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Case report

Orbital hemangioma extirpation via a transnasal endoscopic surgical approach: A case report

Ratih Finisanti [a,1,*], Muhammad Fadjar Perkasa [a,1], Adi Matra Prawira [a], Muhammad Faruk [b]

[a] Department of Otorhinolaryngology - Head and Neck Surgery, Faculty of Medicine, Hasanuddin University, Makassar, Indonesia
[b] Faculty of Medicine, Hasanuddin University, Makassar, Indonesia

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ABSTRACT

Introduction: Orbital tumors are heterogeneous lesions originating from various structures in the eyeball, including the extraconal, conal, and intraconal spaces. One orbital tumor type is a vascular tumor, such as a hemangioma. Hemangiomas are most common in women aged 20–64 years. They are painless and slow-growing, leading to proptosis. The diagnosis can generally be established by clinical examination and computed tomography (CT) or magnetic resonance imaging scans. Orbital hemangioma management can involve regular observation of small and asymptomatic tumors or surgery for large symptomatic tumors.

Presentation of case: We report a 20-year-old Asian woman who presented with swelling on the medial side of her right eyelid that had increased over the last three months. A non-contrast head CT scan showed a mass in the right eye's medial wall. The patient underwent a joint operation with an ophthalmologist to remove the median orbital tumor by transnasal endoscopic orbital surgery. Anatomical pathology examination of the tumor tissue identified hemangiomas.

Conclusion: An endoscopic transnasal approach is a safe and effective way to access and manage an orbital tumor medial to the optic nerve. It is essential to have a multidisciplinary team with experience in endoscopic procedures. This patient had satisfactory results at their three-month postoperative follow-up. They reported no symptoms, and their CT scan did not show a relapsed mass in the right oculi region.

1. Introduction

Orbital tumors are heterogeneous lesions originating from various structures within the orbits [1]. These tumors are divided into primary and secondary lesions as extensions to the orbit from other structures [1,2]. Based on its origin and histology, one orbital tumor type is hemangioma [1]. Orbital hemangiomas are more common in women than men [3]. Nearly 70 % of cases occur in middle age, between 20 and 60 years, and especially in women aged around 40 [4]. The most common intraorbital primary tumors in adults are cavernous hemangiomas, accounting for 4 % of all orbital tumors and 9 %–13 % of all intracranial cavernous hemangiomas [5]. A hemangioma diagnosis can generally be made via preoperative computerized tomography (CT) or magnetic resonance imaging scans. A CT scan can show intraconal and extraconal masses that are round and well-defined [6].

Endoscopic endonasal surgery was first used to treat inflammatory sinonasal disease in the late 1980s. Advances in technology have enabled better visualization, safety, and surgical efficacy. Early reports of applying the endoscopic endonasal approach to lesions of the skull base and orbital apex were restricted to diagnostic biopsies rather than complete excisions. Articles by Herman and by Mir-Salim and Berghaus described the first cases of endonasal endoscopic orbital tumor excision and endonasal microsurgery in 1999 [7].

Currently, the posterior orbital masses located medial and inferior to the optic nerve can be safely accessed endoscopically. This approach can still be developed for use with lesions located lateral to the optic nerve [5,8,9]. Here, we report a hemangioma case successfully treated with functional endoscopic sinus surgery (FESS) using a uninarial approach according to the surgical case report guidelines [10].

* Corresponding author at: Department of Otorhinolaryngology - Head and Neck Surgery, Faculty of Medicine, Hasanuddin University, Jalan Perintis Kemerdekaan KM 11, Makassar, Sulawesi Selatan 90245, Indonesia.
E-mail addresses: ratih.finisanti@gmail.com (R. Finisanti), perkasa715@gmail.com (M.F. Perkasa), matradi88@gmail.com (A.M. Prawira), muhammadfaruk@unhas.ac.id (M. Faruk).
[1] These authors contributed equally to this work.

