

GRANULAR CELL TUMOR OF ORBIT

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INTRODUCTION

- Granular cell tumor is rare benign soft tissue tumor that may involve orbit, periocular skin, lacrimal sac, optic nerve, ciliary body, conjunctiva & other non ocular tissues.

- Reference-. Salour H, Tavakoli M, Karimi S, Rezaei Kanavi M, Faghihi M. Granular cell tumor of the orbit. J Ophthalmic Vis Res. 2013 Oct;8(4):376-9. PMID: 24653826; PMCID: PMC3957045.

53 year old lady, Resident of Pimpri, Pune came to the ophthalmology OPD

- LEFT EYE:
 - Diminution of vision since 1 year
 - Swelling in Left eye upper eyelid since 1 year



- Patient was apparently alright 1 year back when she developed diminution of vision in left eye which was insidious in onset, gradual, progressive and painless in nature.
- It was associated with fullness in left upper eyelid which was insidious in onset, gradual, painless and progressive in nature.
- H/O spectacles use for distant as well as near since 10 years

- No h/o of double vision.
- No h/o fever, Upper respiratory tract infection, headache
- No h/o postural variation, increase in size with coughing or sneezing
- No h/o ocular trauma /surgery
- No h/o any treatment taken in the past

- No known systemic illness

- Sleep-Adequate.
 - Appetite- Mixed diet, Normal.
 - Bowel & Bladder habits: Regular.
 - No addictions.
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- FAMILY HISTORY:- Nothing significant.

- Patient is conscious, cooperative & well oriented to time, place and person.
- Temperature- afebrile.
- Pulse- 84 beats/ minute.
- BP- 130/80 mm Hg
- RR- 18/minute
- No Pallor, icterus, cyanosis or edema noted.
- Lymph node examination-Preauricular, Postauricular, Parotid, submandibular, cervical were non-palpable

- Respiratory System-WNL
- Cardiovascular System-WNL
- Central Nervous System-WNL
- Gastrointestinal system-WNL

OCULAR EXAMINATION

	RIGHT EYE	LEFT EYE
Vision	6/18	6/24
BCVA	6/6 (+1.00/ -0.50 x 120*)	6/12(+1.75 DS)
Near vision Colour vision	N6 (+1.75 DS) Intact	N6 (+1.75 DS) Intact
Extraocular movements	Duction & Versions Full,free ,painless	Duction & Version full,free painless
Head posture and facial symmetry	Normal	Normal
Eyebrows	Normal	Normal
Eyelids	Normal	Discussed on later slide
Eyelashes	Normal	Normal
Orbital margins	Continuous, non-tender	Continuous, non-tender



Inspection

- Nafzaiger's sign & Worm's eye view –ve.
- No Retraction of lid or ptosis noted
- Diffuse swelling present over lateral part of upper eyelid
- Skin over lid appears normal with no redness, pigmentation, venous dilatation, pulsation
- No change with valsalva, postural variation noted



Palpation

- Orbital margins intact
- 4x4mm hemispherical mass with ill defined posterior margins seen, no local rise in temperature, non tender, firm in consistency,transillumination negative, non-Pulsatile with no thrill noted.
- Retropulsion negative
- **Auscultation**
No Bruit heard



- **EXOPHTHALMOMETRY**
done by Hertel's
Exophthalmometer-

- **With base at 110mm**
- (RE) 18 mm
- (LE) 19 mm



ANTERIOR SEGMENT EXAMINATION

	RIGHT EYE	LEFT EYE
Conjunctiva	WNL	WNL
Cornea	Clear	Clear
Anterior Chamber	Normal Depth	Normal depth
Iris	Normal Pattern	Normal Pattern
Pupil	C/C/Reactive to light (No RAPD)	C/C/Reactive to light (No RAPD)
Lens	Grade I nuclear sclerosis	Grade 1 Nuclear sclerosis
Intra-ocular pressure (applanation tonometry)	16 mmhg	16 mmhg

