Content available at: https://www.ipinnovative.com/open-access-journals

IP International Journal of Ocular Oncology and Oculoplasty

Journal homepage: https://ijooo.org/

Case Report

Granular cell tumor: A rare presentation as intraconal orbital space occupying lesion

Malarvizhi Raman[1], Anuradha Alagusundaram[2], Keerthana Athiaman[3,*]

[1]*Sri Muthukumaran Medical College Hospital and Research Institute, Chennai, Tamil Nadu, India*
[2]*Madurai Medical College, Madurai, Tamil Nadu, India*
[3]*Dept. of Oculoplasty, Regional Institute of Ophthalmology, Egmore, Chennai, Tamil Nadu, India*

ARTICLE INFO

Article history:
Received 16-05-2023
Accepted 15-06-2023
Available online 08-08-2023

Keywords:
Granular cell tumor
Benign tumor
S 100 positivity CD 68 positivity

ABSTRACT

Granular cell tumor (GCT) is a rare benign soft tissue tumor. They have been reported in several regions of the body but mostly in the skin and subcutaneous tissue of head and neck. It may involve the orbit, periocular skin, lacrimal sac, optic nerve, ciliary body, conjunctiva and caruncle.
Here we present a rare case of intraconal GCT in a 45 year old woman who presented with unilateral proptosis. She presented with forward protrusion of right eye for the past 3 months,
MRI orbit revealed a well defined T1/T2/FLAIR hypointense lesion in the inferolateral aspect of intraconal compartment of right orbit. Transconjunctival excision of the lesion was done. Histopathological Examination (HPE) and immunohistochemistry confirmed the diagnosis of Granular cell Tumor.

1. Introduction

Granular cell tumor is usually a benign soft tissue lesion of neural origin, first described in 1926 in the tongue. Immunohistochemistry and ultrastructural studies have identified GCT to originate from Schwann cells. It affects people over a wide range of age with a slight female preponderance and a peak incidence in the fourth to sixth decades of life. Head and neck regions are the most affected sites with over two third cases diagnosed in the tongue. GCT in the orbit is very rare.[1,2]

2. Case Report

A 45 year old female came with complaints of forward protrusion of right eye and headache for 3 months. Patient was a known case of type 2 diabetes mellitus on treatment. General physical examination and systemic examination were insignificant. Vitals were stable on examination. Detailed ophthalmological examination was performed. Visual acuity with Snellen's chart was 6/12+1 DSph 6/6 in right eye (RE) and 6/12+1Dcyl *150 6/6p in left eye (LE). Intraocular pressure with Goldmann applanation tonometry was 20 mmHg in both eyes. Colour vision with Ishihara pseudoisochromatic plates was found to be normal in both eyes. Contrast sensitivity with Pelli-Robson was found to be within normal limits in both eyes. Pupils 3 mm in both eyes and equally reacting to light. Orthoptics revealed Right divergent squint to alternate divergent squint, with exotropia of 60 prism diopters. In Worth four dot test BSV was absent. Extraocular movements in 9 gazes showed elevation restriction in the RE. Proptosis evaluation of the right eye revealed an axial proptosis with inferior scleral show and resistance to retropulsion with no corkscrewing of vessels. Hertel's exophthalmometry with a baseline of 101 showed RE-23 mm and LE 20 mm. Fundus examination by indirect ophthalmoscopy revealed

* *Corresponding author*.
E-mail address: keerthanaathiaman@gmail.com (K. Athiaman).