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2. Presentation of case

The ophthalmology department consulted a 20-year-old Asian woman with the chief complaint of swelling in the right eyelid that increasing over the last three months. The patient had a history of bloody tears two weeks before attending the hospital and felt pain when moving the eyeball medially. The patient had no history of decreased vision in the past three months.

Physical examination showed mild swelling in the right eye with limited eye movement (Fig. 1). On the cornea and scleras, the best-corrected visual activity (BCVA) of the right and left eyes was 10/10. Anterior rhinoscopy, otoscopy, and pharyngoscopy did not show any abnormalities.

A non-contrast head CT scan (Fig. 2) showed a mass in the right orbital medial wall with mild prolapse. Laboratory tests and chest radiographs were within normal limits. We planned joint surgery with an ophthalmologist to remove the tumor.

The patient underwent FESS, including an uncinectomy, middle meatal antrostomy, and anterior and posterior ethmoidectomy. Then, the right lamina papyracea was resected, followed by an incision in the orbital septum. The tumor mass was reddish with an uneven surface and soft consistency originating from the right median orbit. We precisely and bluntly released the tumor from the attachment (Fig. 3). Anatomical pathology examination of the tumor tissue determined it was a hemangioma (Fig. 4).

Postoperatively, the patient received analgesics, broad-spectrum antibiotics, and 120 mg of propranolol daily. Patients are followed-up in the Outpatient Department in the first and third postoperative months. The patient's results showed no abnormalities in both eyes with BCVA 10/10 and movements within normal limits, with no ptosis or other complications. Head CT scans with contrast at one and three months postoperatively showed no residual mass in the inferomedial part of the wall and no signs of recurrence.

3. Discussion

Patients often present with proptosis due to a slowly growing orbital mass, which accounts for ~70 % of cases [4,11,12]. Exophthalmos is typically progressive, depending on the anatomical location, and may or may not be axile, painless, and non-pulsatile until a complication occurs (e.g., thrombosis, hemorrhage, or inflammation). Discovery is sometimes accidental. Hyperopia is caused by intra-conical tumors compressing the globe's posterior surface. Compression of the optic nerve is rare and causes papillary edema or even choroidal folds in the fundus, reducing visual acuity [12].

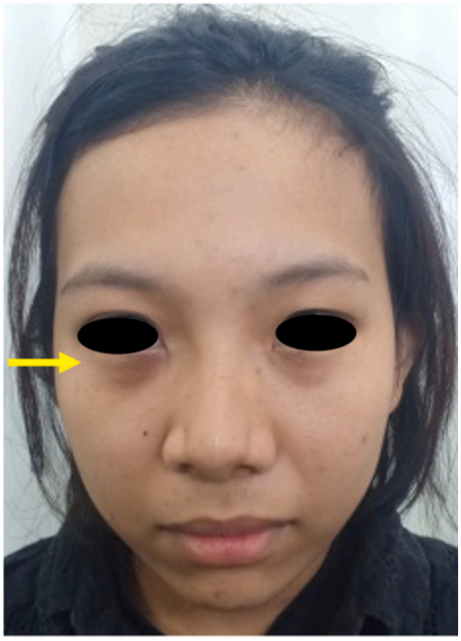

Fig. 1. The patient's ocular appearance. A mild prolapse (yellow arrow, front view) was observed on their lower right eyelid.

Imaging plays a significant role in diagnosis. The cavernous hemangioma is a well-defined, hyper-echogenic, and homogeneous mass on ultrasonography (US) [12]. Color Doppler US shows significant vascular spaces with low flow [6,12,13]. Hemangiomas appear as a well-defined, homogenous mass with a slightly more hyperdense appearance than the ophthalmic muscles on a CT scan. Small calcifications or bone remodeling can occur. Mild contrast enhancement is a common proptosis feature because of its low vascular flow [11]. Magnetic resonance imaging should be used to evaluate the potential compressive impact, particularly on the optic nerve. The lesion is well-defined and oval-shaped. The diagnosis is highly suggested when a typical cavernous hemangioma of the orbit presents as a well-defined homogenous mass with isointensity or mild hypo intensity in T1-weighted sequences and greater intensity than muscles in T2-weighted sequences [12].

