Annals of Medicine and Surgery 79 (2022) 103997

Contents lists available at ScienceDirect

Annals of Medicine and Surgery

journal homepage: www.elsevier.com/locate/amsu

Cross-sectional Study

Atypical intraorbital dermoid and epidermoid cyst: A single institution cross-sectional retrospective study

Bahram Eshraghi [a], Azadeh Shahsanaei [b], Mahdi Abounoori [a], Mohsen Pourazizi [a,*]

[a] Isfahan Eye Research Center, Department of Ophthalmology, Isfahan University of Medical Sciences, Isfahan, Iran
[b] Private Office, Isfahan, Iran

ARTICLE INFO

Keywords:
Dermoid cyst
Epidermoid cyst
Intraorbital cyst
Orbital diseases

ABSTRACT

Purpose: Intraorbital epidermoid and dermoid cyst (DC) has been reported in the literature rarely. The current study evaluates clinicopathologic, radiologic, and management of intraorbital DC cases over ten years.
Methods: In this cross-sectional study, the medical records of patients with intraorbital DC treated at the academic referral center for ocular surgery were retrospectively reviewed. Data reviewed included the patient's demographic characteristics, clinical features, imaging, surgical technique, and pathology report.
Results: Nine patients with a rare presentation of intraorbital DC were reviewed within the study period in five presentations (five intraosseous, one intraconal, one dumbbell-shaped with a large part in anterior orbit, one juxta levator palpebral muscle, and one recurrent case with intracranial extension). They ranged from 8 to 53 years of age, with a median of 29 years, and five (55.6%) were female. Histopathological evaluation revealed two cysts were epidermoid.
Conclusion: The current study provides more clinical and radiologic manifestations of rare presentations of DC that highlight the importance of high clinical suspicion in the approach to atypical DC.

1. Introduction

A dermoid/epidermoid cyst (DC) is a rare, usually congenital benign ocular disorder characterized by painless swelling that may be freely mobile or fixed to the skin and deeper structures [1,2]. They are typically superficial and present in children, especially in children five years old or younger [3–5]. The usual sites of involvement are on the subcutaneous tissues of the head and neck, most often found in the periorbital area on lateral eyebrows [1,2].

Involvement of the intraorbital DC has also been reported in the literature rarely and atypical [3]. As reported in large survey studies, only about 0.5% of the lesions were located in the deep orbit [6].

When DC involves the ocular adnexa or orbit, the presenting signs and symptoms findings are commonly nonspecific and may be confusing, especially in adulthood [1,7]. Therefore, radiological evaluation with and without histological evaluation is essential in making the exact diagnosis.

There are limited data about intraorbital DC in the literature. So herein, we report the clinicopathologic, radiologic, and management of intraorbital DC cases over a 10-year period. Characteristic features of these patients would raise the awareness of the ophthalmologist to the diagnosis of DC and so avoid, mistakenly, considering these cases as other intraorbital pathologies.

2. Methods

We conducted a retrospective review of clinical records of patients with intraorbital presentation. After institutional review board approval at the Isfahan University of Medical Sciences, Isfahan, Iran, this study was retrospectively registered (IR.MUI.MED.REC.1399.1052). Also it registered in ClinicalTrials.gov database (ClinicalTrials.gov Identifier: NCT05410431). Dermoid cyst surgeries performed by a single surgeon (BE) over ten years were reviewed between January 2010 and December 2019. Personal surgery records were used to identify intraorbital DC cases. The study was conducted following the provisions of the Helsinki Declaration [8].