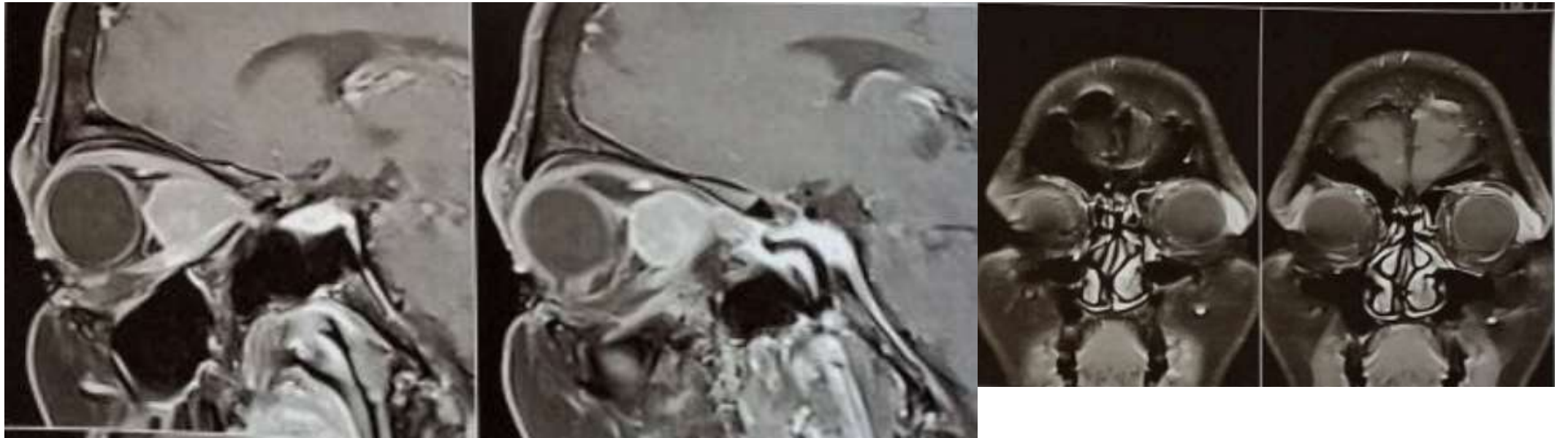
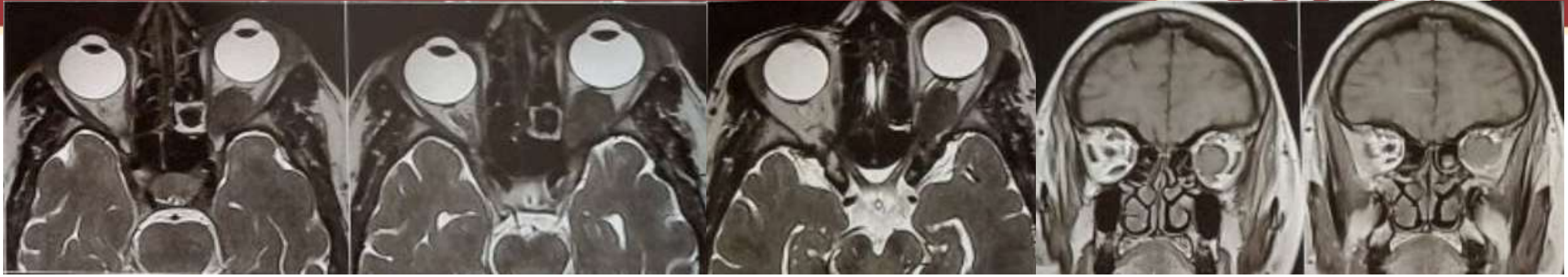
	RIGHT EYE	LEFT EYE
MEDIA	Clear	Clear
OPTIC DISC	Normal size, shape, color, margins with A:V ratio 2:3	Normal size, shape, color, margins with A:V ratio 2:3
CUP DISC RATIO	0.4	0.4
NEURORETINAL RIM	Healthy	Healthy
MACULA	WNL	WNL
FOVEAL REFLEX	+	+
GENERAL FUNDUS	WNL	WNL

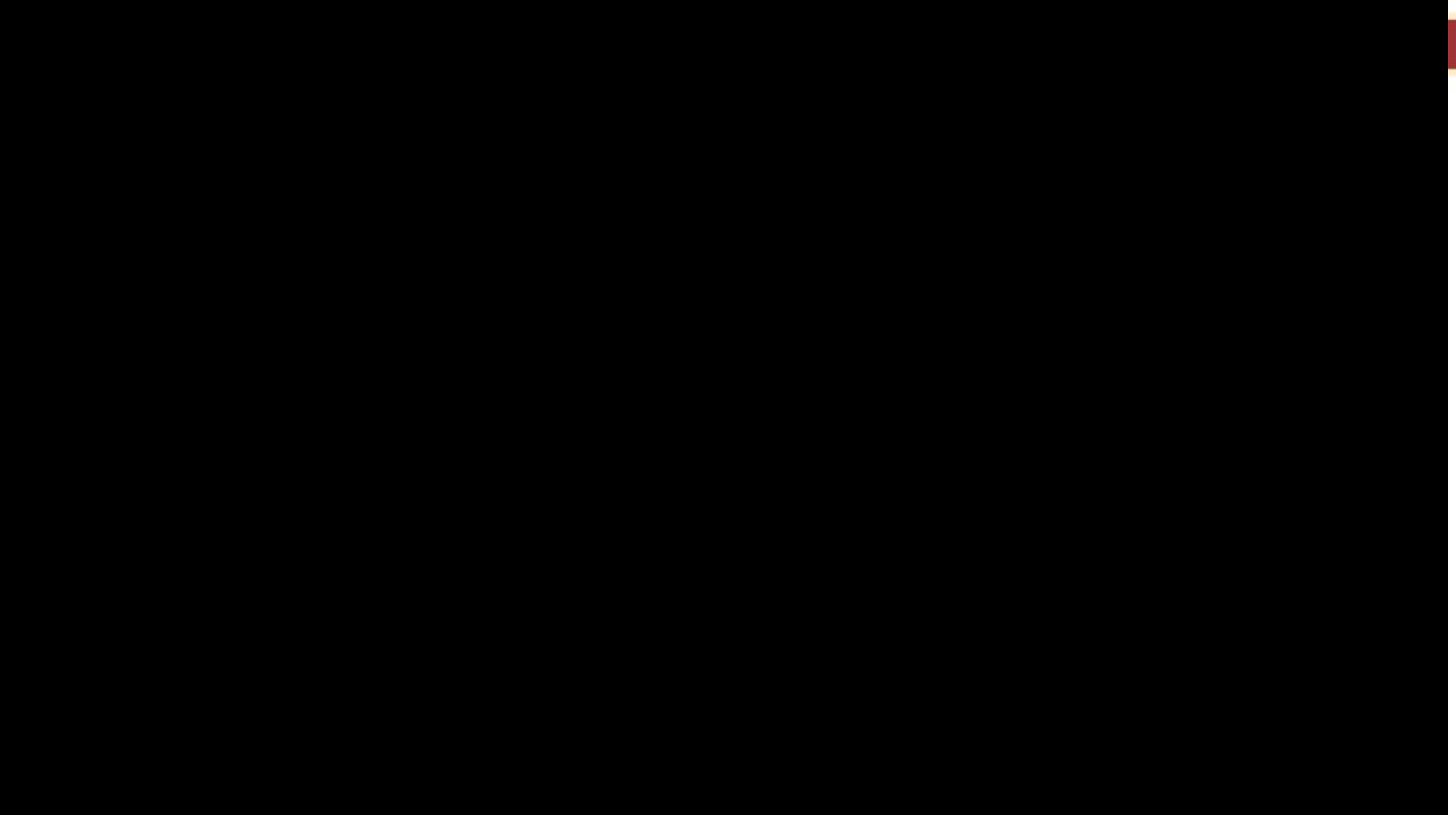
PROVISIONAL DIAGNOSIS

- (LE)Lacrimal Gland tumour
- PLAN OF MANAGEMENT-
- Sx-(LE) Lateral Orbitotomy with mass excision .

