KREP

2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

CASE PRESENTATION

CASE OF ORBITAL MYELOID SARCOMA (CHLOROMA)

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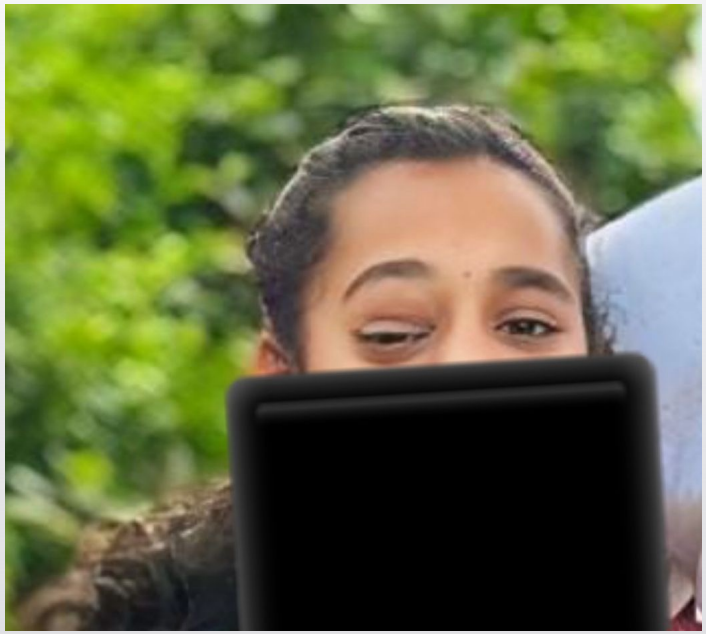
KAHER UNIVERSITY

J.N.MEDICAL COLLEGE ,BELAGAVI

PRESENTOR: DR.SAKSHI ARORA

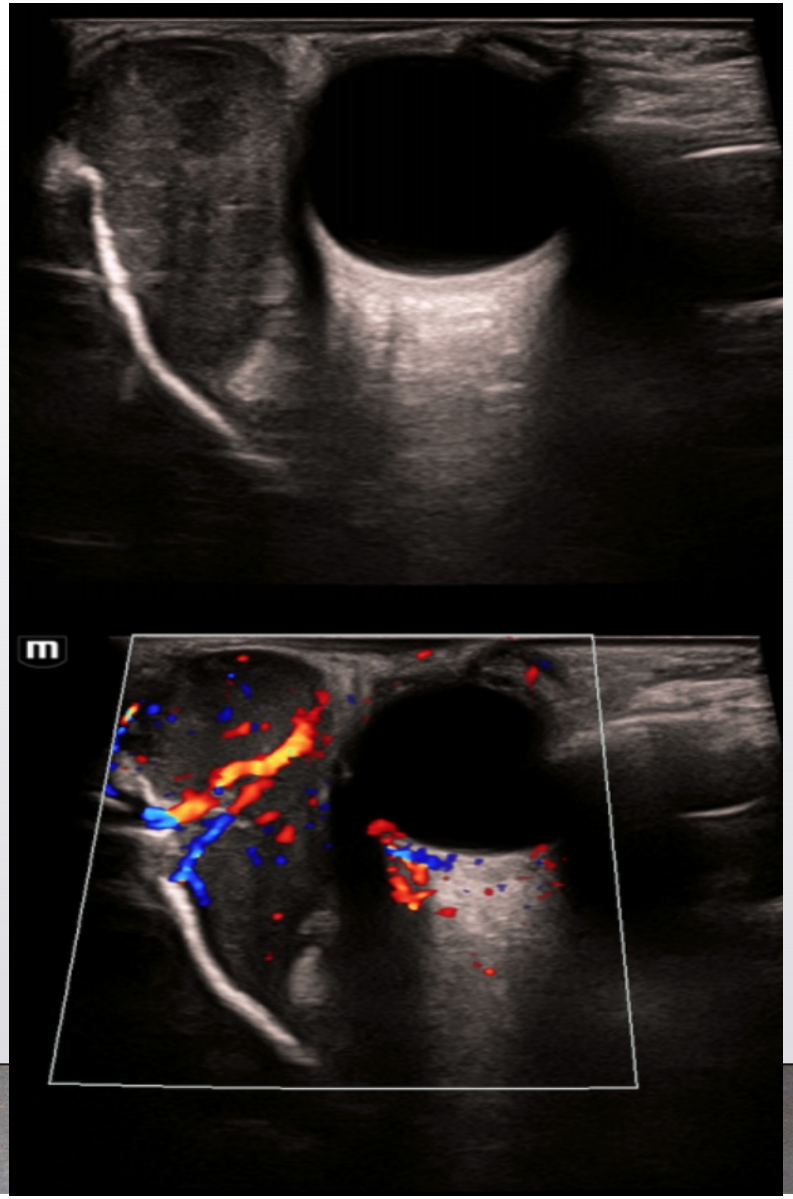
CLINICAL HISTORY OF PATIENT

- **AGE-** Female
- **SEX-** 11 year
- **Brought to OPD with complains of-** painless swelling over right eyelid causing proptosis of the eye with restricted ocular movements & swelling involving left mandibular region since 1 month.
- Not associated with any visual complaints . No history of trauma
- **Blood investigations-** Normal
- **Systemic examination-** Normal