https://doi.org/10.18231/j.ijooo.2023.017

mild temporal pallor in both eyes. A provisional diagnosis of right orbital mass under evaluation was made and proceeded with investigations. B scan ultrasound with 10 MHZ in the RE showed a well defined encapsulated lesion measuring 2*1.6 cm filled with moderate intense echoes abutting the optic nerve in the inferior quadrant. No calcification was noted. Routine Blood work up was within normal limits. MRI orbit showed a well defined T1/T2/FLAIR hypointense lesion in the infero-lateral aspect of intraconal compartment of right orbit showing mild restricted diffusion. The lesion was found to compress the inferior rectus muscle with mild displacement of the optic nerve superomedially. The lesion also showed moderate homogenous enhancement with intravenous contrast with no adjacent bony erosion. After obtaining anesthetic fitness, under general anesthesia, by inferior transconjunctival orbitotomy, total excision of the intraconal lesion was done and sent for HPE. HPE under low power microscopy revealed a mass composed of round to polygonal cells with abundant eosinophilic cytoplasm with eosinophilic cytoplasmic granules and centrally placed round nuclei. The stroma showed focal lymphoid aggregates suggestive of GCT. Specimen was sent for IHC for confirmation of diagnosis. Immunohistochemistry showed S-100-diffuse positive in 100% tumor cells and CD-68 positive in 100% tumor cells in favor of GCT. Postoperatively patient was started on IV antibiotics, tablet prednisolone 30 mg once daily and topical antibiotic eye drops. Patient did well in the postoperative period and was asked to review weekly at ophthalmology outpatient department.

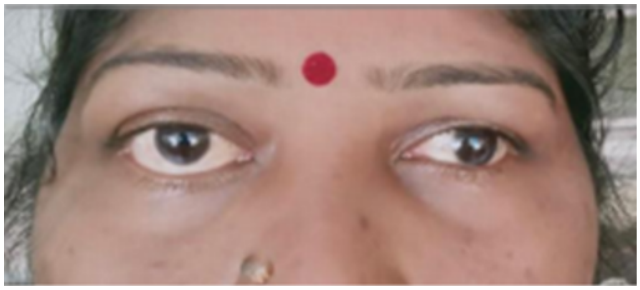

Fig. 1: Pre-operative proptosis

3. Discussion

GCT is a benign soft tissue lesion. Malignant GCTs are extremely rare, accounting for less than 2% of all GCTs.[1,3] It was first described by Abrikossoff in 1926 as a granular cell myoblastoma in the tongue and was thought to originate from mature striated muscle cells.[4] Later, GCTs were found to be of neural origin according to various histochemical and ultrastructure studies. Most patients are middle-aged, with a peak incidence in the fourth through the sixth decades of life. A slight female predominance exists, with an estimated female-to-male ratio