- **Left eye-**
- Hypoechoic lesion seen in retroorbital intraconal compartment of left eye.
- Left optic nerve not visualised separately .



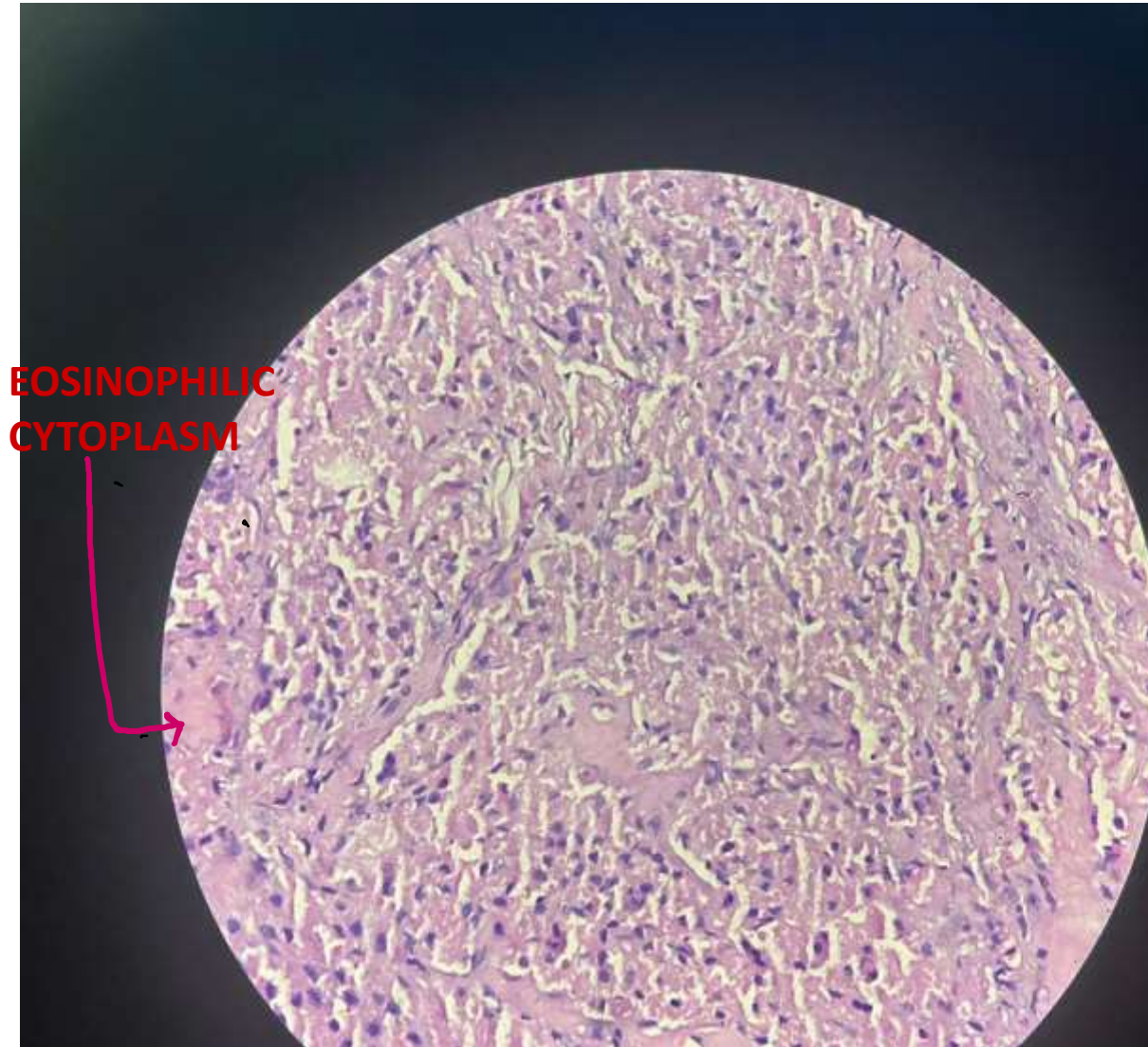


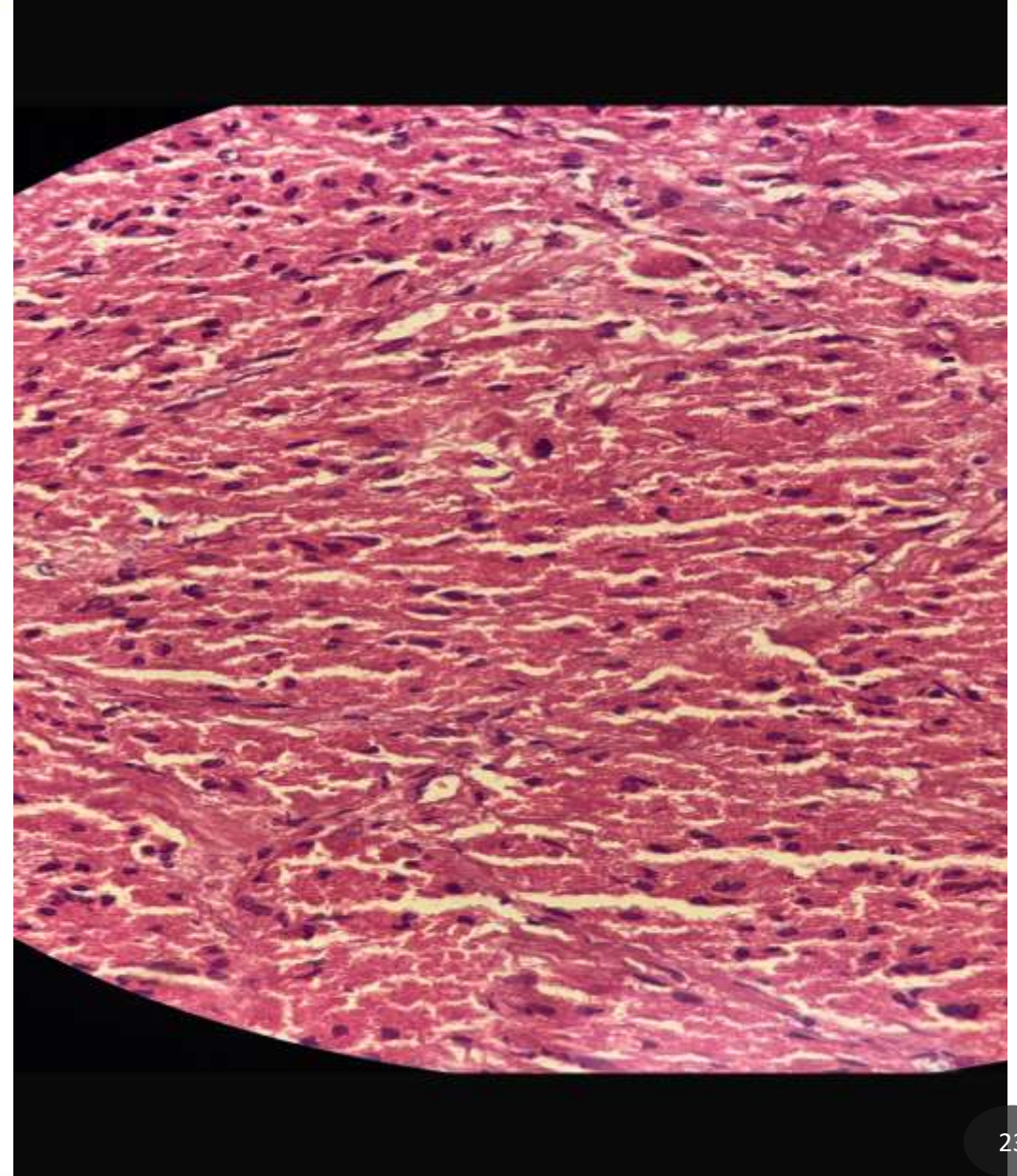
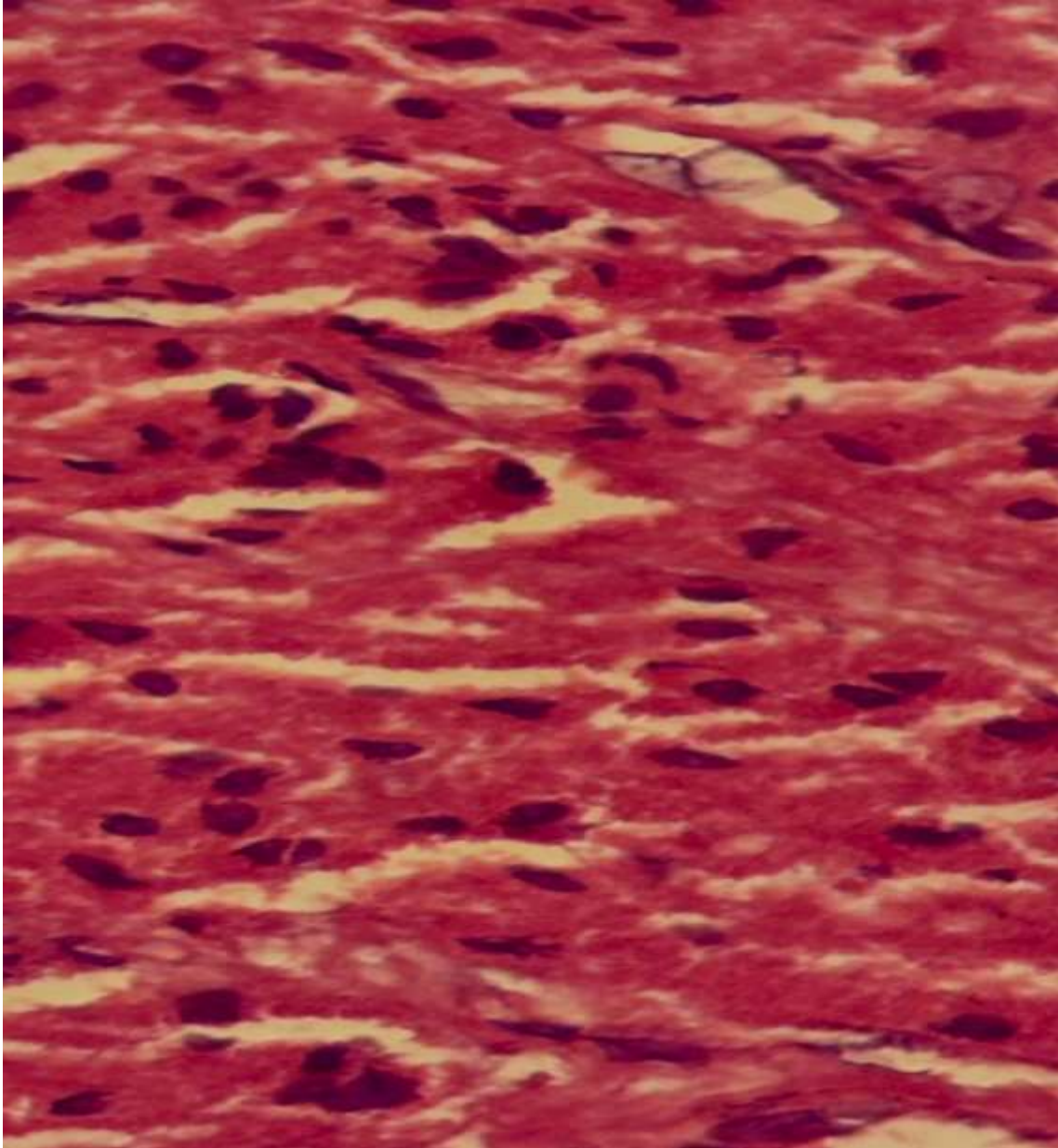