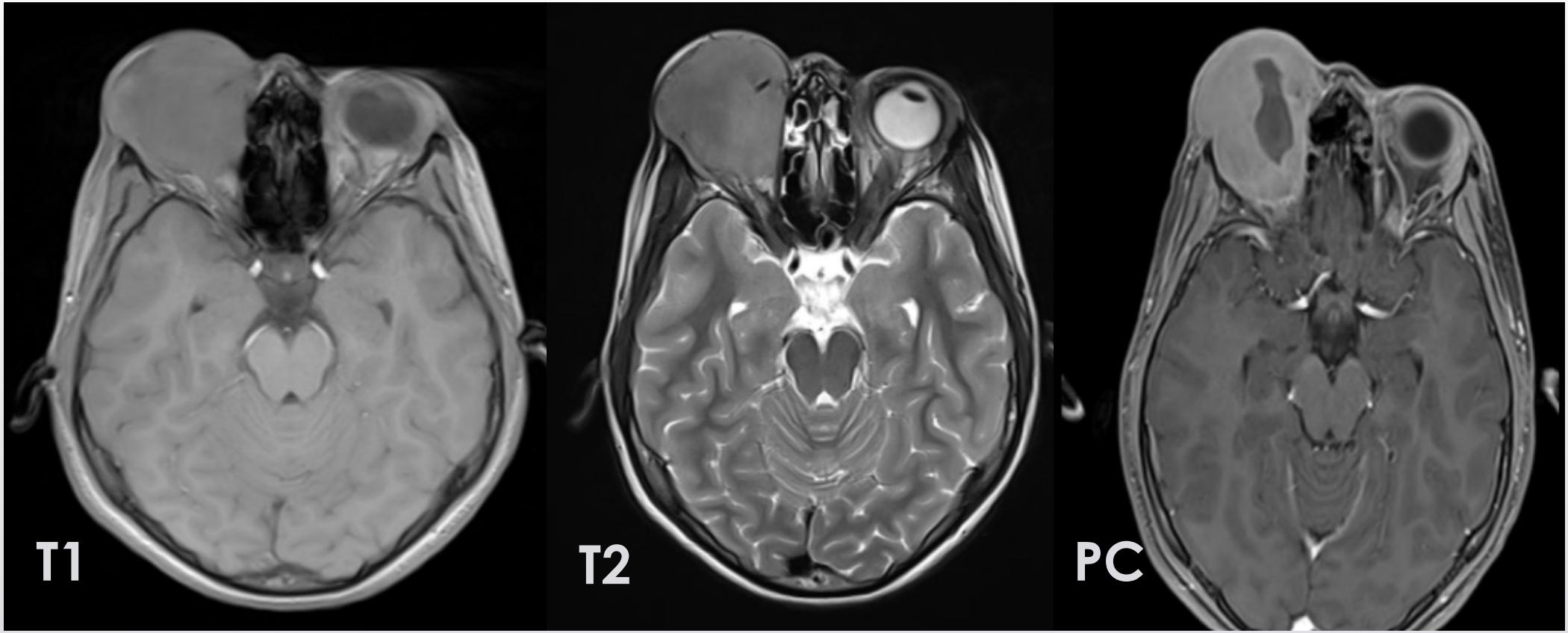


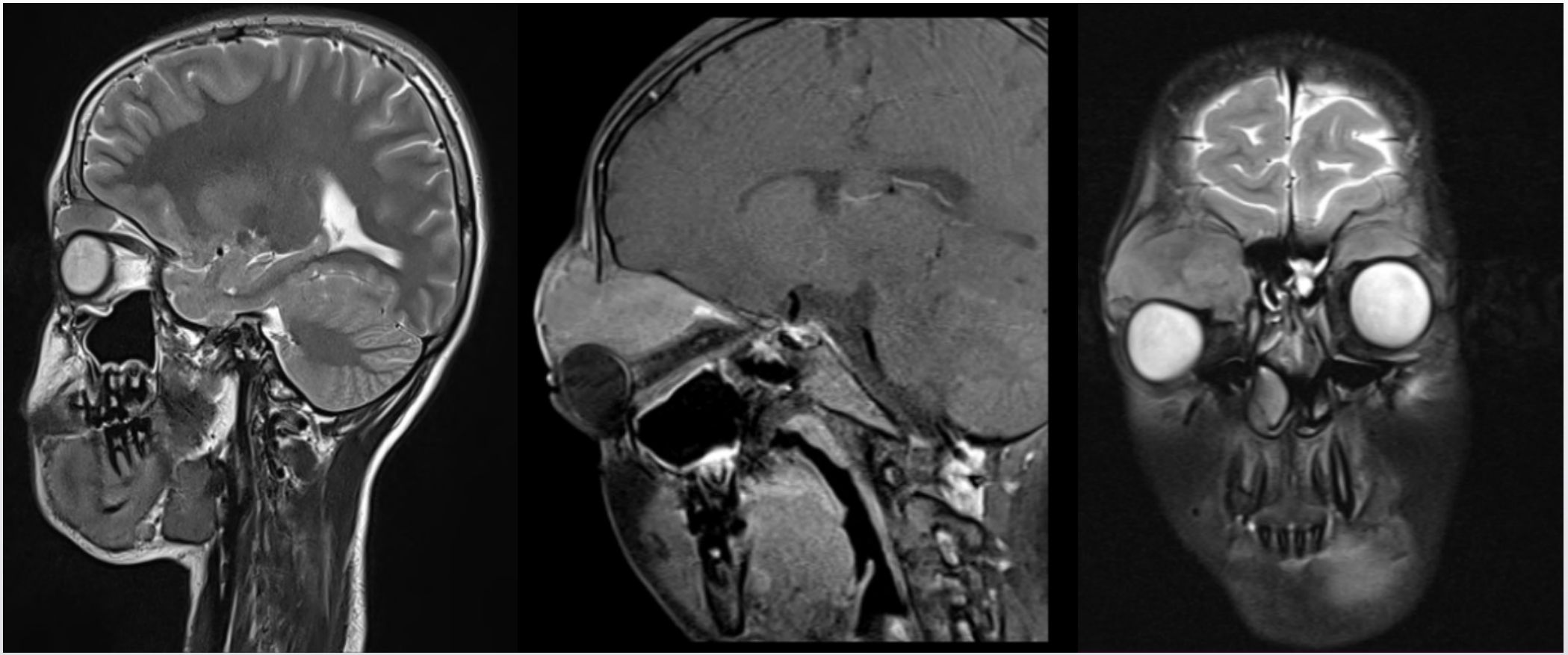
USG FINDINGS

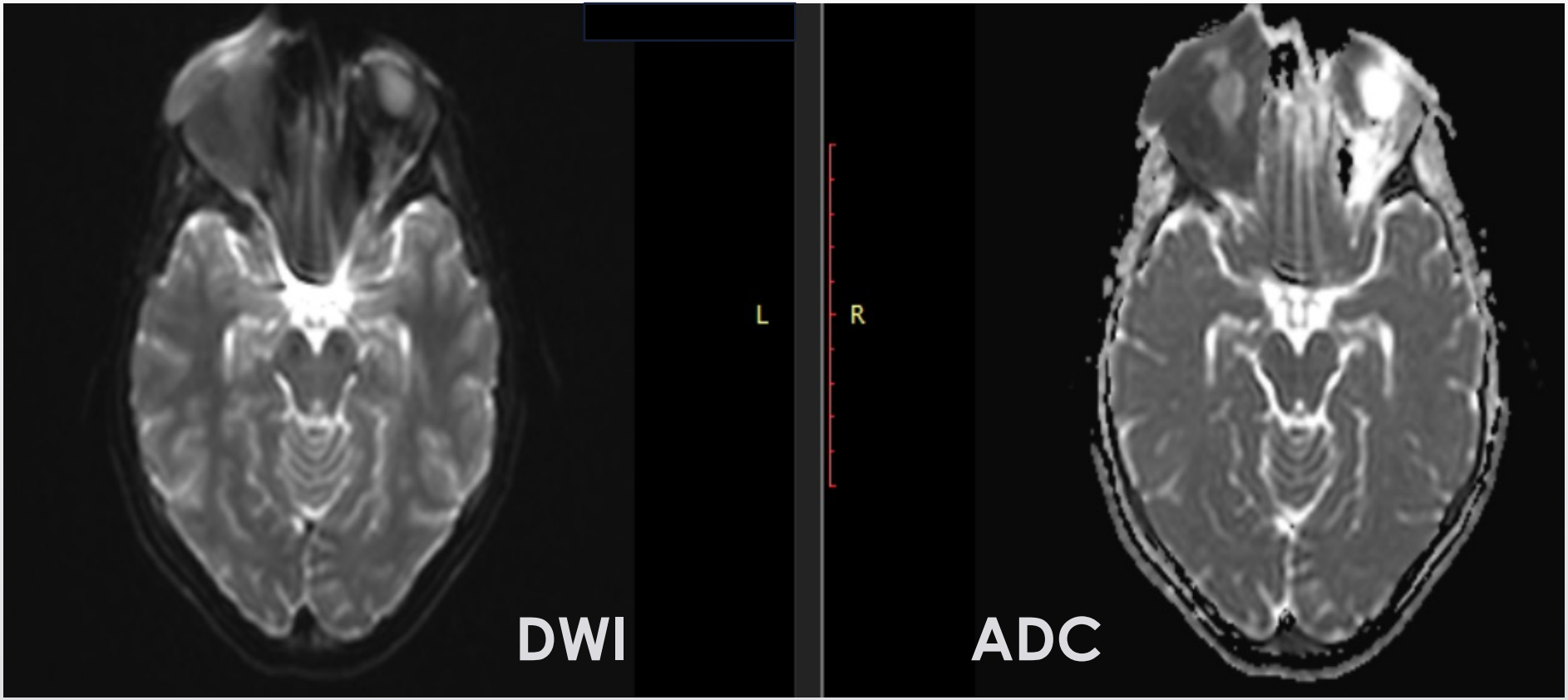
- Heteroechoic lesion involving the extra-conal compartment of right orbit epicentered between the roof of orbit & superior rectus muscle causing inferior displacement of right globe resulting in mild proptosis.
- On colour doppler study- the lesion shows central & peripheral vascularity
- Suggestive of likely neoplastic etiology