Data reviewed included the patient's demographic characteristics, clinical presentation, imaging, treatment modalities, operative report, pathology report, complications, and postoperative outcome. All patient's medical records were examined, and those who had an excision

* Corresponding author. Department of Ophthalmology, Feiz Hospital, Modares St, Isfahan, Iran.
E-mail addresses: eshraghyy@yahoo.com (B. Eshraghi), az.shahsanaei@gmail.com (A. Shahsanaei), Abounoori.mahdi@gmail.com (M. Abounoori), m.pourazizi@yahoo.com (M. Pourazizi).

https://doi.org/10.1016/j.amsu.2022.103997
Received 15 April 2022; Received in revised form 10 June 2022; Accepted 12 June 2022
Available online 20 June 2022

biopsy with histology indicative of dermoid/epidermoid cyst were included. The dermoid/epidermoid cyst was diagnosed by histopathology. Epidermoid cysts are characterized by cystic areas lined by simple squamous epithelium (epidermoid cyst) and containing skin adnexa ("real" dermoid cyst) of tissues from all three germ layers, such as muscle, teeth, bone, cartilage, and so on [9]. Other cystic lesions that did not fit the inclusion criteria were excluded.

The surgical approach was then reviewed regarding the specific anatomic considerations and other factors influencing the surgical approach in each of those cases. Descriptive statistics (SPSS (Statistical Procedures for Social Sciences; Chicago, Illinois, USA) (version 16.0)) were performed to summarize the patient's characteristics.

3. Results

A total of nine patients with a rare presentation of intraorbital DC were reviewed within the study period in five presentations (five Intraosseous, one Intraconal, one dumbbell shape with a large part in anterior orbit, one juxta levator palpebral muscle, and one recurrent case with intracranial extension). Of these, five (55.6%) were female, and four (44.4%) were male. They ranged from 8 to 53 years of age, with a median of 29 years. Regarding histopathological evaluation, seven cysts were dermoid, and two were epidermoid. Table 1 presents the nine patient's demographic, clinical features, imaging, surgical technique, and pathology report (Table 1).

Of the nine current cases, five presented as an intraosseous DC in the superior sphenoid wing. All of them were adults (aged from 22 to 53 years). The most important clinical presentation in these patients was hypophthalmos. The cardinal feature in most intraosseous DC was a heterogeneous and irregular mass with irregular bone erosion in orbital computed tomography (CT) scan (Figs. 1 and 2). Surgery in these cases was complicated by cyst rupture when excision was attempted due to adhesion of the irregular cyst wall to the bone. Complete excision was impossible in these patients. Bone curettage was performed for all 5 cases. Histopathologic examination results showed that the mass was a dermoid cyst. Also, the either early or late complication was not seen after surgery.

An 8-year-old girl presented with marked obvious proptosis in the right eye (Fig. 3). The location of the lesion was intraconal, lateral to the optic nerve. MRI with gadolinium shows a well-defined mass with ring enhancement. She underwent complete excision of the cyst via a transconjunctival approach. Histopathologic examination results showed that the mass was an epidermoid cyst. Also, the either early or late complication was not seen after surgery.

There was one rare case of DC in a 25-year-old female who presented with diplopia and hypophthalmos (Fig. 4). She was a recurrent DC case with a history of anterior orbitotomy three years ago. CT scan demonstrates a large hypodense mass with sphenoid wing erosion associated with intracranial extension. Further evaluation with magnetic resonance imaging (MRI) of orbits confirmed the intracranial extension of the lesion, which demonstrated hypointensity to fat and brain on T1-weighted and hyperintensity to the brain on T2-weighted images. With the second surgery, total removal of encapsulated mass was performed via craniotomy with the help of neurosurgeons. The pathological diagnosis was reported to be an epidermoid cyst. Also, the either early or late complication was not seen after surgery.

Another rare case was a 22-year-old female who presented with s-shape lid puffiness. CT showed a dumbbell-shaped hypodense mass, a large part in the anterior orbit and a small part in the temporal fossa (Fig. 5). This was completely excised using an anterior orbital approach with small bone removal and extraction of both parts. Histopathologic examination results showed that the mass was a dermoid cyst. Also, the either early or late complication was not seen after surgery.

