Case report

Ectopic ocular tissue in testicular teratoma: A case report and review of the literature

Arwa Z. Alromaih a, Amany A. Fathaddin b, Abdullah I. Almater a,c, Hind M. Alkatan a,b,c,*

a Department of Ophthalmology, College of Medicine, King Saud University, Riyadh, Saudi Arabia
b Department of Pathology and Laboratory Medicine, College of Medicine, King Saud University, Riyadh, Saudi Arabia
c King Saud University Medical City, King Saud University, Riyadh, Saudi Arabia

A R T I C L E   I N F O

Keywords:
Teratoma
Ocular tissue
Uvea
Retina
Teratoma
Testicular

A B S T R A C T

Introduction and importance: Teratoma is a common neoplasm in prepubertal and post-pubertal periods. It consists of various types of tissues arising from different germinal layers, endoderm, mesoderm, and ectoderm. Ectopic ocular tissue is a rare phenomenon, with only few reported cases in other locations.

Case presentation: This is a 10-month-old boy who presented with a painless scrotal mass. Following orchidectomy, the excised mass confirmed the presence of uveal and retinal tissues originating in a benign testicular teratoma by histopathological examination.

Discussion: Choroidal and retinal tissue are the most frequently encountered ectopic ocular tissue, while the least observed tissue is the lens. Most of the reported cases of ectopic ocular tissue present in ovarian teratomas. The only 2 previously reported cases of ocular-like tissue in testicular teratoma lack well-defined medullary epithelium, uvea, and retinal tissue as in our case.

Conclusion: To our knowledge, developing ocular tissue in a testicular teratoma is extremely rare. Herein we report a unique case with mature defined ocular tissue within a testicular teratoma in an infant, which should not be overlooked.

1. Introduction

Teratoma is a neoplasm derived from a non-germ cell neoplasia in situ (GCNIS) a sub-set of germ-cell origin [1]. The tumor is composed of various types of tissue representing distinct germinal layers (endoderm, mesoderm, ectoderm) [2]. The teratoma can be either mature (well-differentiated) or immature (primitive) tissue [3]. Most bodily tissues are typically described in teratoma, including skin, bone, cartilage, connective tissue elements, epithelium of the respiratory and gastrointestinal tracts, smooth and striated muscle, and glial and nerve tissues. However, gonadal tissues, liver and ocular structures are very rarely seen [4]. Teratomas may appear anywhere in the body but are commonly seen in the gonads [4]. Among prepubertal testicular tumors, teratoma is the second most common to arise following yolk sac tumor and occurs with ranging frequency between 13% and 60% [3]. In contrast to post-pubertal teratomas, pre-pubertal teratomas are benign and metastasis has not been reported [3]. This case reveals the first-reported, isolated well-recognized uveal and retinal tissues in a mature prepubertal testicular teratoma in the English-written literature. This case report has been prepared and reported in accordance with the SCARE criteria [5].

2. Presentation of case

A full-term healthy 10-month-old boy, presented with an incidental finding of left scrotal painless hard mass. The mass was initially noticed by the parents 7 months prior to presentation. They asked for medical advice when they noted gradual increase in the size of the mass over the last month. The medical history, pregnancy, and delivery were normal and there was no family history of genetic disorders. External examination demonstrated left scrotal non-tender hard mass measuring 4 × 3 cm in size, with no change in skin color, and negative transillumination test. The right scrotum was normal. Ultrasonography of left scrotum showed a 3.4 × 2.7 cm well-circumscribed, oval shaped mass with mixed echogenicity, The mass contained both cystic and solid component, with no clear visualization of the left testicle. Color Doppler revealed mild...
From testicular or extragonadal sites are mostly of immature neuro-ectodermal tissue, which are summarized in Table 1. The most frequently encountered ectopic ocular tissues arise as part of a teratoid lesion such as nevus, meningeal or uveal tissue differentiation in a mature ovarian cystic teratoma, or arising from uveal tissue differentiation in a mature ovarian cystic teratoma [9,10]. Nonetheless, only few reports have described teratomas containing ectopic ocular tissue and no malignant features. Thus, chemotherapy was not required for treatment, and regular follow up was planned.

3. Discussion

Developing ectopic ocular tissue in extra-orbital sites is uncommon [4]. Nonetheless, only few reports have described teratomas containing ectopic ocular tissue, which are summarized in Table 1. The most frequently encountered ectopic ocular tissues are the choroid and retina. Occasionally, the eyelids, cornea, sclera, and vitreous can be seen. However, the lens is rarely observed [4,6]. Most of the ectopic ocular tissues arise within ovarian or sacrococcygeal teratomas. Ocular teratomas originating from testicular or extragonadal sites are mostly of immature neuro-ectodermal tissue without structural organization, such as retina-like, choroid-like, or ciliary body-like tissues. In prepubertal children, mature teratomas are more common and are usually benign. However, it carries high risk of metastasis if present in adults [3]. Till date, there are no reported cases of well-organized ectopic ocular tissue developing in a testicular teratoma in the English-written literature.

