Orbital teratomas: An overview

Sonia Phulke

Consultant, Dept. of Ophthalmology, Drishti Eye Hospital, Sector 11, Panchkula, Haryana, India

***Corresponding Author:**
Email: soniavaidya2010@yahoo.com

Abstract
Teratoma accounts for almost 6.6% of childhood tumours. The most common locations for teratomas are gonadal, sacrococcygeal (57%), retroperitoneal, and mediastinal regions. Teratomas are usually considered as benign tumours, although they grow rapidly, but very rarely malignant. Orbital teratoma usually presents as severe unilateral proptosis or as orbitofacial asymmetry in healthy newborns. Orbital teratomas may remain dormant for years in adults and suddenly present as an acute painful proptosis due to hemorrhage or intracystic accumulation of secretion secondary to accidental trauma. The tumour can vary in size from small to extensive one. The article aims to describe clinical, histopathological, radiological characteristics and management of orbital teratomas.

Keywords: Germ cell layer, Orbit, Orbital teratoma.

Introduction

Orbital teratomas are the large tumour, which are evident at the time of birth. The word "Orbital teratoma" describes the teratoma in orbit only, but periorbital or intracranial extensions are also had described. Congenital teratomas of the head and neck account for 4% of total teratomas and orbital teratomas are rarest among them. The tumour presents as large mass in retro-orbital space with extreme proptosis. The size of the tumor can vary from very small to extensive one. This tumour needs to be resceted immediately after birth to prevent more destruction of adjacent structures. The aim of the present article is to give an overview on orbital teratomas.

"Teratomas" word in Greek means "monstrous growth".

Synonyms are- dysembyeoma, teratoblastoma, organoid tumour, teratoid tumour

Teratomas are group of congenital tumours representing components of normal derivative of all three germinal layers i.e ectoderm, mesoderm and endoderm. According to Marchand Bonnet "a teratoma is formed by wandering embryonic cells derived from blastomers detached from their connexions and able to give rise to other structures by means of differentiation". Duek-Elder (1952) classified the teratoma as follows;

1. A complete foetus implanted in the orbit (orbitopagus parasiticus).
2. A portion of a second foetus in the orbit.
3. A tumour consisting all three germinal layers forming a shapeless mass without regular arrangement.
4. A tumour containing representatives of two germinal layers only.

Normally, germ cells are not found in the orbit or extragonadal sites. The proper differentiation of these germ cell forms distinctive layers like, ectoderm, mesoderm and endoderm, which further forms different normal tissues or organs. Undifferentiated germ cells can remain quiescent in yolk sac until the vascular system develops and organogenesis complete. This undifferentiated germ cell are then carried by the blood stream to all vascular part of the body, among them head and neck regions are most suitable regions. Sometimes these ectopic germ cells abort themselves or sometimes they survive and give rise to germ cell tumour.

The first of teratoma was reported approximately 2000 BC on a babylonial cuneiform tablets.[1] In 1863, Holmes T, first reported the case of orbital teratoma,[2] since then number of cases with varied presentation like a complete parts of a fetus protruding from an infant's orbit and even an entire fetus (orbitopagus paraciticus) implanted in the orbit is being described.[3,4]

Teratoma are the most common congenital tumour, but orbital involvement is very rare, comprises almost - 1.3% of all orbital tumours in children.[5,6]

Teratoma is variant of extragonadal germ cell tumour which typically involves midline structures. Clinically, orbital teartoma can be primarily intraocular, combined orbital and extraorbital and secondary orbital teratoma.[7]

Habal in 1990 proposed a classification of orbital teratomas based on the extension of the tumour.[8]

Type I: Intraorbital: Primary orbital teratoma has its epicentre in the confines of the orbital bony pyramid. These noninvasive tumours grow with mechanical displacement of globe and expansion of orbital cavity. Since the growth is slow, the optic nerve is stretched slowly, preserving neurovisual transmission.

Type II: This type of presentation is rare. This type of orbital teratoma do not primarily arise from orbita, therefore they cause little or no proptosis. However, they tend to be more aggressive, extrude through the

lateral and/or medial wall, and completely destroying these structure.