HPE of Intraconal mass

H and E stained sections show ill defined lesion of polygonal cells arranged in sheets and nests separated by thin collagenous septae. The cells are polygonal shaped with indistinct cell margin. Cells are having small nuclei and abundant granular eosinophilic cytoplasm. Also noted are dense lymphoid aggregates surrounding the tumor. No evidence of mitotic activity or necrosis or haemorrhage within the tumour.

Suggestive of Granular cell tumor



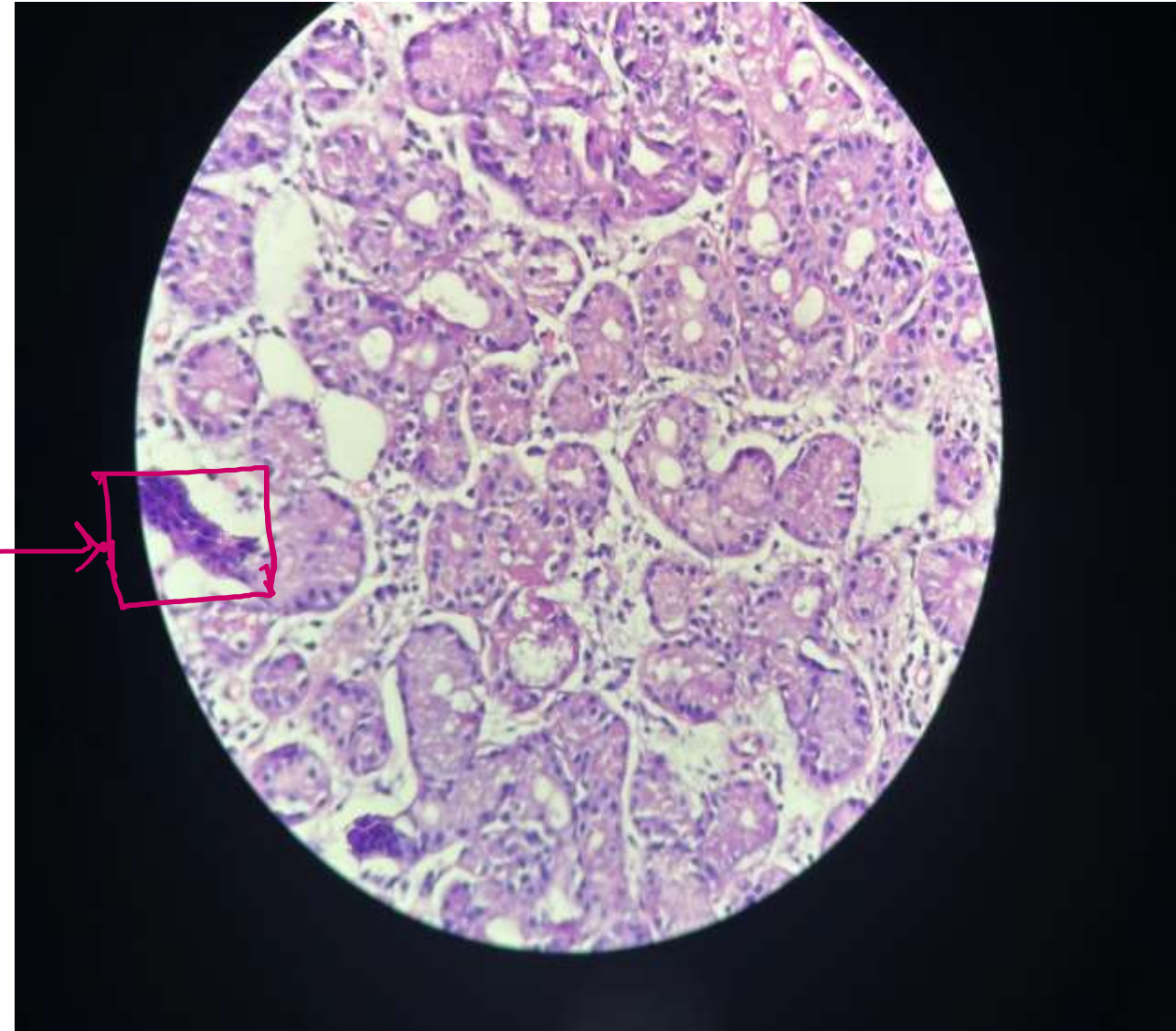


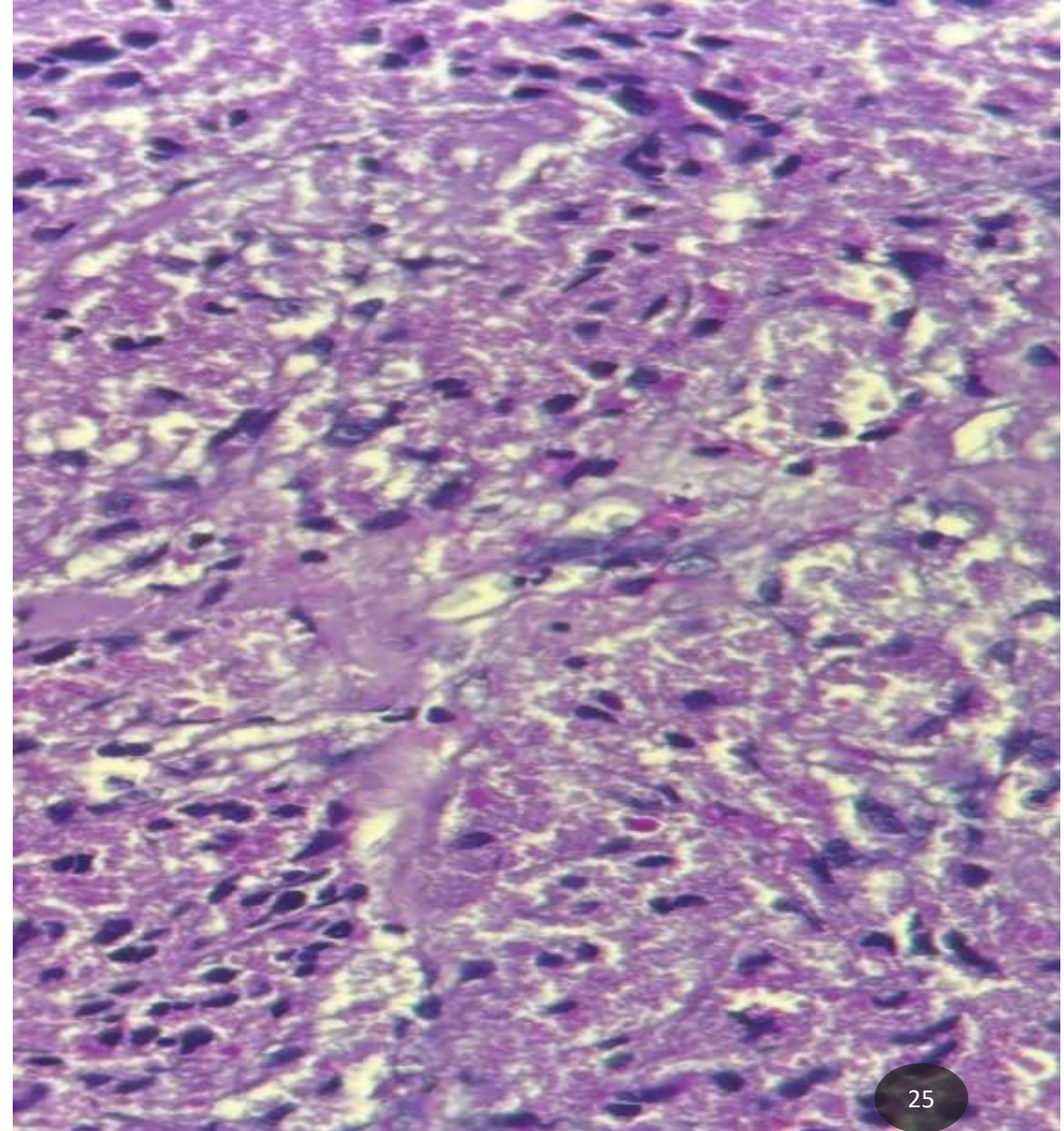
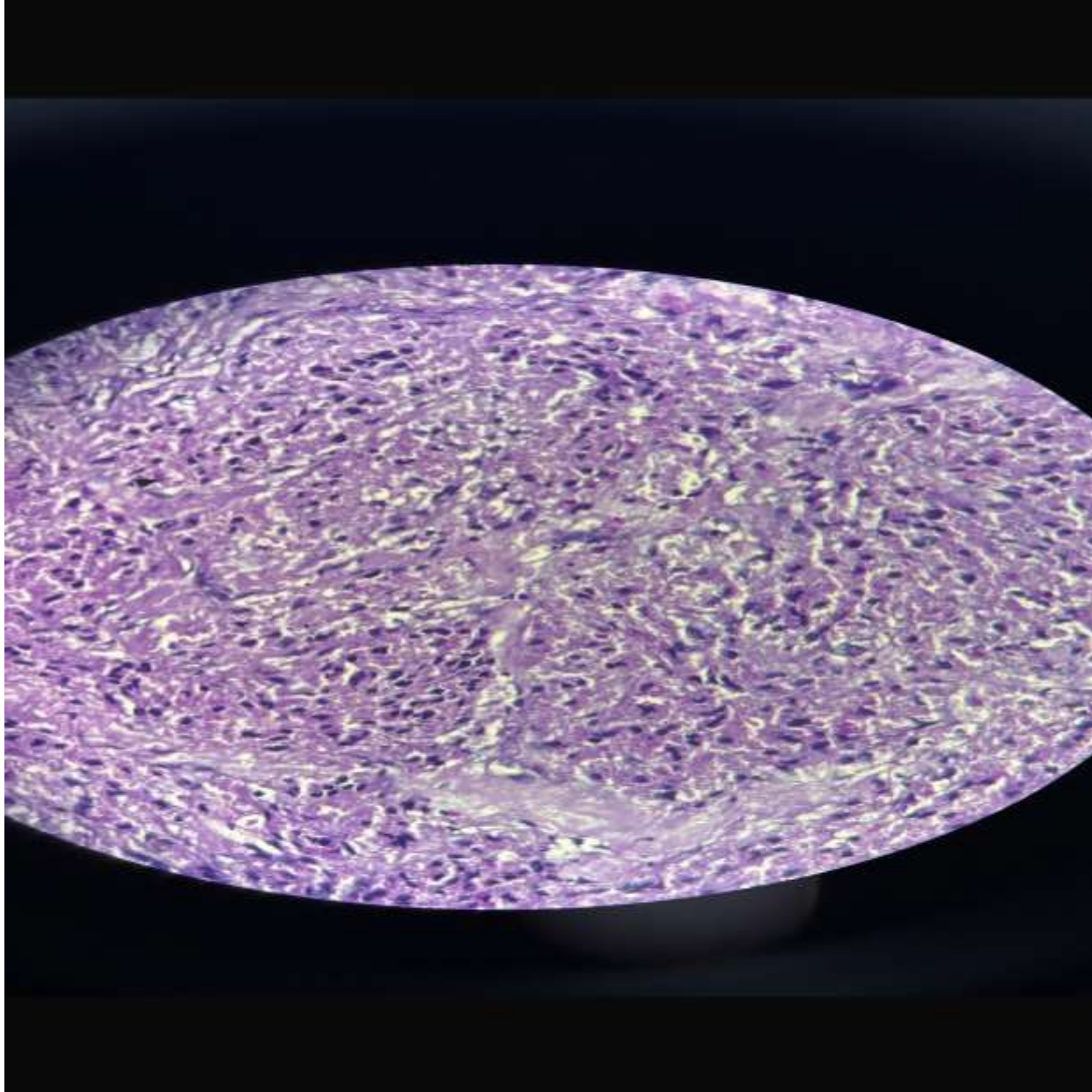
HPE of LACRIMAL GLAND