Histologically, orbital cavernous hemangiomas comprise numerous large vascular spaces surrounded by stroma and endothelial cells, separated by fibrous septa, and covered with a fibrous pseudocapsule. They lack a main arterial supply. Many of these lesions' lumens have intralesional thrombosis due to their typical slow vascular flow and stasis [4].

A cavernous hemangioma's vessel walls will stain positively for classic vascular endothelium markers (cluster of differentiation 31 [CD31] and 34 [CD34]) and negatively for a lymphatic endothelial marker (podoplanin [D2–40]). Additionally, in cavernous hemangiomas, glucose transporter 1 (GLUT-1) immunostaining is negative, and marker of proliferation *Ki-67* expression is either low or negative [4,14].

A panel of 23 international multidisciplinary teams in endonasal skull-base surgery and endonasal orbital tumor developed a modified Delphi method. They created the Cavernous Hemangioma Exclusively Endonasal Resection (CHEER) stage system (Table 1) [5], which is used to more accurately identify the location and stage of orbital hemangiomas that may be amenable to endonasal endoscopic surgical excision. This staging method categorizes orbital hemangiomas into seven different anatomical locations or stages based on the lesion's intraconal or extraconal location, relationship to the ocular artery's muscular trunk, and vertical interaction with the medial rectus muscle. This study indicated that the panel preferred a binarial and two-surgeon strategy over an uninarial and one-surgeon approach with progressing lesion stage [5].

Surgery and observation are the two main methods used to treat orbital cavernous hemangiomas. Sclerotherapy, pingyangmycin intralesional therapy, and stereotactic radiosurgery are alternative treatment options that may be used in uncommon and exceptional cases. Goals and indications for treatment vary on the specific modality. Surgical therapy is required in cases where lesions cause severe symptoms such as optic nerve compression, disfiguring cosmesis, and extraocular gaze restriction [4]. Intraconal lesions inferior and medial to the optic nerve are recommended for the transnasal endoscopic technique, notably cavernous hemangiomas, which can be easily handled with minimal rupture risk, making them excellent for transnasal management [5].

A maxillary antrostomy is typically performed after a sphenoethmoidectomy to provide access to the orbital floor using a 0° optic and an 18 cm rigid endoscope. The floor of the orbit and the bony medial portion of the lamina papyracea are then located and excised using a 45° optic. The periorbita is precisely opened with a sickle knife and endoscopic microscissors after being carefully separated from the underlying bone. The tumor becomes visible and is removed after being separated from the periorbital fat. The dissection corridor for intraconal lesions that are medially and inferiorly positioned is between the medial and inferior rectus muscles. They are identified and isolated using a vessel

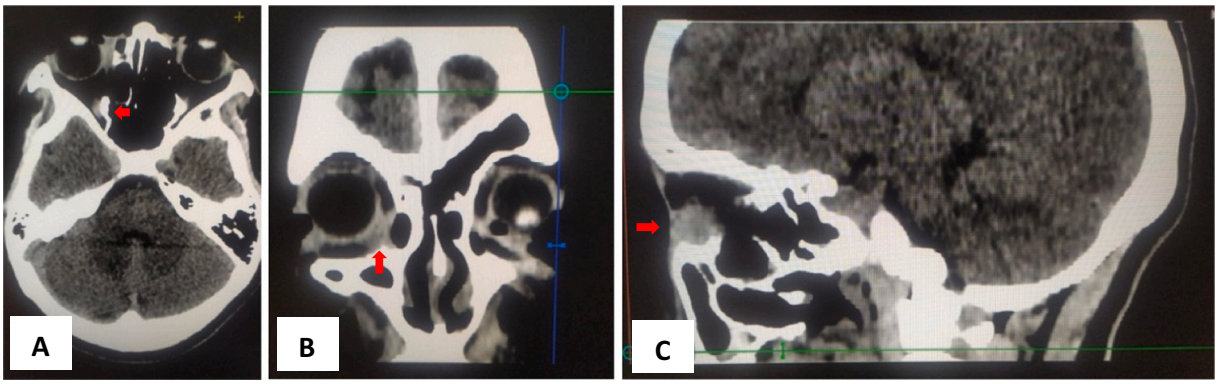

Fig. 2. Non-contrast head CT scan. The (A) axial, (B) coronal, and (C) sagittal views showed a mass on the inferomedial wall of the right orbit (red arrow).