Another rare case was a DC located on the levator muscle of the right palpebral in a 29-year-old man. He presented with mild ptosis and lid puffiness (Fig. 6). The supra-nasal mass was complete remove via orbitotomy, and the clinical diagnosis of DC was confirmed histopathologically. The early complication of surgery was levator edema which formed two weeks after surgery. Also, the late complication of surgery was not observed.

4. Discussion

We presented a series of unusual locations and presentations of

Table 1
Characteristics of 9 patients with intraorbital dermoid/epidermoid cysts.

Pt NO.	Age/ Sex	Location	Examination	CT Findings	MRI Findings	Surgery technique	Pathology
1	8/F	R/Intraconal, lateral to ON	Proptosis, Mild choroidal fold	Not available	Well defined Ring enhancement	Transconjunctival, complete removal	Epidermoid
2	22/F	L/IO, Sup, Sphenoid wing	Ptosis, Lid puffiness	Heterogenous (hypo>), irregular	Not available	Ant orbitotomy, complete removal	Dermoid
3	22/F	R/Supra-temporal, Ant	Lid puffiness, S shape	Hypo, Dumble-shape: large part in ant orbit, a small part in the temporal fossa	Not available	Ant orbitotomy, complete removal	Dermoid
4	25/F	L/Sup, Sphenoid wing	Hypophthalmos, Diplopia (up)	Hypo, large, complete sphenoid wing erosion, intracranial extension	T1:hypo to fat and brain T2:hyper to brain Well defined, heterogeneous, intra cranial extension	History of ant orbitotomy 3 years ago The second procedure was done via craniotomy and teamwork mass was removed	Epidermoid
5	29/M	L/IO, Post-Sup	Mild proptosis, Lid puffiness	Hypo, localized limited intraosseous posterior mass	Not available	Lateral orbitotomy, complete removal	Dermoid
6	29/M	R/Supra-nasal, Ant, on levator muscle	Mild ptosis, Lid puffiness,	Not available	well defined supra-nasal mass, T1: hyper	Ant orbitotomy, complete removal	Dermoid
7	30/F	L/IO, Sup, Sphenoid wing	Hypophthalmos, Diplopia (up), Choroidal fold	Heterogenous (hypo>), large, extension to apex, ant to post	Not available	Lateral orbitotomy, complete removal	Dermoid
8	45/M	L/IO, Sup, Sphenoid wing	Hypohthalmos, Proptosis	Heterogenous (hypo>), large, bone erosion in apex, beside meninge, ant to post	T1 hypo>, T2 hyper>, heterogenous, distinct border	Ant orbitotomy, complete removal	Dermoid
9	53/M	R/IO, Sup, Sphenoid wing	Hypophthalmos, Diplopia (up)	Iso, irregular, distinct border	Not available	Ant orbitotomy, complete removal	Dermoid

F: Female, M: Male, R: Right, L: Left, ON: Optic nerve, Ant: Anterior, Post: Posterior, Sup: Superior, Hypo: Hypodense, Hyper: Hyperdense, IO: Intraosseous.