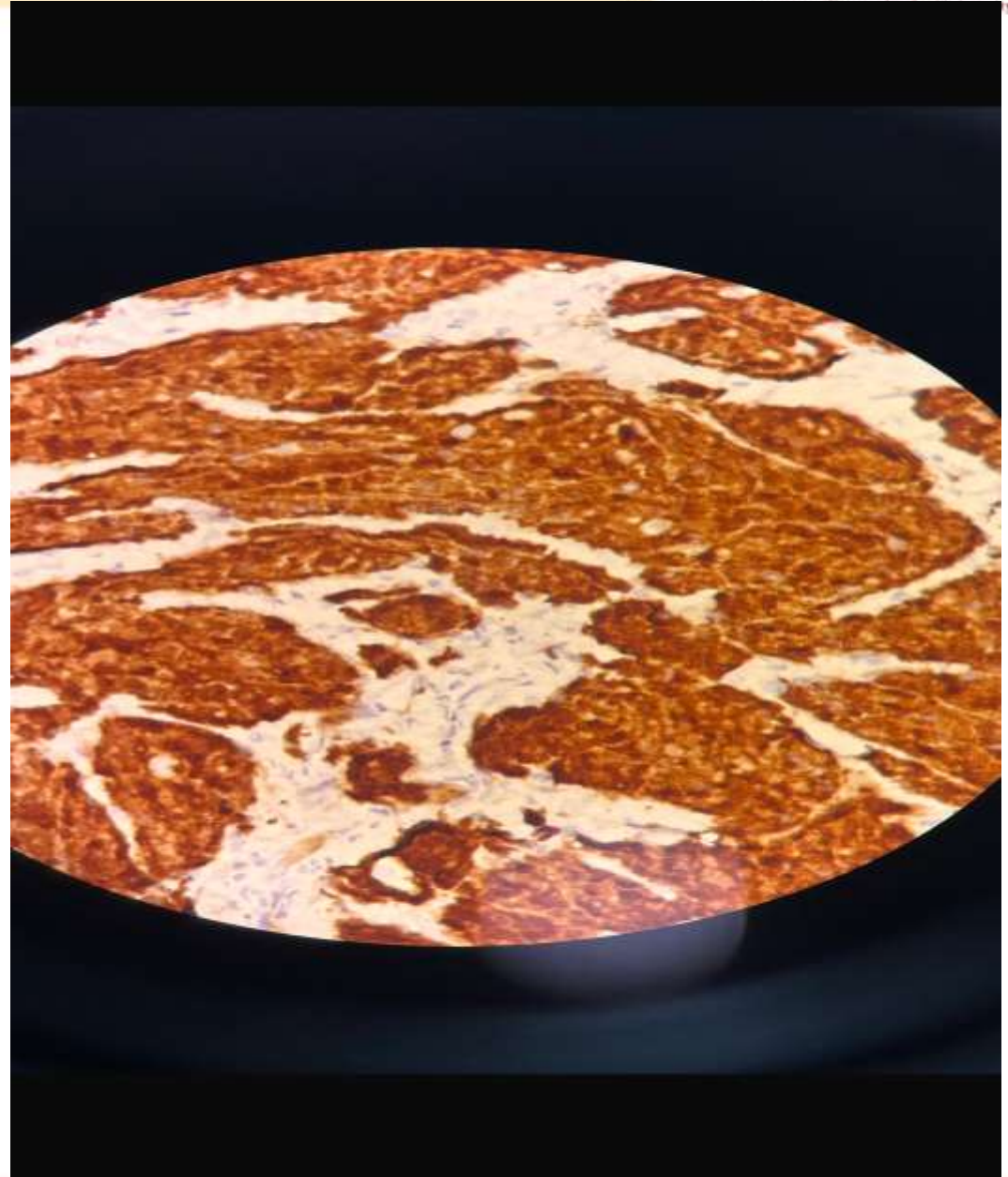
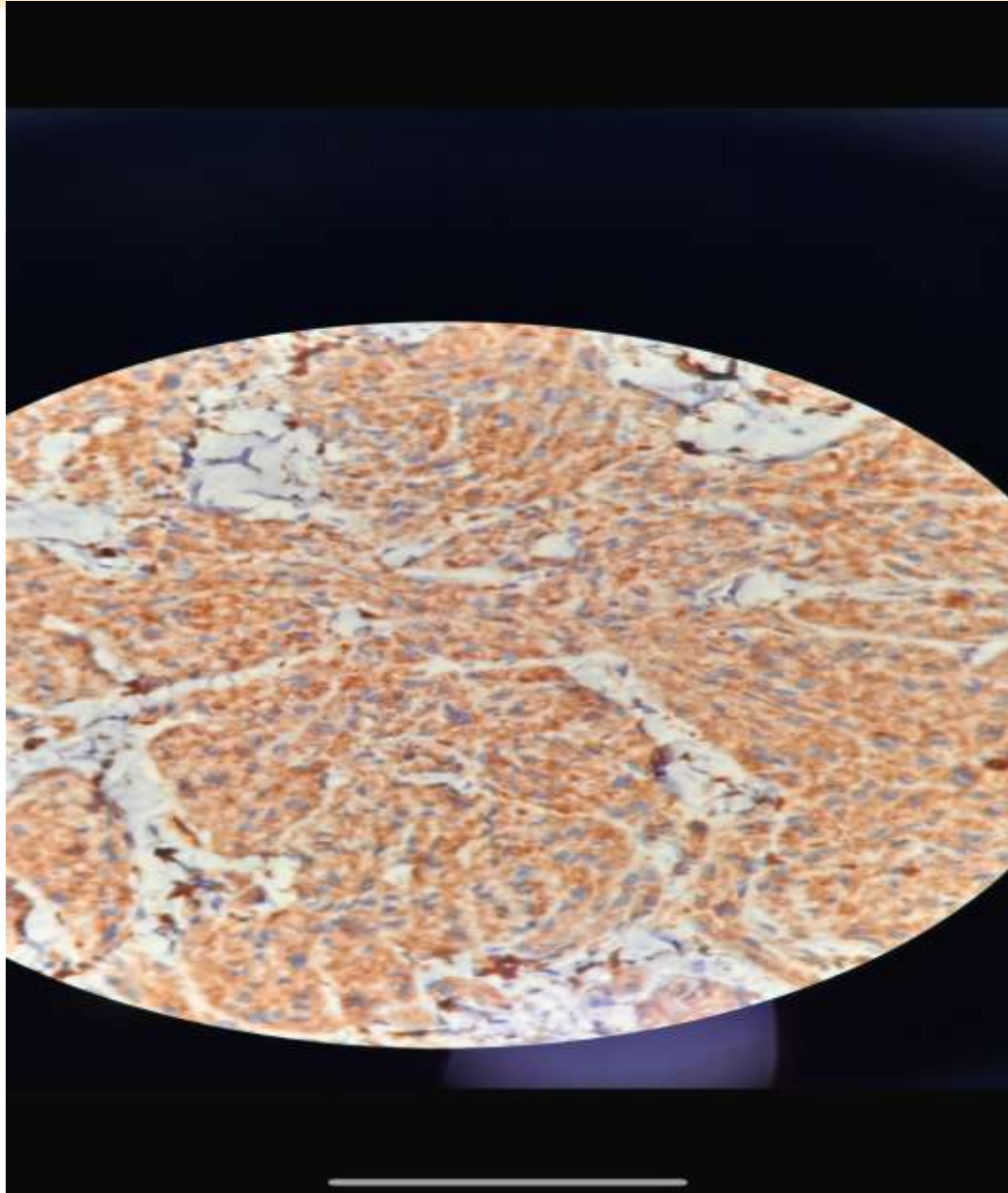
Section shows seromucinous glands surrounded by mild chronic inflammatory infiltrate. Focal lymphoid aggregates seen.

