PLEASE…RESPECT THE CHILD

MONSEF IBRAHIM KHARBOUSH

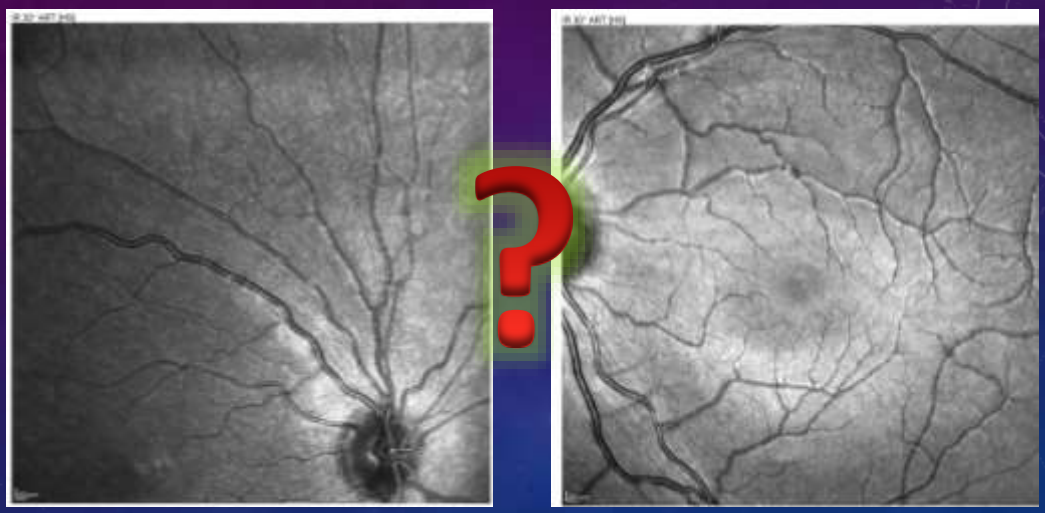
4TH YEAR RESIDENT

ALEXANDRIA MAIN UNIVERSITY HOSPITAL

- A 6 yo girl with no PMH presented to the clinic with vague history of **BOV in her eyes** which started **3-4 months ago**.
- Her Mother stated that they went to many ophthalmologists who reassured her and told her that she is only "Faking" her symptoms

O/E

	OD	OS
UCVA	HM?? CF	**CF 3m**
Lids	Free	free
Conj	Free	free
Pupils	Sluggish	Sluggish
IOP	TND	TND
EOM	FMAD	FMAD
Anterior segment	Clear	clear
Fundus	NDVs	NDVs



OD OS

Does she needs glasses ?! Is she amblyope ?!

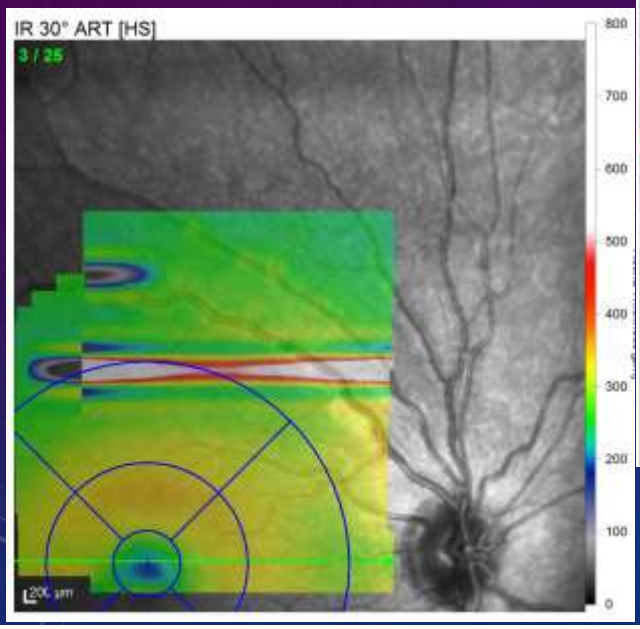
But no Improvement on correction!