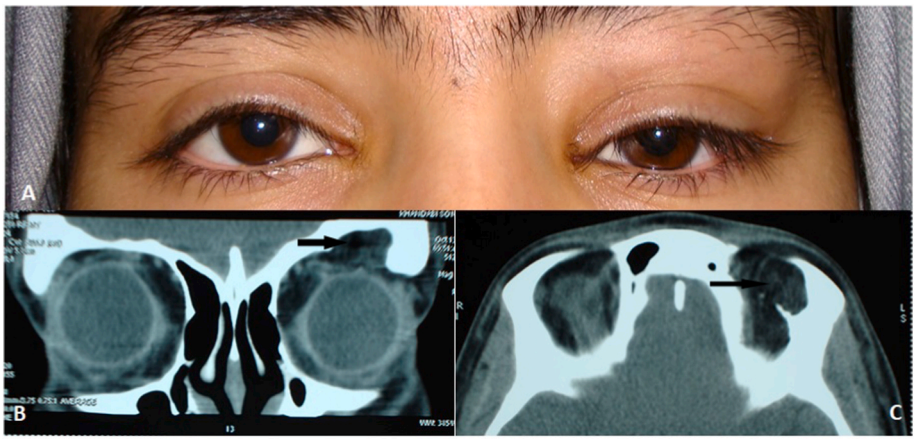

Fig. 1. 22-year-old female with intraosseous dermoid cyst (Case NO.2)**. A:** Clinical image of the patient presented with left ptosis and upper lid puffiness. **B:** Coronal orbital computed tomography (CT) shows an irregular heterogeneous mass located superolateral in the left orbit. **C:** Axial orbital CT image of the same patient.

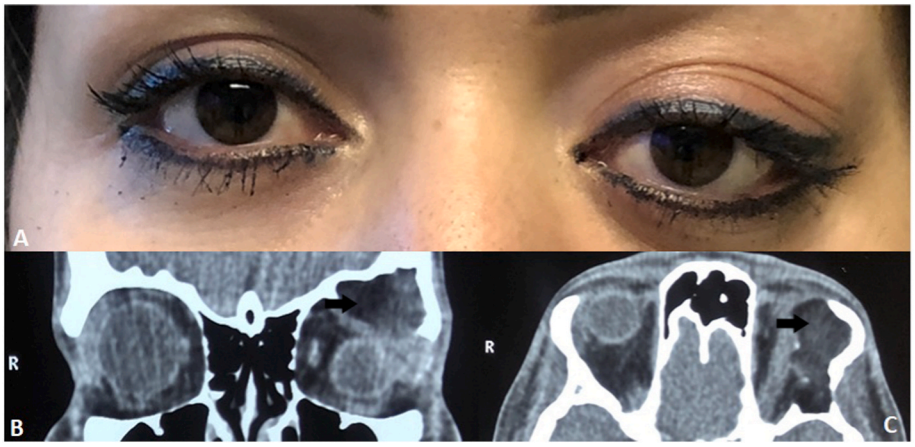

Fig. 2. 30-year-old female with intraosseous dermoid cyst (Case NO.7**). A:** Clinical image of the patient presented with hypophthalmos. **B:** Coronal orbital computed tomography (CT) shows a large hyper/hypo-dens mass with orbital erosion in the left orbit. **C:** Axial orbital CT image of the same patient with extension into the orbital apex.

intraorbital DC. The present report demonstrated that the mean age of patients with intraorbital DC was higher than periocular that usually present in early infancy.

DC arises in the early stages of embryonic development, between the third and fifth week of intrauterine life, so it is considered a congenital cyst probably due to faulty separation of the neuroectoderm and cutaneous ectoderm [10,11]. Ocular DC is divided into superficial and deep types. Superficial type presenting as a superficial slowly growing mass at early childhood [5]. At the same time, the average age of children presenting with deep orbital dermoid is higher [12,13]. Only about 0.5% of the lesions were located in the deep orbit, as reported in large survey studies [14,15]. Intraorbital DC is usually presented as deep orbital DC [5]. Deep orbital DC may not be detected until teenage or later in adult life. Intraorbital DC tends to manifest in the second and third decades of life with atypical sing and symptoms [12]. In our report, eight patients were adults, and one was pediatric.

Various studies have reported that atypical DC has a more complicated presentation with a higher possibility of misdiagnosis. Intraorbital DC depending on the location and size of the cyst, may present proptosis, diplopia, and ocular eye movement restriction [12]. In our report, the most clinical sign of patients was hypophthalmos, proptosis, lid puffiness, and ptosis.

We report a case of an intraconal epidermoid cyst in an 8-year-old that is very rare. Although the DC is not a classical differential diagnosis in the list of intraconal orbital tumors, it should be considered as a differential diagnosis according to the characteristic CT or MRI features.

