), and it was adherent to the sclera (Fig. 2 left). On funduscopy, optic atrophy and an elevated choroidal mass with choroidal folds in the temporal quadrant were noted. The left eye had a visual acuity of 20/20, intraocular pressure of 16 mm Hg, and normal anterior segment and fundus findings.

CT scanning revealed a hyperdense, large oval mass with a cystic component. MRI demonstrated a well-circumscribed oval mass filling the superotemporal right orbit, producing scleral indentation of the right globe. On MRI, the tumor was isointense with respect to the extraocular muscles and cerebral gray matter on T1-weighted images and hyperintense on T2-weighted images (Fig. 2 right). The tumor showed moderate contrast enhancement.

Operation, Pathological Findings, and Postoperative Course

The patient underwent an anterior orbitotomy via a conjunctival approach, and the tumor was totally excised. No connections to optic nerve or bone were detected during surgery. The tumor was adherent to the sclera of the eyeball and to the surrounding orbital fat, and it appeared to have no connections to the periorbita. On gross examination, the tumor measured 2.8 × 2.5 × 1.5 cm and was reddish-purple (Fig. 3A). Histopathological examination disclosed that the tumor consisted of cells with large hyperchromatic, vesicular, eccentrically located nuclei and prominent nucleoli with mild atypia. The cells had eosinophilic cytoplasm, and intranuclear cytoplasmic pseudoinclusions were present (Fig. 3B). There were 11 mitoses per 10 hpf. Immunohistochemically, the tumor stained positively for epithelial membrane antigen (EMA), vimentin, p53, and INI-1 (Fig. 3C and D). The tumor cells showed negative immunoreactivity for desmin and actin. The Ki-67 labeling index was 20%. Based on the histopathological and immunohistochemical findings, a final diagnosis of RM was made. Five months after surgery, the patient was started on a regimen of sunitinib maleate (Sutent) 25 mg daily for 28 days in a 42-day cycle. Blood count and serum chemistry analyses were performed after each treatment cycle. Orbital and brain MRI and clinical evaluation were done at 3-month intervals to assess tumor response. At the 18-month follow-up, there was no clinical or MRI evidence of a recurrent tumor (Fig. 4), and visual acuity in the right eye was counting fingers at 3 m.

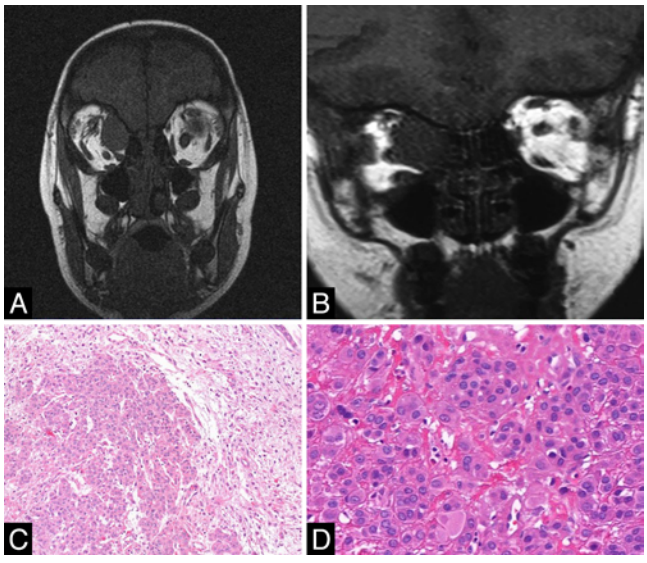


FIG. 1. A and B: Preoperative T1-weighted coronal orbital MR images obtained in 2008 (A) and 2010 (B) showing the superomedially located orbital tumor at initial presentation and at the time of first recurrence, respectively. The tumor is isointense with respect to the extraocular muscles, and the cerebral gray matter in both images. **C:** Photomicrograph obtained in 2008, showing a biphasic tumor composed of sheaths of spindle cells and groups/nodules of epithelioid round cells with eosinophilic cytoplasm. The findings were thought to represent neurothekeoma initially. H & E, original magnification ×10.3. **D:** Higher magnification of the epithelioid round cells. H & E, original magnification ×31.8. Figure is available in color online only.