MRI FINDINGS



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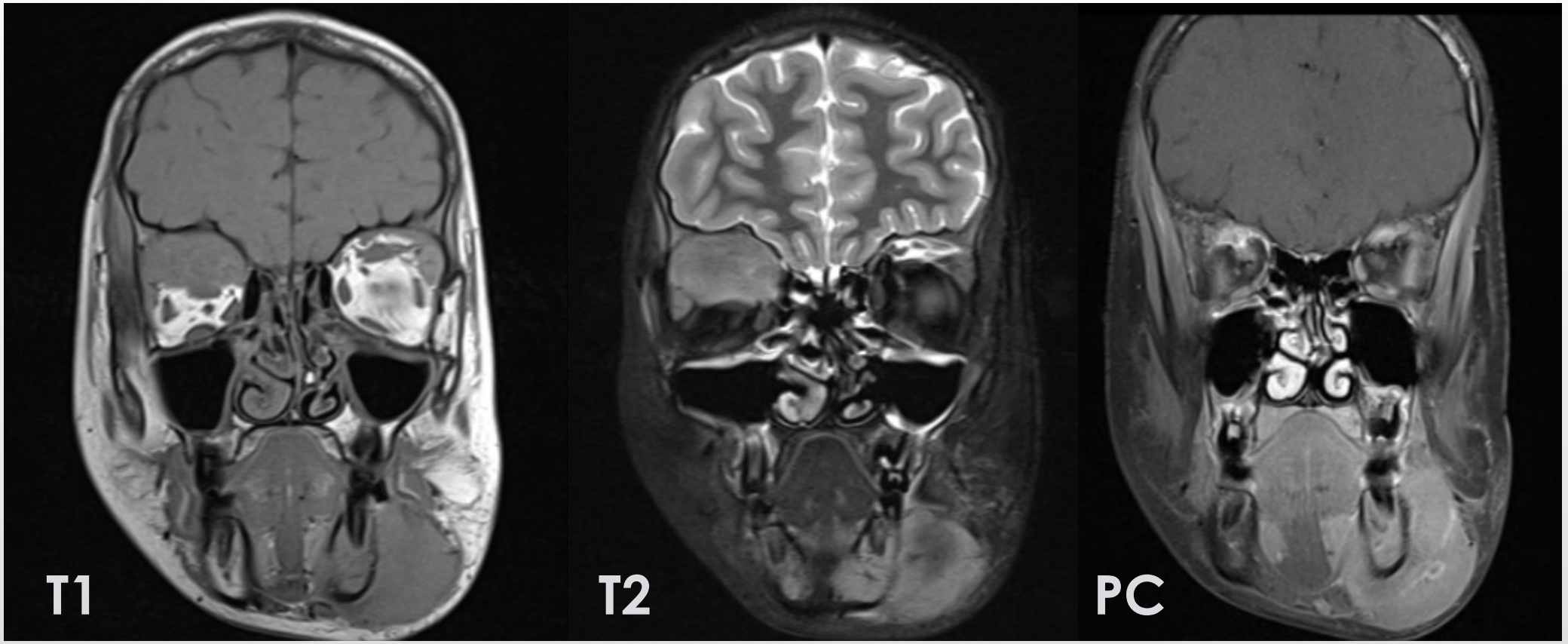
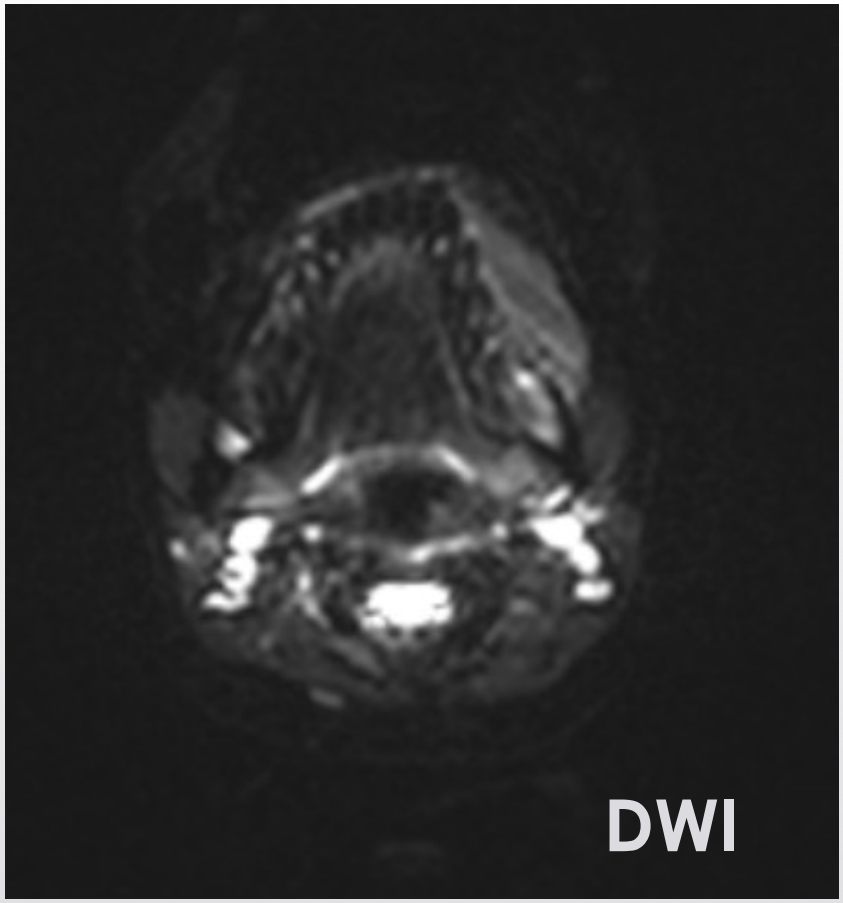
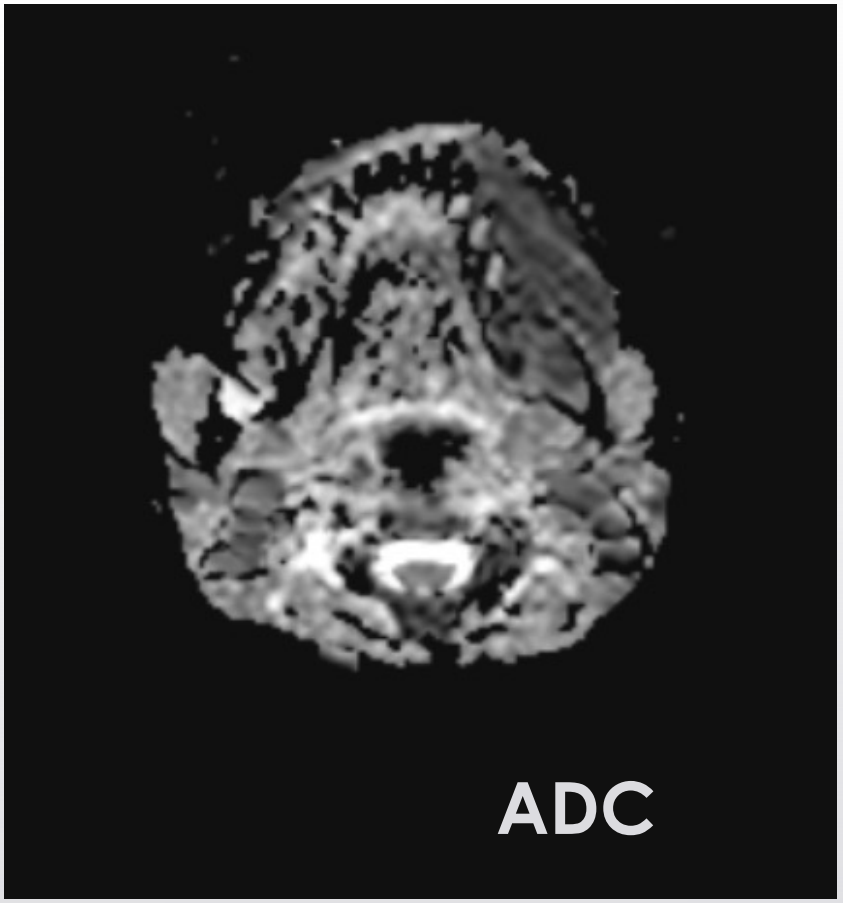