Currently, good quality imaging plays a major role in the detection of the cyst and also provides more data about the location of the lesion, its components, and its effects on adjacent structures [16]. In typical superficial DC preoperative diagnosis is easier but in the case of the intraorbital cyst, almost always radiological investigation with CT or MRI imaging is necessary although the finding of this imaging study can be not specific [12,17]. Although preoperative imaging can be a useful investigation in intraorbital DC and is done mainly for medico-legal reasons, it may not detect completely extensions and depths of lesion or involvement of adjacent structure [17]. So, in the management of intraorbital DC, the ophthalmologist should be aware of additional complementary procedures when removing atypical intraorbital DC. In our report, the heterogeneous pattern of cyst in the radiological investigation was present in most cases. The importance of awareness of atypical DC beyond that in addition to atypical clinical presentation, the radiological investigation is doubt potentially and can be caused mixed missed cases. So histopathological evaluation is essential for confirmation of the diagnosis.

Currently, the best management for typical periocular DC is total removal [10], but complete resection of some intraorbital DC could be complicated by the location of the cyst and the involvement of orbital

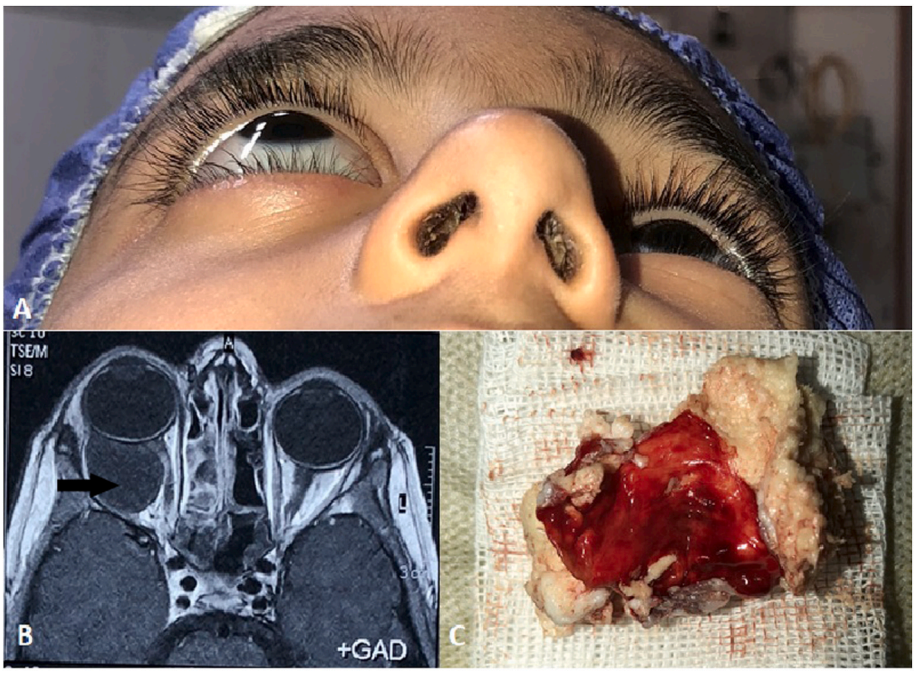

Fig. 3. 8-Year-old girl with intraconal epidermoid cyst (Case NO.1**). A:** Clinical image of the patient presented with right proptosis (worm's-eye view). **B:** Axial contrast-enhanced magnetic resonance imaging (MRI) shows a well-defined mass with ring enhancement on the right orbit. **C:** Photograph of gross pathology after complete removal of mass via transconjunctival approach.