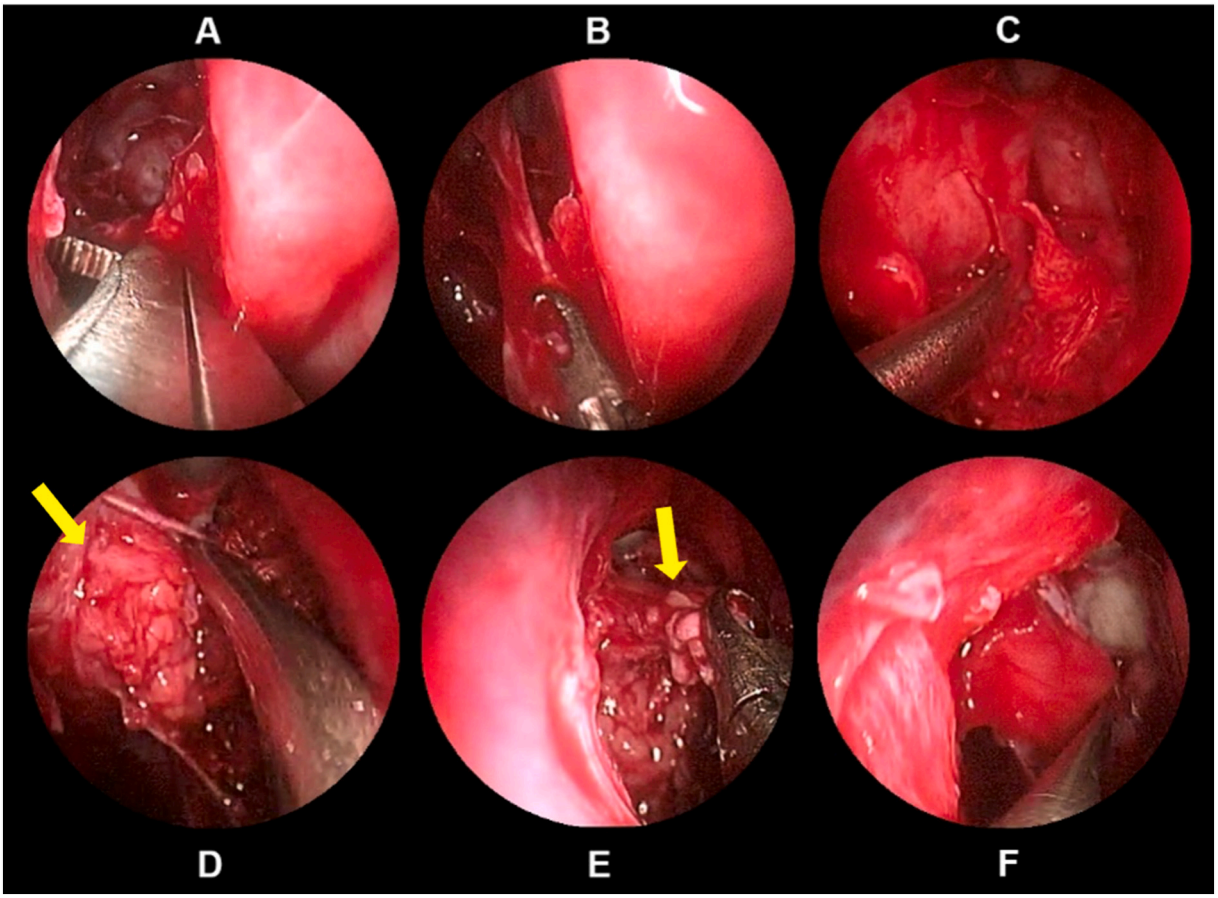

Fig. 3. Transethmoid endoscopic surgery. (A, B) FESS procedures. (C) Right and deepened lamina papyracea resection for tumor visualization. (D) Tumor detachment from surrounding tissue (arrow). (E) Debulking tumor (arrow). (F) Administration of the hemostatic gelatin sponge on surgical wound defects.

loop as they insert on the globe before retraction. A limited bipolar cautery and thorough sharp dissection are used to locate and remove the tumor once the intraconal corridor has been developed [5].