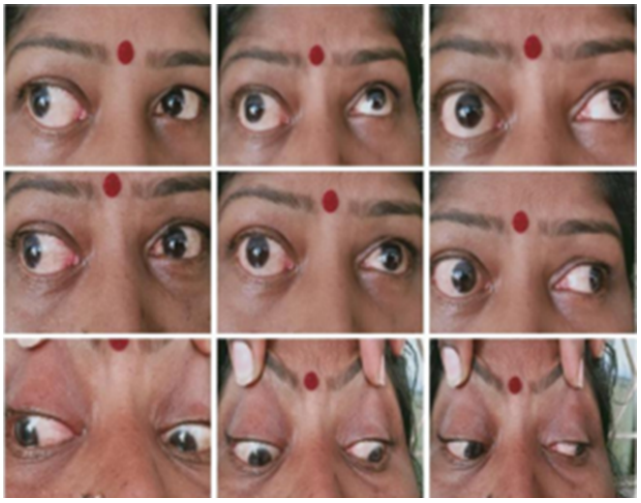

Fig. 2: Ninegaze showing elevation restriction

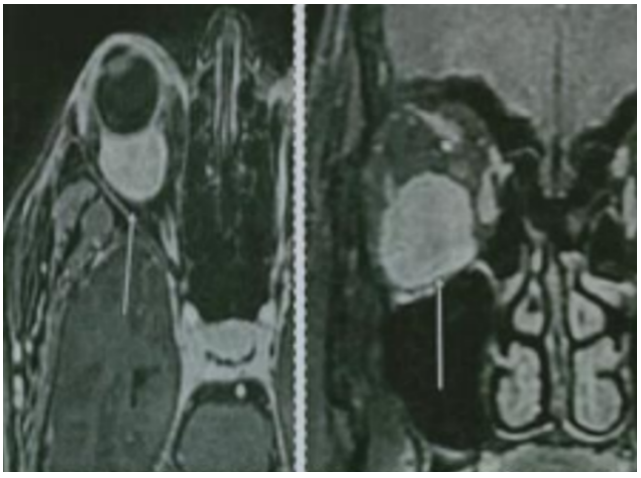

Fig. 3: Masscompressing inferior rectus and displacing optic nerve superomedially

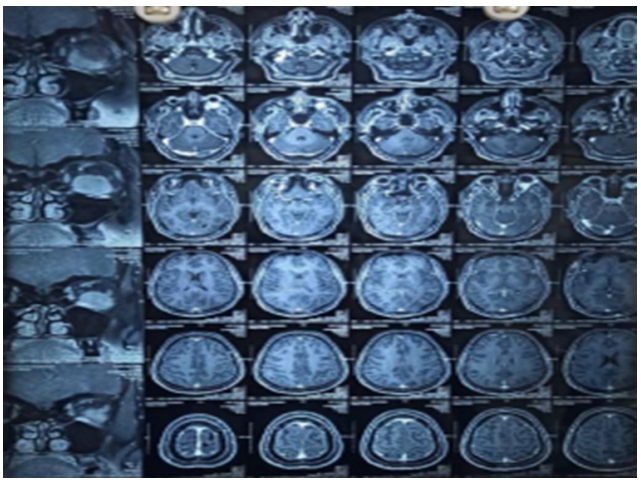

Fig. 4: Intraconalmass in right orbit t1/t2/ flair hypointense and diffusion restriction.

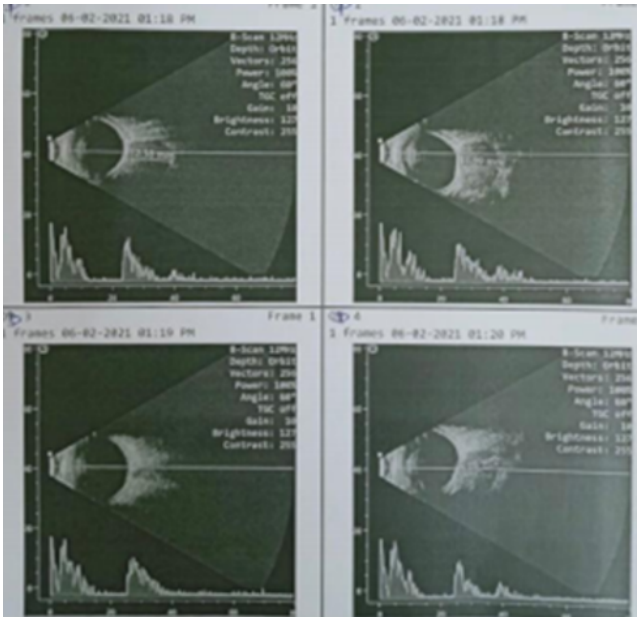

Fig. 5: Well defined encapsulated lesion of size 2x1.6 cm filled with moderate reflective echoes abutting optic nerve in inferior quadrant