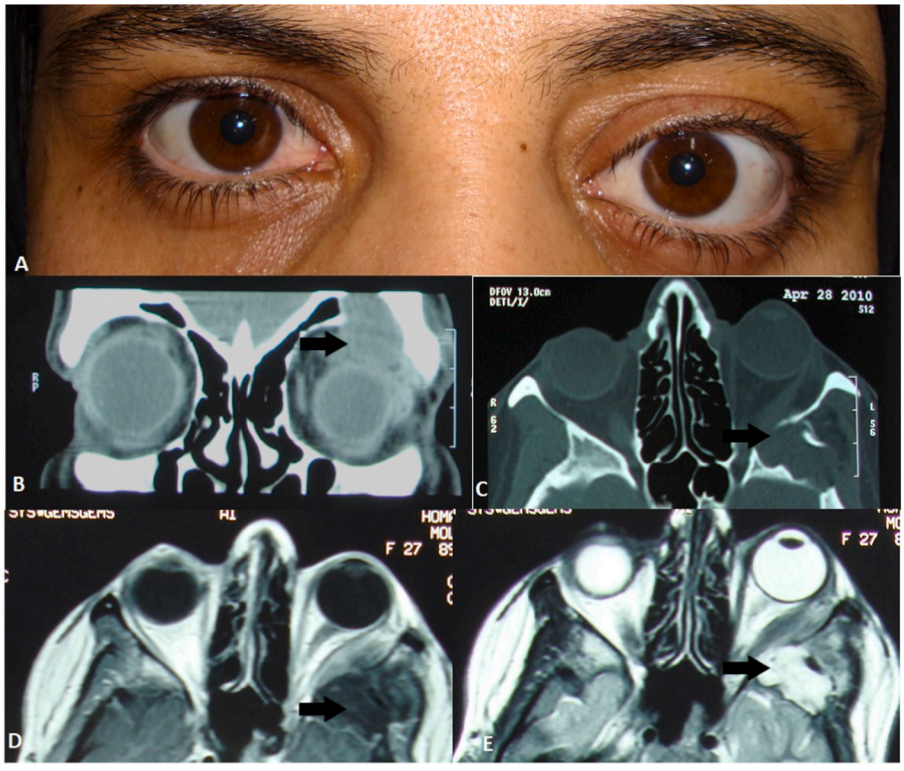

Fig. 4. 25-year-old female with epidermoid cyst associated with intracranial extension. (Case NO.7**). A:** A young woman presented with hypophthalmos and diplopia on the left eye. She had a history of orbitotomy previously**. B and C:** Coronal and axial CT images of the orbits show bone destruction and upward mass extension. **D:** Axial T1 weighted magnetic resonance imaging (MRI) shows hypodense mass to fat and brain with heterogenicity and intracranial extension. **E:** Axial T2 weighted MRI shows the obvious extension of mass lesion to the brain tissue.

structure, so total removal could not be undertaken completely in some cases, including interosseus DC.

The majority of the lateral intraorbital DC have bony involvement, so it is essential to understand the relationship between the cyst and bone in surgical planning. In these cases, all epidermal elements of the cyst must be removed, and burring of the bone may be necessary [7]. We present five cases of a rare intraosseous location of a DC treated using an incision and curettage approach. It may be difficult or impossible to excise complete intraosseous DC. In such cases, some surgeons have advocated evacuating cyst contents to allow for easier dissection around the lesion [18]. This has also been appropriate in the authors' experience with intraosseous DC. In addition, burring of the bone is necessary

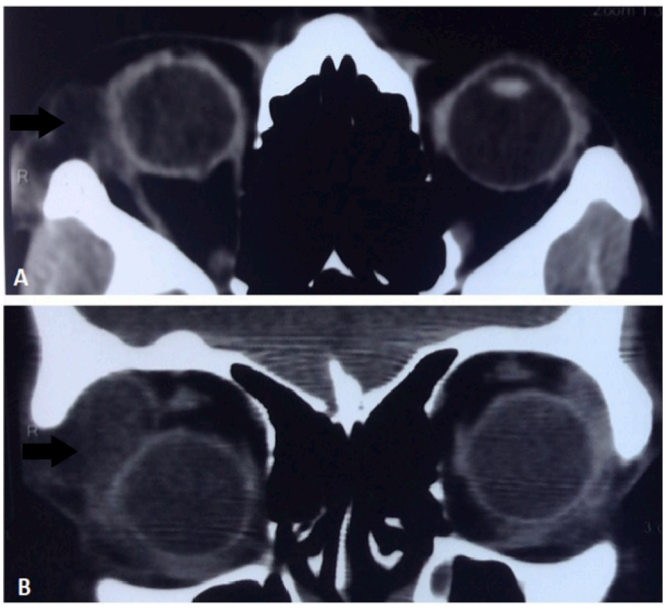

Fig. 5. 22-year-old female with a dumbbell-shape dermoid cyst. (Case NO.3**). A:** Axial and **B:** Coronal orbital computed tomography (CT) shows a hypodense mass. A large part of the mass is located in the anterior orbit and a small part is located in the temporal fossa.