The earliest report of ocular structures in teratomas was published in the French-literature by Verneuil in 1855 [6]. He described a teratoma of the testis with a cyst containing pigment epithelium with characteristics of the pigment epithelium of the eye. He cited Saint-Donat (1696), who described a massive teratoma of the testis containing a bony mass with two orbit-like cavities enclosing black vesicles lined with a uveal structure and containing a “flowing lymph”, which he referred to as aqueous humor. For diagnostic purposes, morphology of testicular teratoma can be easily recognized. Immunohistochemically, NSE is helpful in identifying glial and neural tissue in teratomas. Unlike yolk sac tumors, teratomas do not stain positively for AFP, and elevated serum AFP levels are not reported in patients with these tumors. Ki67 proliferation fraction shows the proliferative activity of the cells.

Sharpe and co-authors [8], were the first to report isolated corneal and conjunctival tissue with developing lenticular structures in an adult woman with mature ovarian teratoma. The rare occurrence of the lens in teratoma was justified by Mann in 1950, who demonstrated that the development of lenses occurred in response to the presence of an optic cup as an inducer structure for the development in the lens, which requires proximity of surface ectoderm [9].

Breinin et al. [4] and Mercur et al. [9], reported cases of teenage girls who presented with malignant ovarian teratoma containing a well-formed optic cup, normal developing embryonic retina, and undifferentiated tissue, closely resembling neuroepithelioma retinae. Interestingly, the development of primary malignant melanoma arising from uveal tissue differentiation in a mature ovarian cystic teratoma has been reported in 2 adult women, which ultimately resulted in widespread metastasis necessitating systemic therapy [10,11]. The occurrence of primary ovarian malignant melanomas was exceptionally rare, because the ovary does not contain melanocytes, and it can only arise as part of a teratoid lesion such as nevus, meningeal or uveal tissue [9,10].

Sergi et al. [12] and Takamatsu et al. [13] reported a fetal sacrococcygeal teratoma with the development of a completely formed eye in the former, and retina-like structure with immature neural components of true rosettes and primitive neural tubes in the latter. They pointed the importance of blood supply and high oxygen concentration for the formation of rods and cones based on their hypothesis that these factors are important in the differentiation of the cells. Dubinski et al., described a rare case of an intracranial teratoma with immature neuro-ectodermal tissue in a malformed eye anlage [14].

4. Conclusion

To the authors’ best knowledge, this is the first case of ectopic welldifferentiated in testicular teratoma of an infant in the English-written literature. This is an interesting histopathological finding with rare occurrence. It is important for this tissue not to be overlooked since immature components with malignant potential might be found in this ectopic tissue. In addition, we have reviewed and

Fig. 1. Histopathological photos of sections from the teratoma showing mixture of benign mature tissue consisting of mucinous glands, skin with skin appendages, and brain tissue in A, B, and C respectively (Original magnification x100 Hematoxylin and eosin).
summarized the reported cases of ectopic ocular tissue in teratomas found in other locations.

Provenance and peer review
Not commissioned, externally peer-reviewed.

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

Ethical approval
IRB is not required for case reports. However, information was obtained and reported in a manner that was compliant with the standards set forth by the Health Insurance Portability and Accountability Act, and the Declaration of Helsinki as amended in 2013.

Consent
General informed written consent was obtained from the patient’s guardians including permission for anonymous use of photos and for

Table 1
Summary of the reported cases of ocular tissue found in a teratoma.

| Study               | Year | Age  | Gender | Site of teratoma | Type of teratoma          | Ocular tissue                                                                 |
|---------------------|------|------|--------|------------------|---------------------------|------------------------------------------------------------------------------|
| Breinin et al. [4]  | 1950 | 19 years | Female | Ovarian         | Malignant, mature         | Well-formed optic cup, embryonic retina, and undifferentiated neuroepithelium retinae |
| Sharpe et al. [7]   | 2018 | 42 years | Female | Ovarian         | Benign, mature            | Corneal and conjunctival tissue with developing lenticular structures          |
| Lengyel et al. [9]  | 2020 | 39 years | Female | Ovarian         | Malignant, cystic         | Uveal tissue                                                                 |
| Takamatsu et al. [12]| 2012 | 29-week fetus | Female | Sacrococcygeal  | Benign, mature, cystic    | Retina-like structure                                                        |
| Sergi et al. [11]   | 1999 | 29-week fetus | Female | Sacrococcygeal  | Benign, mature            | Completely formed eye                                                        |
| Mercur et al. [5]   | 1976 | 14 years | Female | Ovarian         | Malignant, mature         | Well-formed optic cup and embryonic retina                                    |
| Vermeul [6]         | 1855 | Not available | Male   | Testicular      | Not available             | Pigmented epithelium similar to that of the eye                               |
| Dubinski [13]       | 2016 | 3 years | Male   | Intracranial    | Benign, mature            | Malformed eye anlage                                                        |
| Moehrle et al. [10] | 2001 | 56 years | Female | Ovarian         | Malignant, cystic         | Uveal tissue                                                                 |
| Saint-Donat et al   | 1696 | Not available | Male   | Testicular      | Not available             | Black vesicles lined by uveal-like tissue and filled by fluid                |
reporting.