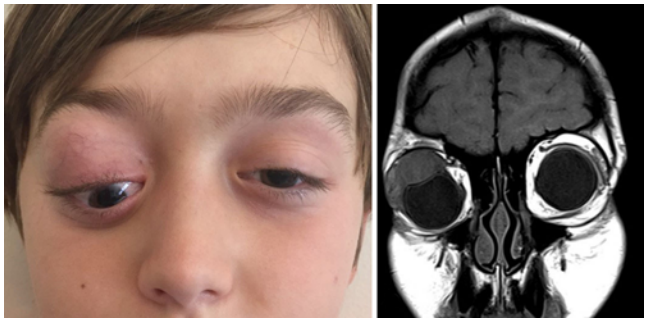


FIG. 2. Left: Preoperative facial photograph obtained at the time of presentation to our institution, showing the inferior displacement of the right globe and right proptosis. **Right:** Preoperative T1-weighted coronal orbital MR image revealing a well-defined orbital mass occupying the superotemporal orbit, producing scleral indentation of the right globe. The tumor is isointense with respect to the extraocular muscles and cerebral gray matter on T1-weighted images. Figure is available in color online only.

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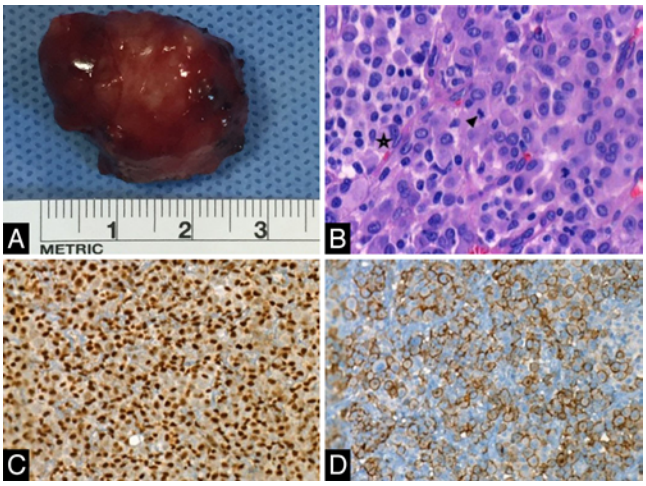


FIG. 3. A: Gross photograph of the excised tumor showing the reddish-purple orbital mass measuring 2.5 cm in largest base diameter. **B:** Photomicrograph showing that the tumor is composed of loosely arranged cells with rhabdoid morphology. The cells have peripheral eccentric nuclei, prominent nucleoli (*star*), and eosinophilic hyaline cytoplasm and display mitosis (*arrowhead*). H & E, original magnification ×61.7. **C:** The tumor cells show strong immunoreactivity for INI-1. Original magnification ×38. **D:** The tumor cells stain positively with EMA. Original magnification ×31.9. Figure is available in color online only.

Review of the histopathology done elsewhere after the previous craniotomies showed that the lesion was misdiagnosed as a neurothekeoma and that it was a meningioma from the beginning. The tumor showed a gradual transition to a more rhabdoid morphology with time. The MIB-1 index was graded as 10% for the first 2 surgical specimens. Review of the IMRT treatment plan disclosed that the medial orbit was irradiated and the lateral orbit was not targeted.

Methods of Literature Search

We performed a literature search of the PubMed database using the keyword combinations "rhabdoid", "meningioma", and "children"/"pediatric". We also performed a literature search using the keyword combinations "orbit", "ectopic", "meningioma" and "orbit", "extradural", "meningioma". We took into consideration only the articles published in the English language. Some of the articles were discarded because they did not fit into the description of RM, pediatric RM, and ectopic orbital meningioma, although they came up under the given keywords. The reference lists of these articles were checked for further references. We realize that we may have missed other cases of RM and ectopic orbital meningioma because the publishing journal or book is not indexed on PubMed and because of search strategy or keyword errors. Although our list may not be complete, we nevertheless made a significant effort to put together a reasonable number of well-documented ectopic orbital meningioma and RM cases to help us understand the presentation, clinical features, and treatment of these lesions better.



FIG. 4. Left: Postoperative facial photograph obtained at the 1.5-year follow-up, showing an absence of recurrent tumor and proptosis. **Right:** Postoperative T1-weighted coronal orbital MR image obtained at the 1.5-year follow-up, showing no residual tumor in the right orbit. Figure is available in color online only.