Choosing a uninarial or binary approach depends on the tumor's location and size and the treating surgeon's preference [15–17]. A uninarial approach is recommended for stage I to III tumors. The multidisciplinary panel that developed the CHEER staging system prefers a three- or four-hand dissection for all but stage I tumors. Therefore, three- or four-hand dissection would be used through a single naris for stage II and stage III. As indicated, the panelists advise using a binarial approach for later-stage tumors, likely because it provides better lateral access through the contralateral naris without the need for angled instrumentation and more workspace for three- or four-hand dissection [5,9,18]. We chose a uninarial approach because the tumor lesions were extraconal.

Preserving orbital volume is the primary reason for considering medial orbital wall reconstruction—especially when resectioning large intraconal or extraconal lesions—placing orbital fat over exposed muscles outside the eye to prevent scarring. A surgeon should avoid orbital fat because it can cause orbital volume decrease, enophthalmos, and diplopia [9].

In a large case series, tumor recurrence was not reported when lesions were removed entirely. However, recurrence may occur when tumors are incompletely removed [19]. Yan and Wu [20] reported three

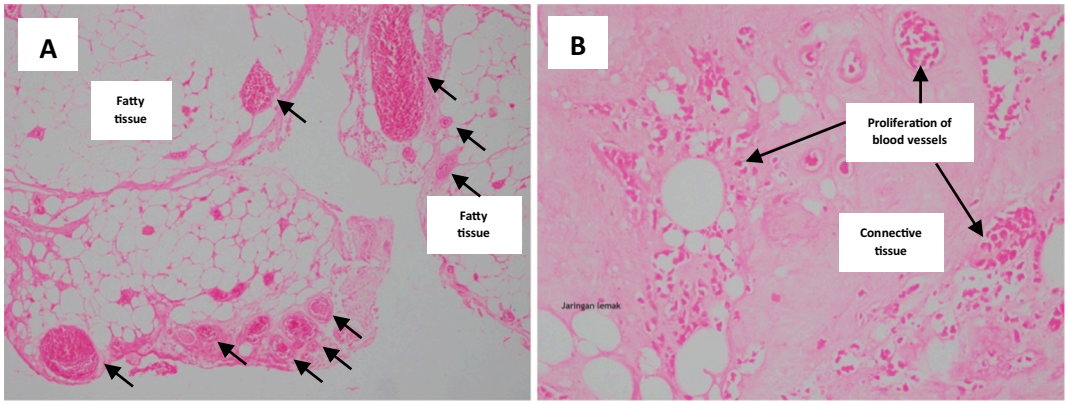

Fig. 4. Hematoxylin-eosin stained sections are shown at (A) 10× and (B) 20× magnification. Histopathological examination showed that the tumor comprised connective tissue, fatty tissue—among which there is a proliferation of blood vessels (arrows) of various sizes containing erythrocytes coated with endothelial cells—and the absence of cytologic atypia.

Table 1
The CHEER staging system [4,5].