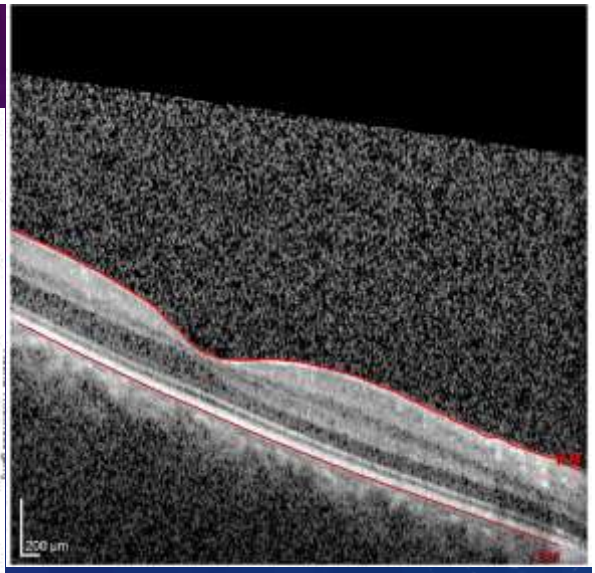
Cycloplegic refraction

What to do NEXT??

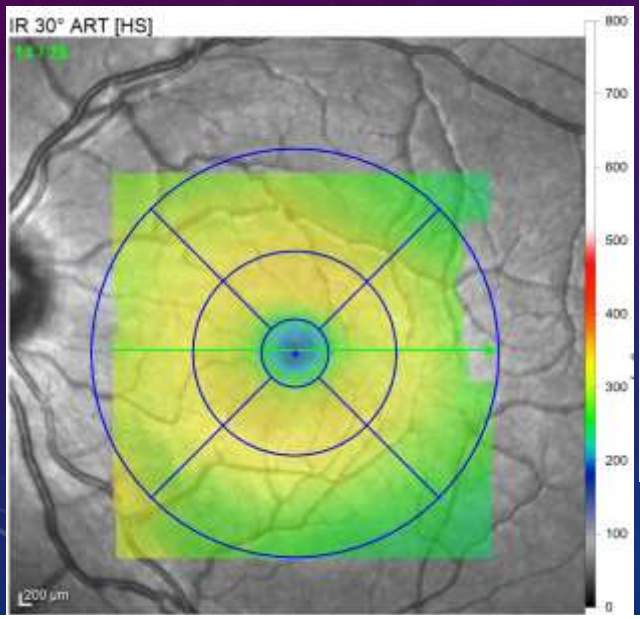
The mother told me: "I told you she is faking it"

OCT OD


IR 30° ART [HS]
3 / 25
800
700
600
500
400
300
200
100
0
200 µm


200 µm

OCT OS


IR 30° ART [HS]
800
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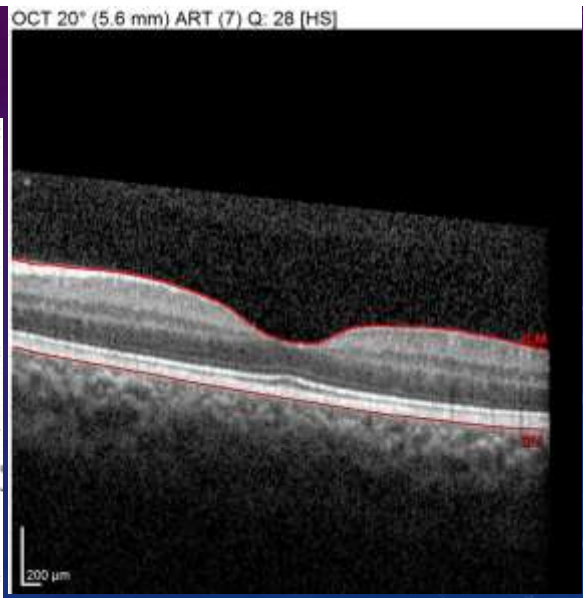