Suggestive of Seromucinous glands with chronic inflammatory pathology.

- LYMPHOID AGGREGATES







IHC MARKER

- S100-Positive
- CD68-Positive

(LE) Granular Cell Tumor Of Orbit

Plan of management- Referred to Medical Oncologist .

Granular cell tumors are rare neoplasms derived from Schwann cells with characteristic pathologic findings. They can exhibit either benign or malignant behavior and most commonly occur in the oral cavity, skin, and gastrointestinal tract. [1]

EPIDEMIOLOGY

Granular cell tumors can appear in all age groups, most commonly in the 4th to 6th decades. These tumors are most often seen in women. [2][3]

ETIOLOGY

Etiology of granular cell tumour is unknown.

TREATMENT

SURGICAL

Complete excision to negative margins with close clinical follow-up is recommended for granular cell tumors, whether benign or malignant.^[3]

CHEMOTHERAPY

There is a limited role for chemotherapy and radiation therapy.

THIS CASE IS PRESENTED BECAUSE OF ITS RARITY.

Reference

1. Rekhi B, Jambhekar NA. Morphologic spectrum, immunohistochemical analysis, and clinical features of a series of granular cell tumors of soft tissues: a study from a tertiary referral cancer center. *Ann Diagn Pathol.* 2010 Jun;14(3):162-7.
2. FISHER ER, WECHSLER H. Granular cell myoblastoma--a misnomer. Electron microscopic and histochemical evidence concerning its Schwann cell derivation and nature (granular cell schwannoma). *Cancer.* 1962 Sep-Oct;15:936-54.
3. Lewin MR, Montgomery EA, Barrett TL. New or unusual dermatopathology tumors: a review. *J Cutan Pathol.* 2011 Sep;38(9):689-96. .
4. Paula & Zhou, Paul & Macdonald, Shannon & Ioakeim Ioannidou, Myrsini & Lee, Grace. (2021). Granular Cell Tumor of the Orbit: Review of the Literature and a Proposed Treatment Modality. *Ophthalmic Plastic & Reconstructive Surgery.* Publish Ahead of Print. 10.1097/IOP.0000000000002038.

THANK YOU