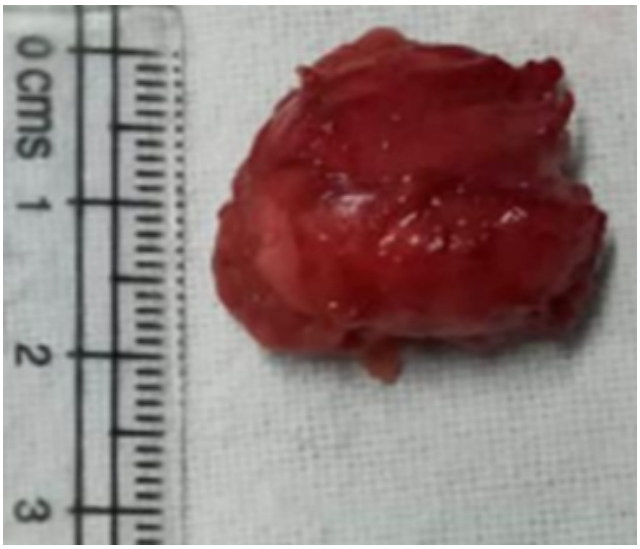

Fig. 6: Excised tumor

of approximately 3:2. According to studies, up to 10% of granular cell tumors are multiple[5] and 10 to 15 % of GCTs were found to be multicentric. A review by Ordóñez and Mackay[6]concluded that GCT is most frequently located in the head and neck area and that about one-fourth of the lesions occurred in the tongue. Other reported sites are the larynx, parotids, breast, esophagus, respiratory, gastrointestinal, and genitourinary tract[7] In ocular adnexa,[8] GCT has been reported to occur in the orbit, extraocular muscles, caruncle, eyelid skin, palpebral conjunctiva, and