Stage	Anatomical details
I	Extraconal cavernous hemangioma of the orbit.
II	An intraconal cavernous hemangioma of the orbit inferior to the medial rectus's horizontal axis and anterior to the ophthalmic artery's inferomedial muscular trunk.
III	Intraconal cavernous hemangioma of the orbit anterior to the inferomedial muscular trunk of the ophthalmic artery and superior to the medial rectus's horizontal axis.
IVA	Intraconal cavernous hemangioma of the orbit posterior to the ophthalmic artery's inferomedial muscular trunk that does not extend into the optic canal.
IVB	Intraconal cavernous hemangioma of the orbit posterior to the ophthalmic artery's inferomedial muscular trunk with extension into the optic canal or an isolated orbital cavernous hemangioma within the optic canal.
VA	Extraconal/intraconal cavernous hemangioma of the orbit with pterygopalatine and/or infratemporal fossa extension via the inferior orbital fissure.
VB	Extraconal/intraconal cavernous hemangioma of the orbit with intracranial extension via the superior orbital fissure.

incidences of recurrence in nine incompletely removed patients. The basic fibroblast growth factor (bFGF) is expressed by orbital cavernous hemangiomas and can promote the development of vascular smooth muscle and endothelial cells, which may contribute to tumor progression. Even after the tumor has been completely excised, the produced and remaining bFGF may retain the ability to promote cavernous hemangioma development [19].

4. Conclusion

An endoscopic transnasal approach is a safe and effective way to access and manage an orbital tumor medial to the optic nerve. It is essential to have a multidisciplinary team with experience in endoscopic procedures. This patient had satisfactory results at their three-month postoperative follow-up. They reported no symptoms, and their CT scan did not show a relapsed mass in the right oculi region.

Patient consent

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

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Ratih Finisanti, Muhammad Fadjar Perkasa, and Adi Matra Prawira: Conceptualization, Methodology, and Software. Ratih Finisanti, Muhammad Fadjar Perkasa, and Adi Matra Prawira: Data curation, Writing-Original draft preparation. Muhammad Faruk: Visualization, Investigation. Muhammad Fadjar Perkasa: Supervision. Ratih Finisanti and Muhammad Faruk: Software, Validation. Ratih Finisanti and Muhammad Faruk: Writing-Reviewing and editing. All authors read and approved the final manuscript.

Conflicts of interest

N/A.

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References

[1] T.E. Darsaut, G. Lanzino, M.B. Lopes, S. Newman, An introductory overview of orbital tumors, Neurosurg. Focus. 10 (2001) 1–9, https://doi.org/10.3171/foc.2001.10.5.2.
[2] P. Yuliawati, N.P.D. Lestari, A case of sinonasal squamous cell carcinoma with secondary involvement of the orbit, Eur. J. Med. Heal. Sci. 3 (2021) 18–20, https://doi.org/10.24018/ejmed.2021.3.1.656.