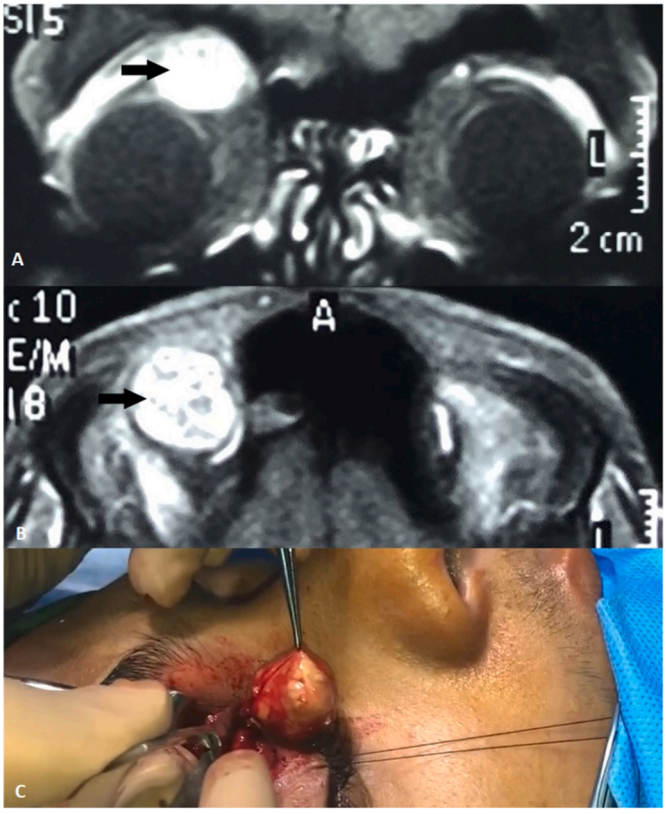

Fig. 6. 29-year-old male with a dermoid cyst on levator palpebral muscle (Case NO.6**). A and B:** Coronal and Axial T1 weighted magnetic resonance imaging (MRI) shows a well-defined mass in the supra nasal area of the right eye**. C:** Intraoperative image of complete removal of the dermoid cyst via anterior orbitotomy.

in cases where there is an intimate connection of the cyst wall to the bone.

Since the published literature describing intraorbital DC is limited, the significance of the current study is providing more clinical and radiologic manifestations of rare presentations of DC that highlight the importance of high clinical suspicion in the approach to atypical DC. Our study also had several limitations, including the small population of the study and the retrospective nature of the study.

In conclusion, we indicated that in addition to the atypical clinical presentation of intraorbital DC, the radiological investigation is doubted potentially and can be caused mixed missed cases. Also, in the management of intraorbital DC, the ophthalmologist should be aware of additional complementary procedures when removing atypical intraorbital DC.

Sources of funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Consent

Not applicable.

Author contributions

BE: study concept or design, data analysis, revise the manuscript, AS: data collection, data analysis interpretation, revising the manuscript, MP: study concept or design, data analysis, writing the paper, MA: data collection, revising the manuscript, editing, and finalizing the manuscript,

Ethics approval

Ethics approval for this report was obtained from the Ethics Committee of Isfahan University of Medical Sciences, Isfahan, Iran in accordance with the Declaration of Helsinki.

Registration of research studies

Name of the registry: Unique Identifying number or registration ID: IR. MUI.MED.REC.1399.1052.