Discussion

Neurothekeoma (nerve sheath myxoma) is a rare, benign myxomatous neoplasm of the cutaneous terminal nerve sheath and most commonly occurs in the head, neck, and upper extremities.[28] Ophthalmic neurothekeomas are extremely rare neoplasms that have been reported to occur in the eyelids, orbit, and cornea.[36] Neurothekeoma is a benign lesion and has a low rate of recurrence with complete excision, regardless of location. Although our case was diagnosed histopathologically as neurothekeoma elsewhere, review of histopathology at our institution revealed that the lesion was a meningioma from the beginning with gradual transition to a more rhabdoid morphology.

Ectopic orbital meningioma is a rare entity. Our search of the PubMed database revealed only 23 cases of ectopic orbital meningioma that were previously reported in the literature.[2,7–9,11,14,19,29,31,39,44–46,48,52,54] The histopathological subtype was available for 21 patients. Thirteen (62.0%) of these 21 patients had meningothelial, 4 (19.0%) had fibroblastic, 2 (9.5%) had endotheliomatous, and 2 (9.5%) had angioblastic histopathological subtypes (Table 1). There has been no reported case of ectopic orbital meningioma with rhabdoid morphology to date.

RM represents a rare histopathological variant of meningioma.[30] The term "rhabdoid" refers to the resemblance of the tumor cells to a rhabdomyoblast without true skeletal muscle differentiation. Perry et al. identified 8 cases (0.37%) of 2161 intracranial meningiomas with a rhabdoid morphology over a period of 27 years.[38] Ko et al. reported that the prevalence of RM was 3.3% in a series of 485 intracranial meningiomas.[25] Our review of 54 publications in the English-language literature detailing 210 cases of intracranial RM showed that the tumor occurs in patients ranging from 2 to 84 years old, with a mean age of 44 years. In 14 of 54 articles, the affected patients were in the pediatric age group (< 18 years old). Together, with 3 more cases published in non-PubMed journals, we were able to retrieve a total of 17 pediatric cases (Table 2).[4–6,10,13,15–18,32,35,37,38,40,41,49,51] The duration of symptoms before the diagnosis of RM was reported to be less than half a year.[23]

Intracranial RMs are usually isointense to gray matter on T1-weighted images and hyperintense on T2-weighted images. Contrast enhancement is seen in the majority of lesions. Another MRI feature is the presence of a cystic

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TABLE 1. Patient demographics and histopathological features in 24 cases with ectopic orbital meningioma

Authors & Year	Age (yrs), Sex	Orbital Location	Surgery	Extent of Resection	Histopathology	Recurrent/ Residual Tumor	Secondary Treatment	FU (mos)
Craig & Gogela, 1949	54, M	Superomedial	Craniotomy	Subtotal excision	Fibroblastic	Residual	None	7
D'Alena, 1964	29, M	Inferior	Orbitotomy	Subtotal excision	Fibroblastic	None	Exenteration+EBRT	96
Tan & Lim, 1965	9, F	Medial	Orbitotomy	Excision*	Meningothelial	NA	None	NA
Macmichael & Cullen, 1969	20, F	Lateral	Orbitotomy	Excision*	Meningothelial†	None	None	3
Takahashi et al., 1973	37, F	Superomedial	Craniotomy	Excision*	Angioblastic	NA	None	NA
	34, F	Inferior	Orbitotomy	Excision*	Angioblastic	NA	None	NA
	48, F	Lateral	Craniotomy	Total excision	Endotheliomatous	NA	None	NA
	55, F	Superior	Craniotomy	Subtotal excision	Endotheliomatous	NA	None	NA
Wolter & Benz, 1976	33, M	Superomedial	Orbitotomy	Excision*	Meningothelial	None	None	3
Lloyd, 1982	19, F	Medial	NA	NA	NA	NA	NA	NA
Johnson et al., 1993	10, M	Superomedial	Orbitotomy	Subtotal excision	Meningothelial	Residual	Craniotomy	12
	7, M	Superomedial	Orbitotomy	Subtotal excision	Meningothelial	Residual	External ethmoid-ectomy	4
Spraul et al., 1996	30, M	Medial	Orbitotomy	Total excision	Fibroblastic	None	None	18
Arai et al., 1997	27, F	Superomedial	Craniotomy	Total excision	Fibroblastic	None	None	1
Farah et al., 1999	53, M	Medial	Orbitotomy	Total excision	Meningothelial	None	None	42
Yokoyama et al., 1999	7, M	Inferomedial	Craniotomy	Total excision	Meningothelial	None	None	NA
Decock et al., 2009	66, M	Lateral	Orbitotomy	Excision*	Meningothelial	None	None	15
Pushker et al., 2013	30, F	Superior	Orbitotomy	Excision*	Meningothelial	Present	None	18
	40, M	Superomedial	Orbitotomy	Excision*	Meningothelial	Present	None	24
	9, M	Superomedial	Orbitotomy	Excision*	Meningothelial	None	None	3
Gündüz et al., 2014	56, F	Superior	Orbitotomy	Subtotal excision	Meningothelial	Residual	EBRT	74
	27, M	Lateral	Orbitotomy	Subtotal excision	Meningothelial	Residual	EBRT	24
Tendler et al., 2017	9, F	Superomedial	Orbitotomy	Excision*	NA	Present	Surgical debulking+PBRT	6
Present case	11, M	Superotemporal	Orbitotomy	Total excision	Rhabdoid	Present	Sunitinib	111