T1
T2
PC


DWI


ADC

MRI FINDINGS

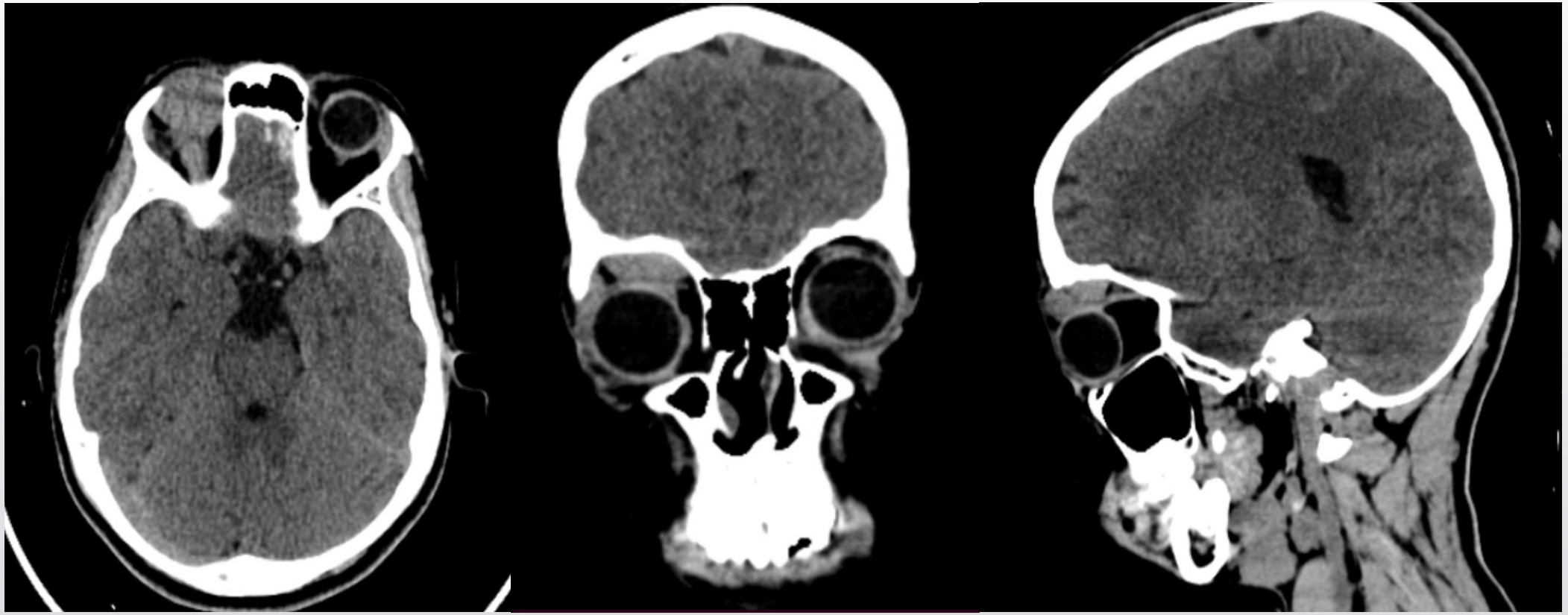
RIGHT ORBIT

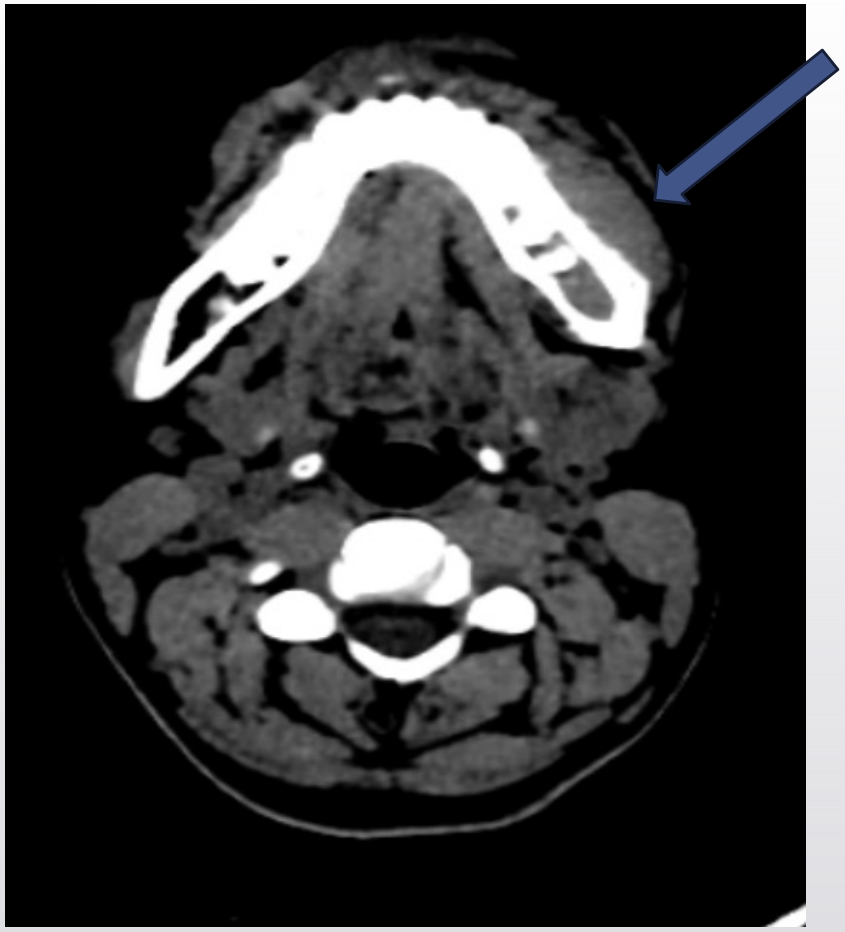
- Heterogenously enhancing T1 & T2 iso to hypointense soft tissue density mass lesion with central non enhancing necrotic area in extra-conal compartment of right orbit epicentered between the roof of orbit & superior rectus muscle causing inferior displacement of right globe resulting in mild proptosis.
- Anteriorly, lesion is seen to cause mass effect & anterior displacement of upper eyelid
- On DWI sequence, the lesion shows mild diffusion restriction
- Lesion is separately visualized from right lacrimal gland
- No adjacent bony erosions noted
- No intra-conal extensions noted