OCT 20° (5.6 mm) ART (7) Q: 28 [HS]
200 µm

CONFRONTATION VISUAL FIELD

OD

OS

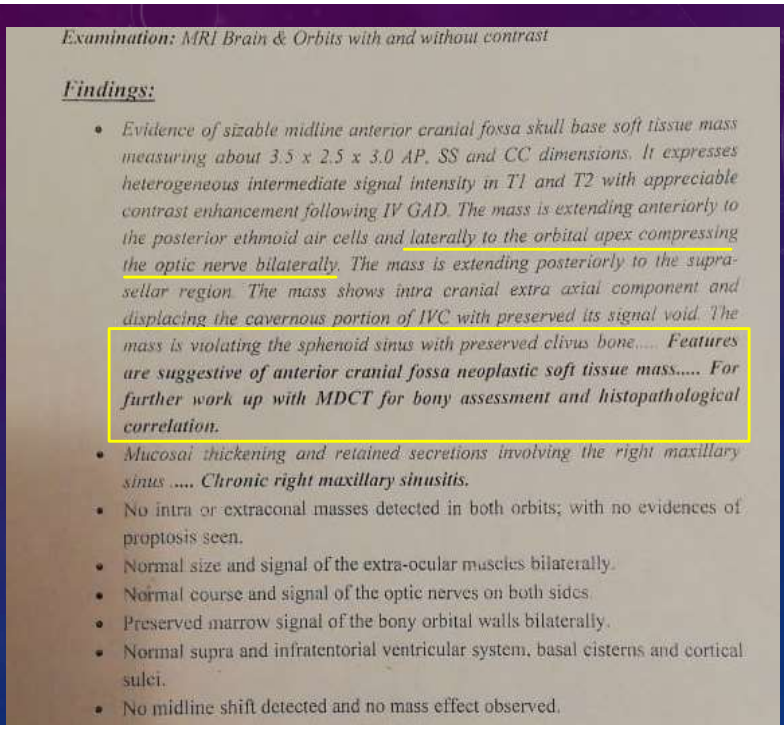
TESTING FOR FUNCTIONAL VISUAL LOSS

- Menace reflex: absent OD – present OS
- Eye contact: absent
- Fogging the left eye with +10D on a trial frame, asking the girl to reach her mother walking: She hit the wall, changed her direction following only her mother's voice

The girl is not FAKING

MRI BRAIN AND ORBIT WAS ORDERED

MRI



Examination: MRI Brain & Orbits with and without contrast

Findings:

- *Evidence of sizable midline anterior cranial fossa skull base soft tissue mass measuring about 3.5 x 2.5 x 3.0 AP, SS and CC dimensions. It expresses heterogeneous intermediate signal intensity in T1 and T2 with appreciable contrast enhancement following IV GAD. The mass is extending anteriorly to the posterior ethmoid air cells and laterally to the orbital apex compressing the optic nerve bilaterally. The mass is extending posteriorly to the supra-sellar region. The mass shows intra cranial extra axial component and displacing the cavernous portion of IVC with preserved its signal void. The mass is violating the sphenoid sinus with preserved clivus bone..... **Features are suggestive of anterior cranial fossa neoplastic soft tissue mass..... For further work up with MDCT for bony assessment and histopathological correlation.***
- *Mucosal thickening and retained secretions involving the right maxillary sinus ..... **Chronic right maxillary sinusitis.***
- No intra or extraconal masses detected in both orbits; with no evidences of proptosis seen.
- Normal size and signal of the extra-ocular muscles bilaterally.
- Normal course and signal of the optic nerves on both sides.
- Preserved marrow signal of the bony orbital walls bilaterally.
- Normal supra and infratentorial ventricular system, basal cisterns and cortical sulci.
- No midline shift detected and no mass effect observed.

HISTOPATHOLOGY

Clinical Diagnosis : Petroclival, Sphenoid and ethmoid sinus mass

Nature of Specimen : Endoscopic biopsy

Receiving Date : 01-05-2018 **Delivery Date :** 04-05-2018

PATHOLOGY REPORT

Gross :

Multiple irregular rubbery to firm tissue pieces entangling tiny bony tissue, collectively measured 3x3 cm, totally submitted.

Microscopic :

Sections examined from the specimen received revealed pieces of tumor tissue showing dense diffuse infiltration by intermediate size lymphoid cells with high mitotic activity and showing focal stary sky apperance. Tumor is infiltrating bony tissue.