EBRT = external-beam radiotherapy; FU = follow-up; NA = not available; PBRT = proton-beam radiotherapy.

* Data not available as to whether subtotal or total excision was achieved.

† Diagnosis made based on the histopathological findings.

component within the lesion.[12] Our case of orbital RM had similar findings. Differential diagnosis of orbital RM based on clinical and neuroimaging findings may include dermoid cyst, rhabdomyosarcoma, simple epithelial cyst, mucocele, adenoid cystic carcinoma, lymphangioma, and many other entities.[43]

Histopathologically, RMs usually exhibit signs of anaplasia, high mitotic activity, and markedly increased MIB-1 (Ki-67) index, a marker of cell proliferation.[47] In general, an MIB-1 labeling index more than 10% shows malignant meningeal tumors. In our case, there was a significant increase of MIB-1-positive cells from the first and second specimens (10%) to the third specimen (20%), suggesting an aggressive transition. Microscopically, RMs have the typical cytological feature of sheets of rhabdoid cells, which often present as plump cells with eccentric nuclei and prominent nucleoli. The cells have abundant eosinophilic cytoplasm and paranuclear intracytoplasmic hyaline inclusions.[30] Ultrastructural examination demonstrates the presence of interdigitating cell processes joined by numerous desmosomes and paranuclear whorls of intermediate filaments typical of the rhabdoid phenotype.[33] On immunohistochemical staining, the tumor cells are abundantly vimentin positive.[24] EMA is variably positive in conventional meningiomas and RMs.[22,38] Staining with GFAP, homatropine methylbromide 45 (HMB 45), and muscle markers, including desmin and S100 protein, is generally negative in RMs, although cases with partial positive staining have also been reported.[32] The histopathological and immunohistochemical findings in our case were in accordance with the previously reported results. Our case further demonstrated positive staining with p53 protein. Histopathological differential diagnosis of RM includes atypical rhabdoid–teratoid tumors, metastatic melanomas, germ cell tumors, and rhabdomyosarcomas.[32]

It is well documented that tumors with rhabdoid morphology have an aggressive course. However, the outcome in pediatric patients with RMs seems to be better than that of their adult counterparts, with a mortality rate of 12.5% at 84 months compared with a mortality rate of 42.9% at 12 months in adults.[32] Nevertheless, the small number of reported pediatric RMs available in the literature makes

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TABLE 2. Review of reported pediatric cases of RM

Authors & Year	Age (yrs), Sex	Surgery	Histopathology	Recurrence	Secondary Treatment	FU (mos)	Outcome*
Perry et al., 1998	13, F	NA	Rhabdoid papillary	Present	None	204	Alive
Rittierodt et al., 2001	9, M	Excision†	Rhabdoid	Present	None	96	Alive
Hojo & Abe, 2001	15, M	Subtotal excision	Rhabdoid papillary	Present	EBRT	72	Alive
Huang et al., 2002	13, F	Total excision	Rhabdoid	NA	NA	NA	NA
Wakabayashi et al., 2005	12, F	Subtotal excision	Rhabdoid papillary	Present	EBRT	156	Dead
Nozza et al., 2005	12, F	Subtotal excision	Rhabdoid	None	EBRT+chemo	20	Alive
Gu et al., 2005	15, M	Subtotal excision	Rhabdoid	Present	NA	4	Alive
Martínez-Lage et al., 2006	14, F	Total excision	Rhabdoid	None	EBRT	6	Alive
Jing et al., 2006	5, F	NA	Rhabdoid	NA	NA	NA	NA
Cai et al., 2008	2, F	Subtotal excision	Rhabdoid	None	None	30	Alive
Chaturvedi et al., 2008	13, F	Subtotal excision	Rhabdoid	None	EBRT	6	Alive
Morina et al., 2010	8, F	Total excision	Rhabdoid	None	EBRT	18	Alive
Buccoliero et al., 2011	3, M	Subtotal excision	Rhabdoid	None	Chemo	33	Alive
El Jamal et al., 2011	16, F	Subtotal excision	Rhabdoid	None	EBRT	14	Alive
Jia et al., 2014	11, F	Total excision	Rhabdoid papillary	None	EBRT	7	Alive
Tian et al., 2014	9, M	Subtotal excision	Rhabdoid	Present	EBRT	12	Dead
Reddy et al., 2015	6, F	Subtotal excision	Rhabdoid	Present	EBRT	21	Alive
Present case	11, M	Total excision	Rhabdoid	Present	Sunitinib	111	Alive