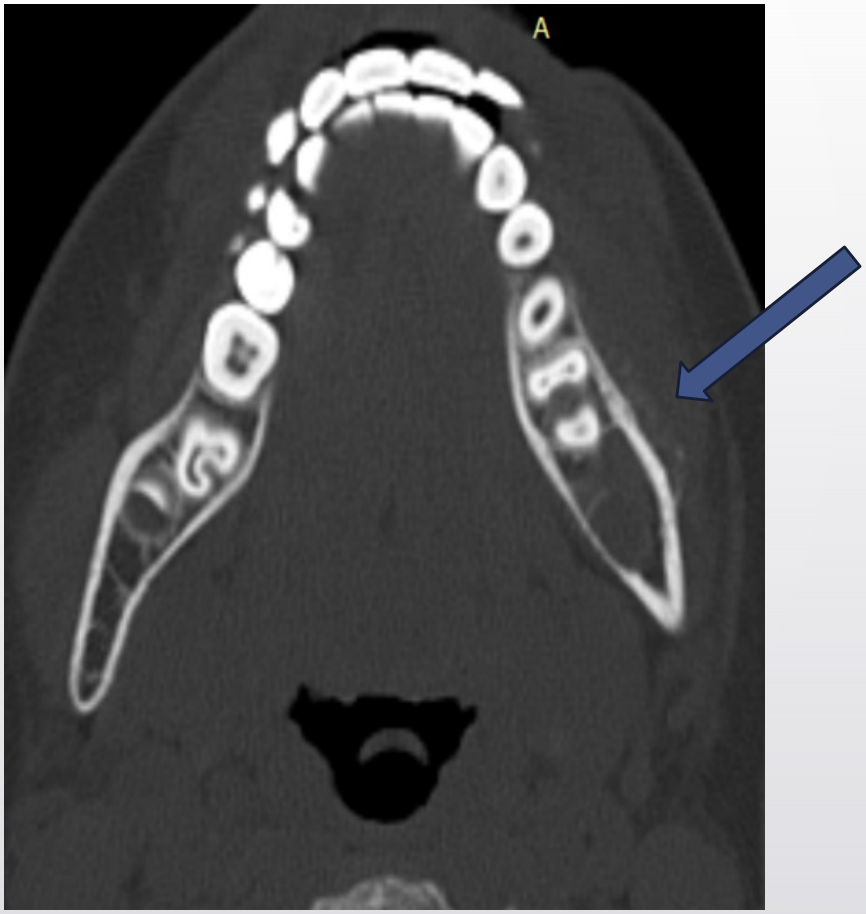
LEFT MANDIBLE

- Heterogenously enhancing T1 isointense & T2 hyperintense soft tissue density mass lesion involving body of mandible & inferior alveolar arch on left side
- On DWI sequence, the lesion shows diffusion restriction
- There is seen widening of mandibular canal, inferior alveolar canal & submental foramen with thickened extra cranial V3 segment of mandibular nerve.

CT FINDINGS



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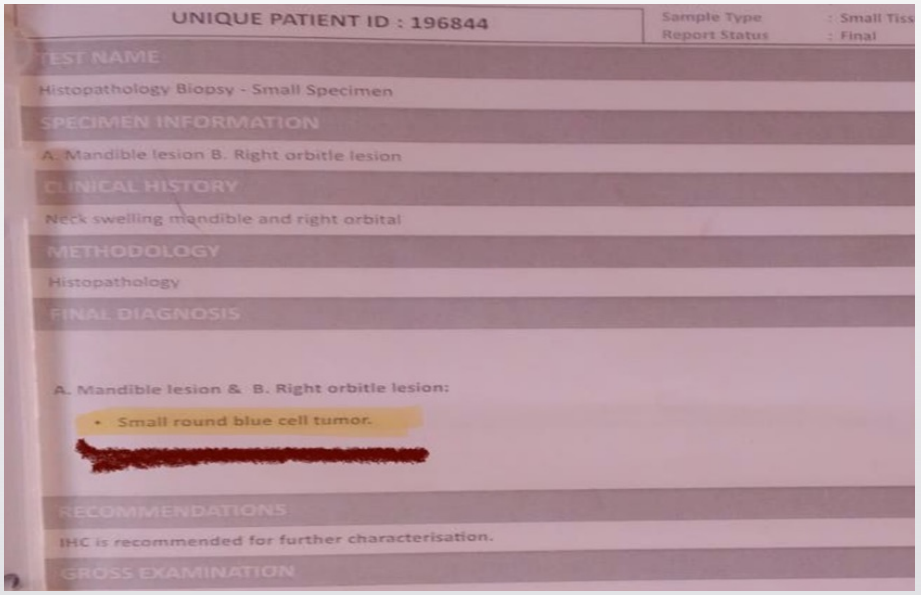
CT FINDINGS

- A well defined soft tissue density mass lesion noted involving the extra-conal compartment of right orbit which is seen to cause compression of superior rectus muscle inferiorly & inferior displacement of the eyeball
- Left eyeball appears normal
- Both optic nerves are normal
- Soft tissue density mass lesion noted involving body of mandible & inferior alveolar arch on left side with thick periosteal reaction noted around the lesion.

FOLLOW UP

- Patient underwent biopsy for orbital & mandibular lesion- came out as SMALL ROUND BLUE CELL TUMOR


UNIQUE PATIENT ID : 196844

Sample Type : Small Tiss

Report Status : Final

TEST NAME

Histopathology Biopsy - Small Specimen

SPECIMEN INFORMATION

A. Mandible lesion B. Right orbitle lesion

CLINICAL HISTORY

Neck swelling mandible and right orbital

METHODOLOGY

Histopathology

FINAL DIAGNOSIS

A. Mandible lesion & B. Right orbitle lesion:

- Small round blue cell tumor.

RECOMMENDATIONS

IHC is recommended for further characterisation.

GROSS EXAMINATION

TABLE 9.1.1 Clinical and Histologic Differential Diagnosis of Small Round Cell Tumors