Type III: This is an extremely rare variety. Intracranial orbital teratomas are more aggressive and spread as a retrograde extension into intracranial structure. They produce pressure through their space occupying nature. It also present as proptosis but at a milder form than type I.[8]

Teratoma accounts for almost 6.6% of childhood tumours, 80% of which occurs in females. The most common locations for teratomas are gonadal, sacrococcygeal (57%), retroperitoneal, and mediastinal regions. Teratoma of the head and neck constitutes 9% of all teratoma cases, 70-80% of them contain neural tissue and 20-40% remains undifferentiated or contains immature cells.

Teratomas are usually considered as benign tumours, although they grow rapidly, but very rarely malignant.[9-12] The presence of embryonal tissues or immature cells is supposed to be classified as malignant tertoma.[13] Some authors has suggested that the presence of endodermal and mesodermal features along with primitive neuro-ectodermal tissues may be classified and treated as benign teratoma.[14] While those with massive confluent growth of neuro-ectodermal tissues and little of other two germ cell layers may be considered as primary malignant teratoma.[14] Previous reported cases of malignant teratoma had also shown very different clinical as well as histological presentations.[9-12] one of these case was primarily intracranial with optic nerve involvement in a 19 year old male with, who had shown diffuse cerebrospinal fluid spread with metastasis in cervical and lumbar regions. In spite of giving chemotherapy and radiotherapy and even no recurrence of tumour on autopsy, patient died because of generalized metastasis.[9] Garden and McCanis has reported a case malignant cell population appeared 3 years after the excision of a benign orbital teratoma and consisted of cells believed to be malignant germ cells on histopathology.[11]

Orbital teratoma usually presents as severe unilateral proptosis or as orbitofacial asymmetry in healthy newborns. Orbital teratomas may remain dormant for years in adults and suddenly present as an acute painful proptosis due to hemorrhage or intracystic accumulation of secretion secondary to accidental trauma.[15,16] Unusual presentation like recurrent orbital cellulitis[17] have also been reported. They usually presents as a unilateral lesion, only one bilateral case has been reported.[13,18] The tumour can vary in size from small to extensive one.

There is no family history or no teratogenic influence of any drugs. It is more common in women and frequently described in left orbit (60%).[19-20] The number of reported cases is more commonly seen in whites than in blacks or Asians.[20,18] The eye is morphologically well developed, but vision potential is very low because of exposure keratopathy or optic nerve compression.[13] Rare associations like microcornea, corectopia with cataract and congenital colobomatous microphthalmic globe has also being reported.[16,21] The bony wall of orbit is expanded without any surrounding bony destructions. However Hassan HM et al in 2008 described a rare case of benign orbital tertaoma who had presented as extraconal mass with bony defects and malformed sphenoid bone.[17] Another patient of orbital teratoma associated with craniofacial anomaly (trigonocephaly) was reported by Sadove et al in 1991.[22] There is evidence of calcification and ossification in tumour mass on imaging.[13] Heterogenous structure with focal punctuate calcification and area of fat density are the characteristics features of orbital teratoma on imaging. Imaging helps in both delineation and characterization of the mass as well in planning for the surgical excision of the mass.[15] MRI has specific role in differentiating between teratoma and rhabdomyosarcoma.[23]

Differential diagnosis of orbital teratoma includes capillary haemangioma, lymphangioma, microphthalmos with cyst and malignant tumours like rhabdomyosarcoma, leukemia and neuroblastoma, dermoid cyst and meningoencephalocele.

Histopathologically teratoma contains all three germ cell layers. The most commonly observed germ cells in teratoma are surface ectoderm producing squamous epithelial lined cyst, hair follicles and sweat glands. Mesodermal layer is represented by the bone, muscle, cartilage and fat. Endoderm is least commonly noticed germ cell layer and it may produce gastro-intestinal tissue cyst lined by respiratory type pseudo-stratified columnar epithelium. This glandular tissue is responsible for the rapid progression of tumours. Differentiation between mature or immature teratoma can be made only histologically.