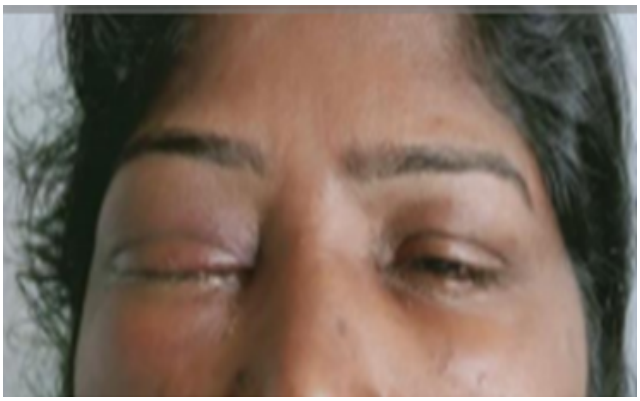

Fig. 7: Immediate post-operative picture

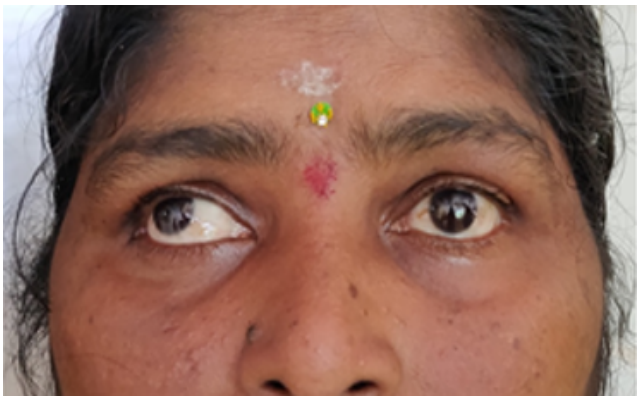

Fig. 8: Late post-operative picture

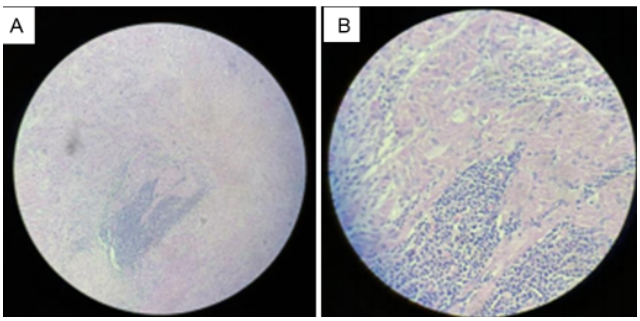

Fig. 9: A: Low power 10 X; **B:** High power 40 X

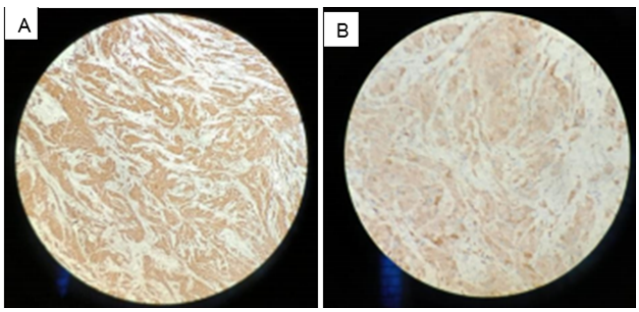

Fig. 10: A: S100 Positivity; **B:** CD 68 Positivity

lacrimal drainage apparatus[2] and furthermore GCT in the orbit was found to be rare. The most common complaints of orbital GCT patients are diplopia, proptosis, and restriction of mobility[1] Malignant orbital GCT was scarcely reported. For determining the malignancy of the tumor as early as possible and to avoid metastasis, histological specimen examination and the rate of tumor progress can be two indicative parameters.[9] A slowly developing tumor can be suggestive of a benign tumor and a majority of GCTs are usually benign. Immunohistochemistry and ultrastructural studies have identified GCT to originate from Schwann cells.[10] In histology, GCT cells feature acidophilic granular cytoplasm packed with lysosomes.

4. Conflict of Interest

None.

5. Source of Funding

None.

References

1. Li XF, Qian J, Yuan YF, Bi YW, Zhang R. Orbital granular cell tumours: clinical and pathologic characteristics of six cases and literature review. *Eye (Lond)*. 2016;30(4):529–37. doi:10.1038/eye.2015.268.
2. Salour H, Tavakoli M, Karimi S, Kanavi MR, Faghihi M. Granular cell tumor of the orbit. *J Ophthalmic Vis Res*. 2013;8(4):376–9.
3. Rejas RAG, Campos MS, Cortes AR, Pinto DDS, Desousa M. The neural histogenetic origin of the oral granular cell tumor:an immunohistochemical evidence. *Med Oral Patol Oral Cir Bucal*. 2011;16(1):6–10. doi:10.4317/medoral.16.e6.
4. Abrikossoff A. *Über Myome, ausgehend von der quergestreiften willkürlichen Muskulatur*. 1926;260:215–233.
5. Said-Al-Naief N, Brandwein M, Lawson W, Gordon R, Lumerman H. Synchronous lingual granular cell tumor and squamous carcinoma: A case report and review of the literature. *Arch Otolaryngol- Head Neck Surg*. 1997;123(5):543–7.
6. Jaeger MJ, Green WR, Miller NR. Granular cell tumor of the orbit and ocular adnexa. *Surv Ophthalmol*. 1987;31(6):417–23. doi:10.1016/0039-6257(87)90033-6.
7. Ribeiro SF, Chahud F, Cruz AA. Oculomotor disturbances due to granular cell tumor. *Ophthal Plast Reconstr Surg*. 2012;28(1):23–7. doi:10.1097/IOP.0b013e3182141c54.
8. Noonan JD, Horton CE, Old WL, Stokes TL. Granular cell myoblastoma of the head and neck:review of the literature and 10 year experience. *Am J Surg*. 1979;138(4):611–4.
9. Callejo SA, Kronish JW, Decker SJ, Cohen GR, Rosajr H. Malignant granular cell tumor metastatic to the orbit. *Ophthalmology*. 2000;107(3):550–4.
10. Jardines L, Cheung L, Livolsi V, Hendrickson S, Brooks J. Malignant granular cell tumors:report of a case and review of the literature. *Surgery*. 1994;116(1):49–54.

Author biography

Malarvizhi Raman, Professor and HOD

Anuradha Alagusundaram, Associate Professor

Keerthana Athiaman, Post Graduate

Cite this article: Raman M, Alagusundaram A, Athiaman K. Granular cell tumor: A rare presentation as intraconal orbital space occupying lesion. *IP Int J Ocul Oncol Oculoplasty* 2023;9(2):87-90.