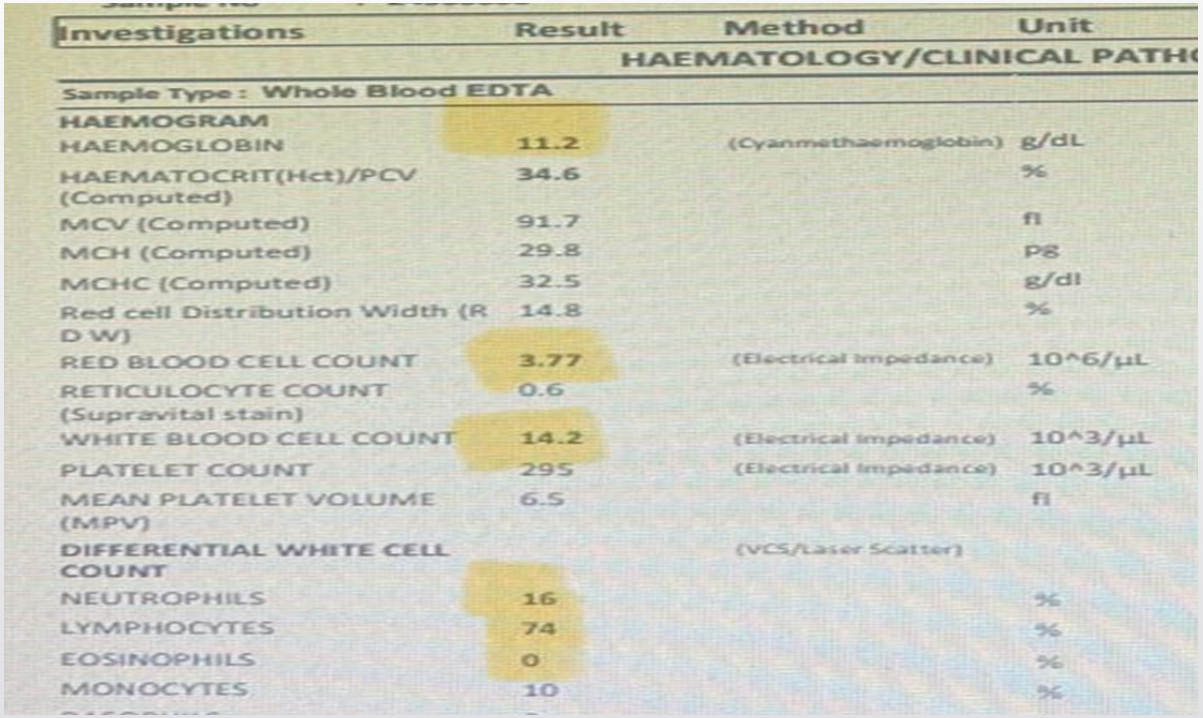
Diagnosis	Patient Age	Location	Distinguishing Histologic Features
Embryonal rhabdomyosarcoma	Young children; peak 3–12 years	Head and neck, genitourinary tract, biliary tract, and retroperitoneum	May have focal rhabdomyoblasts; may have edematous/myxoid stroma and cambium layer
Alveolar rhabdomyosarcoma	Adolescents and young adults; peak 10–25 years	Extremity, head and neck, trunk	Focal rhabdomyoblastic differentiation; usually some alveolar structure
Ewing sarcoma	Children and young adults; peak 10–25 years	Osseous; extraskeletal sites include deep soft tissues	Solid sheets of cells, often with necrosis
Desmoplastic small round cell tumor	Wide age range; mean age 22 years	Intra-abdominal or pelvic	Desmoplastic stromal response; nests of small round blue cells
Poorly differentiated synovial sarcoma	Adults; 30–35 years	Extremity, often close to large joints	Small round blue cells with necrosis and high mitotic index
Round cell liposarcoma	Adults; 20–50 years	Deep soft tissue of the extremity	Focal myxoid areas and lipoblastic differentiation
Undifferentiated round cell sarcoma	Any, including children	Head and neck, trunk, parameningeal	Small round blue cells with no specific features
Small cell osteosarcoma	Peak in children and young adults; 10–20 years	Osseous	Focal osteoid production
Mesenchymal chondrosarcoma	Any; peak 10–30 years	Osseous, soft tissues, and meninges	Biphasic tumor with small blue cells and chondroid matrix material
Neuroblastoma	Infants and young children; 0–2 years	Adrenal gland and sympathetic ganglia	Neurofibrillary matrix, rosette formation, and ganglionic differentiation
Melanotic neuroectodermal tumor of infancy	Infants; 0–1 years	Maxilla and mandible	Biphasic tumor: small round blue cells and larger pigmented epithelioid cells
Lymphoma/leukemia	Any	Lymph nodes, head and neck, mediastinum, intra-abdominal	Variable; "starry sky" or mixed with other inflammatory cells
Small cell carcinoma	Older adults	Lung	Nuclear molding and fragile chromatin
Merkel cell carcinoma	Older adults	Dermis or subcutaneous soft tissue of face and extremity	Nuclear molding and apoptosis

Based on the above follow up, the following differentials to be considered:

1. Rhabdomyosarcoma
2. Orbital lymphoma
3. Metastasis- likely neuroblastoma
4. Leukemia

FOLLOW UP 2

- Patient underwent bone marrow biopsy & CSF cytology
- CSF Cytology- negative for malignant cells
- BONE MARROW BIOPSY- ***Feature suggestive of acute leukemia***



Investigations	Result	Method	Unit
		HAEMATOLOGY/CLINICAL PATH	
Sample Type : Whole Blood EDTA			
HAEMOGRAM			
HAEMOGLOBIN	11.2	(Cyanmethaemoglobin)	g/dL
HAEMATOCRIT(Hct)/PCV (Computed)	34.6		%
MCV (Computed)	91.7		fl
MCH (Computed)	29.8		pg
MCHC (Computed)	32.5		g/dl
Red cell Distribution Width (R D W)	14.8		%
RED BLOOD CELL COUNT	3.77	(Electrical Impedance)	10^6/µL
RETICULOCYTE COUNT (Supravital stain)	0.6		%
WHITE BLOOD CELL COUNT	14.2	(Electrical Impedance)	10^3/µL
PLATELET COUNT	295	(Electrical Impedance)	10^3/µL
MEAN PLATELET VOLUME (MPV)	6.5		fl
DIFFERENTIAL WHITE CELL COUNT		(VCS/Laser Scatter)	
NEUTROPHILS	16		%
LYMPHOCYTES	74		%
EOSINOPHILS	0		%
MONOCYTES	10		%

HAEMATOLOGY/CLINICAL PATHOLOGY

Investigations	Result	Units	Biological Range.
Bone Marrow(Proc,Reporting,Special stains)By Appt.			
ASPIRATION	Received 7 unstained slides for reporting		(BM/305/57)
PREPARATION AND STAINING	Satisfactory		
CELLULARITY	Hypercellular		
ERYTHROPIESIS	Normomegaloblastic maturation		
MYELOPOIESIS	Increased with increase in immature granular series of cells with predominant promyelocytes and myeloblasts		
MEGA KARYOCYTES	Normal in number and morphology		
PLASMA CELLS	02 %		
PARASITES	Nil		
ABNORMAL CELLS	Few myeloblasts and promyelocytes with cytoplasmic vacuoles, few showing Auer rods are seen		
M:E RATIO	8 : 1		
OTHER FINDINGS	Marrow Differential count : Myeloblasts - 23 % Promyelocytes - 42 % Myelocytes - 08 % Metamyelocytes - 04 % Band forms - 02 % Neutrophils - 06 % Lymphocytes - 15 %.		
IMPRESSION	Features suggestive of Acute Leukaemia - AML M2/M3		

Investigations	Result	Method	Unit	Reference Range
HISTOPATHOLOGY				
Sample Type : FLUID				
CYTO NO:	1262/24			
SITE:	Cerebrospinal Fluid			
GROSS:	Received 1 ml clear, colorless fluid.			
MICROSCOPY:	Pap and Giemsa stained smears show occasional lymphocytes. The of atypical cells in the smears studied.			
IMPRESSION *	Cerebrospinal Fluid Cytology - Negative for malignant cells.			