Research registration

Not applicable.

Guarantor

Hind M. Alkatan

CRediT authorship contribution statement

Arwa Z. Alromaih: Review of chart for data collection, literature review and first draft of the case report.

Amany A. Fathaddini: Histopathological review of slides and images.

Abdullah I. Almater: Literature review and first draft of the case report.

Hind M. Alkatan: Study design, histopathological examination, and final tissue diagnosis, taking images, and overall review for editing of the manuscript as the corresponding author.

Declaration of competing interest

None.

References

[1] H. Moch, P.A. Humphrey, T.M. Ulbright, V.E. Reuter, WHO Classification of Tumours of the Urinary System And Male Genital Organs, 4th ed., International Agency for Research on Cancer, Lyon, 2016.

[2] K.I. Al-Obaidy, M.T. Idrees, Testicular tumors: a contemporary update on morphologic, immunohistochemical and molecular features, Adv. Anat. Pathol. 28 (4) (2021 Jul) 258–275.

[3] B.S. Carver, H. Al-Ahmadie, J. Sheinfeld, Adult and pediatric testicular teratoma, Urol. Clin. N. Am. 34 (2) (2007 May) 245–251.

[4] G.M. Breinin, The eye in teratomas, Arch. Ophthalmol. 43 (3) (1950 Mar 1) 482.

[5] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, A. Thoma, et al., The SCARE 2020 guideline: updating consensus Surgical Case Report (SCARE) guidelines, Int. J. Surg. 84 (2020 Dec) 226–230.

[6] L. Mercur, T.G. Farkas, P.M. Anker, R. Brill, M.L. Mund, Developing eye in an ovarian teratoma, Arch. Ophthalmol. 94 (4) (1976 Apr 1) 597–598.

[7] A. Verneuil, Inclusion scrotale et testiculaire. Arch gen de med 1 and 2, 1855.

[8] R. Allan Sharpe, C.T. Welsh, L.J.P. Perry, Ocular structures in a mature ovarian teratoma (cited 2021 Dec 5), Am. J. Ophthalmol. Case Rep. 13 (2018 Mar 1) 20–21. Available from: https://pubmed.ncbi.nlm.nih.gov/30505981/.

[9] I. Mann, The Development of the Human Eye Vol. chap 3, Grane & Stratton, Inc, New York, 1950.

[10] K. Lengyel, F. Young, A. Kucukmetin, N. Cresti, R. Plummer, A. Ralte, et al., BRAF wild-type, PTEN mutant malignant uveal melanoma arising within a mature ovarian teratoma: a case report and review of the literature, Int. J. Gynecol. Pathol. 39 (4) (2020 Jul 1) 321–326.

[11] M. Moehrle, H. Fischbach, B. Nuessle, G. Rassner, Primary malignant melanoma arising in a cystic necrotic ovarian teratoma (cited 2021 Dec 5), Eur. J. Obstet. Gynecol. Reprod. Biol. 99 (2) (2001 Dec 1) 268–271. Available from: https://pubmed.ncbi.nlm.nih.gov/11788186/.

[12] C. Serpi, V. Ehemann, B. Beedgen, O. Linderkamp, H.F. Otto, Huge fetal sacrococcygeal teratoma with a completely formed eye and intratumoral DNA ploidy heterogeneity, Pediatr. Dev. Pathol. 2 (1) (1999 Jan 10) 50–57.

[13] M. Takamatsu, H. Aoki, Y. Hirose, K. Kobayashi, H. Tomita, T. Runo, Teratoma showing the features of retinal structure: a case of sacrococcygeal teratoma (cited 2021 Dec 5), Oncol. Lett. 3 (5) (2012 May) 1025–1026. Available from: https://pubmed.ncbi.nlm.nih.gov/22783384/.

[14] D. Dubinski, M. Mittlemann, G. Marquardt, D.S. Tews, A. Noack, B. Behmanesh, et al., Immature teratoma of the tectum mesencephali with histopathological detection of rudimentary eye anlage in a 3-year-old boy: report of a rare case, Neuropathology 36 (6) (2016 Dec) 556–560.