Laboratory tests include B- Human Chorionic Gonadotropin (B-HCG) and a-fetoprotein (AFP) levels. B-HCG is an indicator of testicular, embryonal or choriocarcionama.[3] AFP may correlate with diagnosis of malignant teratoma and their recurrence and response to treatment.[24]

Prenatal Ultrasound will probably be the most helpful future tool in diagnosis of orbital teratoma. Prenatal USG will also help the surgeons to plan the removal of tumour in a planned way under optimal conditions.

Management

In general, immediate surgical excision of tumour as soon as the diagnosis is made is recommended. In the past, complete excision with exteneration was most acceptable surgical option for orbital teratoma, but incomplete excision can cause recurrence of local tumour.[15,25-26]

However, number of cases is being reported in literature where exenteration was deferred and

maximum possible orbital and ocular tissues were successfully preserved with only enbloc removal of mass for good cosmesis. Early surgical intervention can preserve vision also to some extent. However teratoma with involvement of optic nerve and exposure keratopathy, extreme proptosis and absent papillary reflex present as a challenge for the cosmetic correction.

P J Damato and F J Damato described a case, where almost 3 cm size of orbital teratoma was removed in toto and eyeball was preserved in a newborn baby.[27]

Jaychandran Sadaksharan had described a case of recurred orbital teratoma, which was successfully removed in toto without compromising globe.[15]

Jared J Mee reported a case, where teratoma had late presentation with good postoperative visual outcome.[28] Lawerence Gnanaraj et al reported a case with massive congenital teratoma was successfully removed by eye lid sparing extenteration technique.[29]

Cristina Gonzalez reported a case of massive congenital teratoma of approximately 6 cm in size on MRI in a healthy new born baby which was successfully removed and ocular prosthesis was applied for cosmetic purpose.[30]

The prognosis of teratoma depends upon the age of presentation, site and nature of tumour (benign or malignant). The prognosis is usually good if removed completely in early age.[31] All teratoma should be followed up regularly to look for any recurrence or malignant transformation.[32] Residual or recurrent teartoma with or without malignant feature requires chemotherapy and radiotherapy.[33]