Chemo = systemic intravenous chemotherapy.

* Alive indicates complete cure or stable with disease.

† Data were not available as to whether subtotal or total excision was achieved.

conclusive prognostic remarks difficult. Vaubel et al. suggested that some RMs lack features of malignancy (WHO grade I or II) and may not behave as aggressively as RMs classified as WHO grade III.[50] One interesting feature of RM is that in a significant number of cases, the rhabdoid cells are observed predominantly at the time of recurrence, as in our case.[33] This finding suggests that transition from a focal rhabdoid phenotype to a predominantly rhabdoid one can occur, usually associated with recurrence and local aggressive behavior.[22,38]

Despite extensive resection and radiotherapy, 41.4% of intracranial RMs recur and require additional treatment.[55] In the current case, the tumor was initially located in the superonasal orbit but occurred in a superotemporal location at the time of presentation to us. Incomplete tumor excision, tumor seeding, and/or lack of coverage of the temporal orbit by the previous IMRT might have played a role in the occurrence of the tumor at a different site in our case.

Several different treatments other than surgery have been tried for intracranial meningiomas, including traditional chemotherapeutic options, hormonal therapies, interferon alfa-2b, and molecularly targeted therapies, but all have been disappointing.[21] In our case, despite the fact that total tumor resection was achieved, sunitinib maleate was initiated as adjuvant treatment to prevent recurrence. Sunitinib maleate is an orally administered, small-molecule multiple tyrosine kinase inhibitor that targets platelet-derived growth factor receptor, vascular endothelial growth factor receptor, FMS-like tyrosine kinase, tyrosine protein kinase KIT, macrophage colony stimulating factor 1 receptor, and RET proto-oncogene, most of which are abundantly expressed in meningiomas.[21,34] It has antiangiogenic, cytostatic, and antimigratory activity for meningioma tumor cells.[1,21,34] Inhibiting these targets represents an attractive therapeutic approach for meningiomas. In particular, previously irradiated meningioma cells are more sensitive to adjuvant sunitinib therapy.[1,20,21]

In summary, we present an interesting case of ectopic orbital RM occurring in an 11-year-old boy. The patient had undergone 2 previous craniotomies and IMRT for a superomedially located orbital lesion that had been diagnosed histopathologically as neurothekeoma at an outside institution. At presentation to us about 6 years after the second craniotomy and IMRT, the lesion was located in the superotemporal orbit. The tumor was totally excised and diagnosed histopathologically as RM. Review of histopathology showed that the initial diagnosis was incorrect and the tumor represented a meningioma from the beginning, demonstrating a gradual transformation into a more rhabdoid morphology. We speculate that tumor seeding or inadequate coverage of the lateral orbit by the previous IMRT might have played a role in the occurrence of the tumor at a different orbital site. This case is interesting because ectopic orbital meningioma is an extremely rare tumor in adults as well as in children and rhabdoid morphology in an ectopic orbital meningioma has never been reported before in any age group.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Gündüz, Yeşiltaş. Acquisition of data: all authors. Analysis and interpretation of data: Gündüz, Yeşiltaş. Drafting the article: Gündüz, Yeşiltaş. Critically revising the article: Gündüz, Yeşiltaş. Reviewed submitted version of manuscript: Gündüz, Yeşiltaş. Approved the final version of the manuscript on behalf of all authors: Gündüz. Study supervision: Gündüz.

Correspondence

Kaan Gündüz: Ankara University Faculty of Medicine, Ankara, Turkey. drkaangunduz@gmail.com.

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