Diagnosis :

Sphenoid and ethmoid sinus mass, Endoscopic biopsy, HIGH GRADE LYMPHOMA, COMPATIBLE WITH BURKITT'S LYMPHOMA

IMMUNOHISTOCHEMISTRY

IMMUNOHISTOCHEMISTRY REPORT

Technique :

Sections were prepared from the paraffin block, processed in Benchmark Ultra machine (Ventana) and stained by rabbit monoclonal antibodies using Ultra view HRP DAB as chromogen and hematoxylin as counter-stain.
The sections were treated against:
-KI67
Sections were prepared from the paraffin block then treated by monoclonal antibodies and detection kit (EnVision FLEX) using DAB as chromogen and hematoxylin as counter-stain, using Omnis machine
The sections were treated against:
-CD20
-CD3
-CD10

Results :

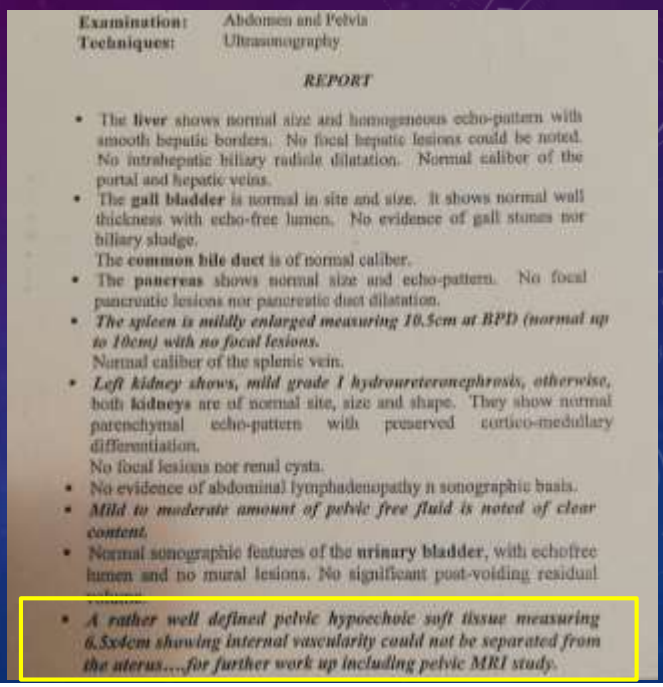
Neoplastic cells are POSITIVE for CD20
Neoplastic cells are NEGATIVE for CD3
Neoplastic cells are POSITIVE for CD10
100 % of Neoplastic cells are positive for Ki 67.

Conclusion :

***Block, FINDINGS ARE CONSISTENT WITH* BURKITT'S LYMPHOMA.**

The girl was sent to the pediatric oncology for management

U/S ABDOMEN & PELVIS

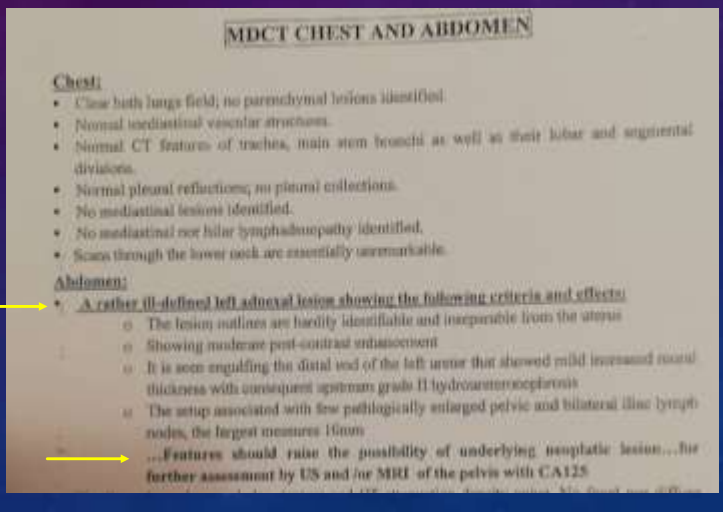


Examination: Abdomen and Pelvis
Techniques: Ultrasonography

REPORT

- The **liver** shows normal size and homogeneous echo-pattern with smooth hepatic borders. No focal hepatic lesions could be noted. No intrahepatic biliary radicle dilatation. Normal caliber of the portal and hepatic veins.
- The **gall bladder** is normal in site and size. It shows normal wall thickness with echo-free lumen. No evidence of gall stones nor biliary sludge.
  The **common bile duct** is of normal caliber.
- The **pancreas** shows normal size and echo-pattern. No focal pancreatic lesions nor pancreatic duct dilatation.
- ***The spleen is mildly enlarged measuring 10.5cm at BPD (normal up to 10cm) with no focal lesions.***
  Normal caliber of the splenic vein.
- ***Left kidney shows, mild grade 1 hydroureteronephrosis, otherwise,*** both **kidneys** are of normal site, size and shape. They show normal parenchymal echo-pattern with preserved cortico-medullary differentiation.
  No focal lesions nor renal cysts.
- No evidence of abdominal lymphadenopathy n sonographic basis.
- ***Mild to moderate amount of pelvic free fluid is noted of clear content.***
- Normal sonographic features of the **urinary bladder**, with echofree lumen and no mural lesions. No significant post-voiding residual
- ***A rather well defined pelvic hypoechoic soft tissue measuring 6.5x4cm showing internal vascularity could not be separated from the uterus….for further work up including pelvic MRI study.***