ClinicalTrials.gov Identifier: NCT05410431.

Guarantor

Dr. Bahram Eshraghi.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.103997.

References

[1] J.A. Shields, C.L. Shields, Orbital cysts of childhood–classification, clinical features, and management, Surv. Ophthalmol. 49 (3) (2004) 281–299.

[2] G. Bonavolonta, D. Strianese, P. Grassi, et al., An analysis of 2,480 space-occupying lesions of the orbit from 1976 to 2011, Ophthalmic Plast. Reconstr. Surg. 29 (2) (2013) 79–86.
[3] B.E. Howard, M.M. Masood, J.M. Clark, B.D. Thorp, Intraorbital dermoid cyst with zygomaticofrontal suture erosion in an adult, J. Craniofac. Surg. 30 (2) (2019) 514–515.
[4] B.A. Golden, M.S. Jaskolka, R.L. Ruiz, Craniofacial and orbital dermoids in children, Oral Maxillofac. Surg. Clin. 24 (3) (2012) 417–425.
[5] R. Ahuja, N.F. Azar, Orbital dermoids in children, Semin. Ophthalmol. 21 (3) (2006) 207–211.
[6] H.Z.B. Alaya, M. Gaha, Y. Limem, et al., Kyste dermoïde intraorbitaire. À propos d'une observation et revue de la littérature, J. Fr. Ophtalmol. 36 (2) (2013) 172–177.
[7] J. Bajric, G.J. Harris, The spectrum of orbital dermoid cysts and their surgical management, Orbit 39 (4) (2020) 266–275.
[8] G. Mathew, R. Agha, J. Albrecht, et al., STROCSS 2021: strengthening the reporting of cohort, cross-sectional and case-control studies in surgery, International Journal of Surgery Open 37 (2021), 100430.
[9] M. Dutta, J. Saha, G. Biswas, S. Chattopadhyay, I. Sen, R. Sinha, Epidermoid cysts in head and neck: our experiences, with review of literature, Indian J. Otolaryngol. 65 (1) (2013) 14–21.
[10] J.F. Martinez-Lage, J. Ramos, A. Puche, M. Poza, Extradural dermoid tumours of the posterior fossa, Arch. Dis. Child. 77 (5) (1997) 427–430.
[11] A.S. Barkley, S.M. Susarla, A. Lee, Frontotemporal dermal sinus tract with 2 connected intradiploic dermoid cysts: a rare case and review of the literature, World Neurosurg 127 (2019) 350–353.
[12] R.P. Sherman, J. Rootman, J.S. Lapointe, Orbital dermoids: clinical presentation and management, Br. J. Ophthalmol. 68 (9) (1984) 642–652.
[13] B. Eshraghi, H. Hashemian, M. Safizadeh, M. Akbari, Periocular dermoid cysts: common sites in referral cases in an Iranian population, Journal of Current Ophthalmology 25 (4) (2013) 309.
[14] J.A. Shields, C.L. Shields, R. Scartozzi, Survey of 1264 patients with orbital tumors and simulating lesions: the 2002 Montgomery Lecture, part 1, Ophthalmology 111 (5) (2004) 997–1008.
[15] S.J. Chawda, I.F. Moseley, Computed tomography of orbital dermoids: a 20-year review, Clin. Radiol. 54 (12) (1999) 821–825.
[16] H. Zaghouani Ben Alaya, M. Gaha, Y. Limem, et al., [Intraorbital dermoid cyst. Case report and review of the literature], J. Fr. Ophtalmol. 36 (2) (2013) 172–177.
[17] A. Sari, T. Yener, A. Sari, Y. Basterzi, Atypical presentation of an intraorbital dermoid cyst, J. Craniofac. Surg. 19 (6) (2008) 1715–1716.
[18] A.A. El-Ghafar, H.A. ElKhair, Evacuation of dermoid cysts before excision, Journal of the Egyptian Ophthalmological Society 106 (4) (2013) 235.