References

1. GROSS RW, CLATWORTHY HW Jr, MEEKER IA Jr. Saccrococcygeal teratoma in infants and children: report of 40 cases. Surg Gynecol Obstet. 1951Mar;92(3);341-354.
2. Holmes T. Congenital tumor removed from the orbit. Trans. Pathol. Soc. London. 1863;14:248–248.
3. Grosfeld JL, Billmire DF. Teratomas in infancy and childhood. Curr Prob Cancer. 1985 Sep;9(9):1-53.
4. Mizuo G. Eine seltene from von teratoma orbitae. Arch Augenheilkd 1910; 65:365-383.
5. Cockerham KP, Bejjani GK, Kennerdell JS, Maroon JC. Surgery for orbital tumors. Part II: transorbital approaches. Neurosurg focus. 2001 May 15;10(5):E3.
6. Harley RD,pediatric ophthalmology, Vol 2, 2'nd ed, Philadelphia. WB, Saunders. 1983:1223-1271.
7. Kivela T, Tarkkanen A. Orbital germ cell tumours revisited: a clinicopathological approach to classification. Surv Ophthalmol 1994 May-Jun;38(6):541–54.
8. Habal MB. Orbital teratomas: a new approach for surgical treatment. J Craniofac Surg 1990 Jan;1(1):8-14.
9. Samii M, Ramina R, Koch G, Reusche E. Malignant teratoma of the optic nerve: case report. Neurosurgery. 1985 May;16(5):696-700.
10. SARADARIAN AV. Malignant teratoma of the orbit: six and one half year's observation. Arch Ophthal. 1947 Feb;37(2):253.
11. Garden JW, McCanis J.C. Congenital orbital-intracranial teratoma with subsequent malignancy: Case report. Br J Ophthalmol. 1986;70(2):111-3.
12. Soares EJC, Lopar K da S, Andrade J de S, Fa-Leiro LC, Alves JCR. Orbital malignant teratoma; a case report. Orbit. 1983;2:235-242.
13. Levin ML, Leone CR Jr, Kincaid MC. Congenital orbital teratomas. Am J Ophthalmol. 1986 Oct 15;102(4):476–81.
14. Kleinman GM, Young RH, Scully RE. Primary neuroectodermal tumours of the ovary. A report of 25 cases. Am J Surg Pathol. 1993 Aug;17(8):764-78.
15. Orbital teratoma: Radiologist's perspective-Jayachandran Sadaksharam, Khushboo Singh. J of cranio-maxillary diseases. July.2014, vol3;2.
16. M singh et al. Orbit. orbital teratoma with tooth in an adult; rare association with cataract and corectopia. Orbit, 2013; 32(5): 327–29.
17. Hassan HM, Mc Andrew PT, Yagan A, et al. Mature orbital teratoma presenting as a recurrent orbital cellulitis with an ectopic tooth and sphenoid malformation. Orbit 2008;27:309–312.
18. Chang DF, Dallow RL, Walton DS. Congenital Orbital Teratomas: report of a case with visual preservation. J Pediatr Ophthalmol Strabismus 1980; Mar-Apr;17(2):88-95.
19. Herman TE, Vachharajani A, Siegel MJ. Massive congenital orbital teratoma. J Perinatol.2009;29:396-7.
20. Alkemade PP. Congenital teratoma of the orbit. Ophthalmologica. 1976; 173(3-4):274–85.
21. Singh U, Subramanian A, Bal A. Congenital orbital teratoma presenting as microphthalmos with cyst. Indian J ophthalmol. 2009 Nov-Dec;57(6):474-5.
22. Sadove AM, Kalsbeck JE, Ellis FD, Eppley BL, Elluru R. Orbital teratoma associated with trigonocephaly. Plast Reconstr Surg 1991 Dec;88(6):1059-63.
23. De Potter P, Shields JA, Shields CA,editors. MRI of the Eye and Orbit. J.B Lippincott company; Philadelphia, PA:1995. p.304.
24. Tsuchida Y, Saito S, Ishida M, Omi K, Urano Y. Yolk sac tumour (endodermal sinus tumour) and alpha-fetoprotien: A report of three cases. Cancer. 1973 Oct;32(4):917-21.
25. Mamalis N, Garland PE, Argyle JC,Apple DJ. Congenital orbital teratoma: a review and report of two cases. Surv Ophthalmol 1985 Jul-Aug;30(1):41-6.
26. Kearney JA. True filial teratoma and cyst in the orbit of newborn child. Am J Ophthalmol.1926;9:416-426.
27. NEONATAL ORBITAL TERATOMA. Damato PJ and Damato FJ. Br J Ophthalmol.1962 Nov;46(11):685-91.
28. Jared J Mee, Penny A McKelvie and Alan A McNab FRANZCO. Orbital teratoma: late presentation with normal vision. Clinical and Experimental Ophthalmology. 2002 Feb;30(1):41–43.
29. Lawrence Gnanaraj, Skibell, Bentley C, Coret-Simon, Judithet, Halliday, William, Forrest, Christophe, DeAngelis, Dan. Massive Congenital Orbital Teratoma. Ophthalmic Plastic and Reconstructive Surgery. 2005 Nov;21(6);445-470.
30. Cristina González, Carlos A. Restrepo, Gloria I. Salazar, Patricia Monsalve, M. Congenital orbital teratoma. Colomb Med. 2012;43:82-85.
31. Gonzalez-Crussi, F. in: Extragonadal Teratomas. Armed Forces Institute of Pathology, Washington; 1982:1–49 (100-1008. (Atlas of Tumor Pathology, 2nd ser, fasc. 18).
32. Maurya R.P, Singh V.P, Prashant B, Singh M.K, Kumar M. Congenital orbital teratoma with bilateral

anophthalmia: A rare presentation. World J. Pathol 2012;1:35-38.
33. Tuncbay E, Ovul I, Oztop F. Orbito-cranial teratoma. A case report. Acta Neurochir (Wien) 1981;56(1-2):73-80.