[3] A. Hentati N. Matar H. Dridi S. Bouali H. Jemel , Bilateral orbital cavernous hemangioma., Asian J. Neurosurg. 13 (n.d.) 1222–1224. doi:10.4103/ajns.AJNS_96_17.
[4] C.M. Low, J.K. Stokken, Typical orbital pathologies: hemangioma, J. Neurol. Surg. Part B Skull Base. 82 (2021) 20–26, https://doi.org/10.1055/s-0040-1722633.
[5] F. Signorelli, C. Anile, M. Rigante, G. Paludetti, A. Pompucci, A. Mangiola, Endoscopic treatment of orbital tumors, World J. Clin. Cases. 3 (2015) 270–274, https://doi.org/10.12998/wjcc.v3.i3.270.
[6] L. Zhang, X. Li, F. Tang, L. Gan, X. Wei, Diagnostic imaging methods and comparative analysis of orbital cavernous hemangioma, Front. Oncol. 10 (2020), 577452, https://doi.org/10.3389/fonc.2020.577452.
[7] E.R. Reshef, B.S. Bleier, S.K. Freitag, The endoscopic transnasal approach to orbital tumors: a review, Semin. Ophthalmol. 36 (2021) 232–240, https://doi.org/10.1080/08820538.2021.1890794.
[8] R.A. Rimmer A.E. Graf J.H. Fastenberg J. Bilyk G.G. Nyquist M.R. Rosen M.R.M.P. Rabinowitz M.R.M.P. Rabinowitz , Management of orbital masses: outcomes of endoscopic and combined approaches with no orbital reconstruction., Allergy Rhinol. (Providence). 11 (n.d.) 2152656719899922. doi:10.1177/2152656719899922.
[9] C. Banks, Q. Husain, B.S. Bleier, Endoscopic endonasal intraconal orbit surgery, worldJ. Otorhinolaryngol. - Head Neck Surg. 6 (2020) 100–105, https://doi.org/10.1016/j.wjorl.2019.07.001.
[10] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230, https://doi.org/10.1016/j.ijsu.2020.10.034.
[11] D. Vogele, N. Sollmann, A. Beck, B. Haggenmüller, S.A. Schmidt, B. Schmitz, T. Kapapa, Y. Ozpeynirci, M. Beer, C. Kloth, Orbital tumors—clinicalradiologic and histopathologic Correlation, Diagnostics 12 (2022) 2376, https://doi.org/10.3390/diagnostics12102376.
[12] E. Ayoub, A. Farid, C. Yahya, E.B. Nizar, H. Meryem, L.A.M. Youssef, B. Meriem, M. Maàroufi, A. Badreeddine, Cavernous hemangioma of the orbit: case report and a review of the literature, Radiol. Case Rep. 17 (2022) 4104–4107, https://doi.org/10.1016/j.radcr.2022.07.101.
[13] A.H.I. Habib, K.H. El-Kaffas, A.S. Mustafa, S.N. Mashour, Role of ultrasound and colored doppler examination in the diagnosis and the classification of the superficial soft tissue vascular anomalies, Egypt. J. Radiol. Nucl. Med. 53 (2022) 89, https://doi.org/10.1186/s43055-022-00753-9.
[14] T.H. Osaki, F.A. Jakobiec, P.R. Mendoza, Y. Lee, A. Fay, Immunohistochemical investigations of orbital infantile hemangiomas and adult encapsulated cavernous venous lesions (Malformation versus Hemangioma), ophthalmic, Plast. Reconstr. Surg. 29 (2013) 183–195, https://doi.org/10.1097/IOP.0b013e31828b0f1f.
[15] L. Rangel-Castilla J.J. Russin R.F. Spetzler Surgical management of skull base tumors., Reports Pract. Oncol. Radiother. J. Gt. Cancer Cent. Pozn. Polish Soc. Radiat. Oncol. 21 (n.d.) 325–35. doi:10.1016/j.rpor.2014.09.002.
[16] K. Melder, N. Zwagerman, P.A. Gardner, E.W. Wang, Endoscopic endonasal approach for intra- and extraconal orbital pathologies, J. Neurol. Surg. B. Skull Base. 81 (2020) 442–449, https://doi.org/10.1055/s-0040-1713940.
[17] D. Widodo, F. Perkasa, K.J. Sjukur, M. Faruk, R. Al-Abqary, Combined transcranial and transnasal endoscopic approach in transnasal-penetrating intracranial injury: A rare case report, Int. J. Surg. Case Rep. 97 (2022), 107422, https://doi.org/10.1016/j.ijscr.2022.107422.
[18] E. El Rassi, N.D. Adappa, P. Battaglia, P. Castelnuovo, I. Dallan, S.K. Freitag, P. A. Gardner, R. Lenzi, D. Lubbe, R. Metson, K.S. Moe, L. Muscatello, H. Mustak, J. F. Nogueira, J.N. Palmer, N. Prepageran, V.R. Ramakirshnan, R. Sacks, C. H. Snyderman, S.T. Stefko, M. Turri-Zanoni, E.W. Wang, B. Zhou, B.S. Bleier, Development of the international orbital cavernous hemangioma exclusively endonasal resection (CHEER) staging system, Int. Forum Allergy Rhinol. 9 (2019) 804–812, https://doi.org/10.1002/alr.22316.
[19] C. Deng, W. Hu, Multiple cavernous hemangiomas in the orbit, Medicine (Baltimore) 99 (2020), e20670, https://doi.org/10.1097/MD.0000000000020670.
[20] J. Yan, Z. Wu, Cavernous hemangioma of the orbit: analysis of 214 cases, Orbit 23 (2004) 33–40, https://doi.org/10.1076/orbi.23.1.33.28992.