Orbital Myeloid Sarcoma (Chloroma)

- Myeloid sarcoma (MS) of the orbit is an uncommon condition occurring in children, generally coupled to myeloproliferative neoplasms.
- Myeloid sarcoma (MS) is an extra-medullary solid tumor caused by an abnormal proliferation of primitive immature precursors of the granulocytic series of white blood cells.
- MS is also called "chloroma" because of its green color secondary to the presence of intracellular myeloperoxidase. Subsequently, because of its macroscopic appearance variability, the tumor was renamed granulocytic sarcoma in 1966.
- MS is a rare disease, often related to other underlying unrecognized myeloproliferative conditions.MS occurs in 2.5–9.1% of patients with acute myeloid leukemia (AML). Less frequently it occurs as a initial manifestation of AML in non-leukemic patients, or in association with myelodysplastic disorders or chronic myeloid leukemia (CML) with impending blast crisis.
- In pediatric population, orbit is one of the most common sites of occurrence.

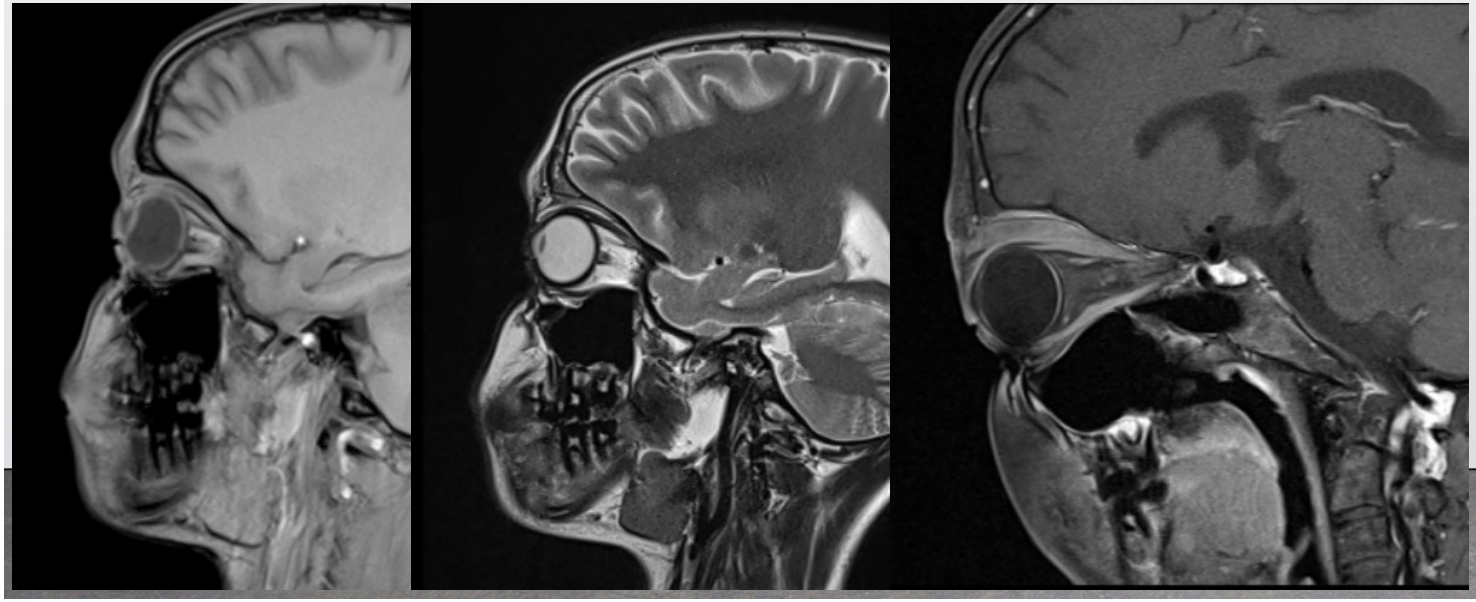
- When myeloid sarcoma presents as an isolated finding which is very rare, this may mimic inflammatory/infective or lymphoproliferative diseases .
- Even though it can be asymptomatic, proptosis is the most common presenting feature. It commonly presents as unilateral exophthalmos.
- Proptosis is usually due to leukemic infiltrates, retrobulbar hemorrhage, orbital muscle infiltration, or venous blockage. In such cases, acute leukemia may develop shortly afterward with a median time ranging from 1-25 months.
- Other clinical differential diagnosis includes-
  - vascular lesion
  - Lymphoma
  - metastatic neuroblastoma
  - rhabdomyosarcoma
- Presentation as eyelid swelling mimicking preseptal cellulitis is possible.

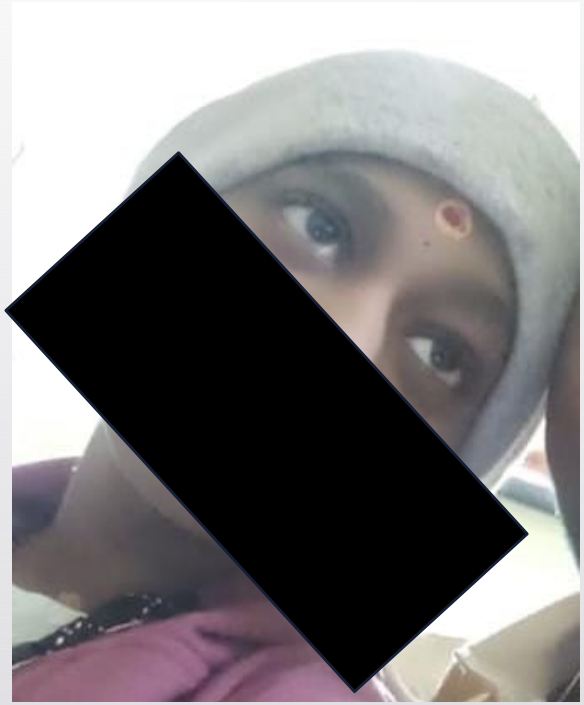
TREATMENTS AVAILABLE

- There is no current consensus on the best therapy due to the rarity of the myeloid sarcoma.
- Orbital myeloid sarcoma has been generally thought to be an antecedent event to the evolution of AML, and treatment options are customized to the individual patient.
- Therapeutic options include surgery, radiotherapy, and chemotherapy.
- Given the systemic nature of AML, chemotherapy is the mainstay of management in most cases.

TREATMENT FOLLOW UP OF PATIENT

- Patient has taken 5 cycles chemotherapy (vincristine)
- Complete resolution of orbital and mandibular swelling





THANK YOU