CT CHEST AND ABDOMEN



MDCT CHEST AND ABDOMEN

Chest:

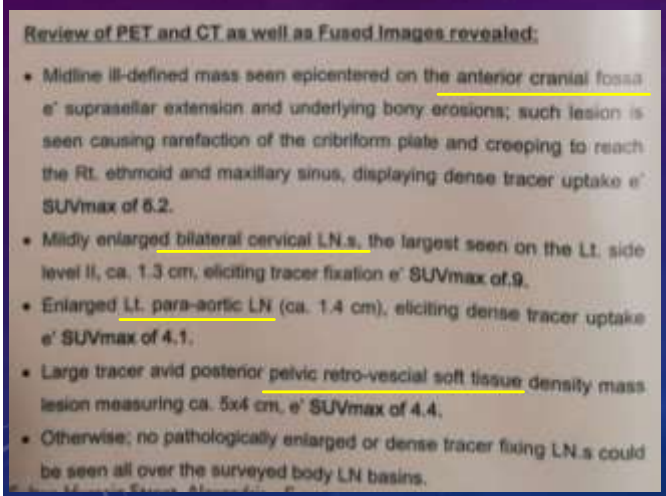
- Clear both lungs field; no parenchymal lesions identified.
- Normal mediastinal vascular structures.
- Normal CT features of trachea, main stem bronchi as well as their lobar and segmental divisions.
- Normal pleural reflections; no pleural collections.
- No mediastinal lesions identified.
- No mediastinal nor hilar lymphadenopathy identified.
- Scans through the lower neck are essentially unremarkable.

Abdomen:

- A rather ill-defined left adnexal lesion showing the following criteria and effects:
  - The lesion outlines are hardly identifiable and inseparable from the uterus
  - Showing moderate post-contrast enhancement
  - It is seen engulfing the distal end of the left ureter that showed mild increased mural thickness with consequent upstream grade II hydroureteronephrosis
  - The setup associated with few pathologically enlarged pelvic and bilateral iliac lymph nodes, the largest measures 10mm

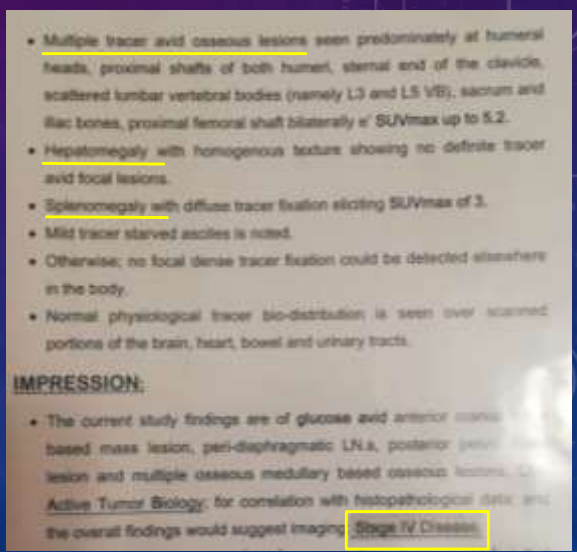
...Features should raise the possibility of underlying neoplastic lesion...for further assessment by US and /or MRI of the pelvis with CA125

PET SCAN



Review of PET and CT as well as Fused Images revealed:

- Midline ill-defined mass seen epicentered on the anterior cranial fossa e' suprasellar extension and underlying bony erosions; such lesion is seen causing rarefaction of the cribriform plate and creeping to reach the Rt. ethmoid and maxillary sinus, displaying dense tracer uptake e' SUVmax of 6.2.
- Mildly enlarged bilateral cervical LN.s, the largest seen on the Lt. side level II, ca. 1.3 cm, eliciting tracer fixation e' SUVmax of 9.
- Enlarged Lt. para-aortic LN (ca. 1.4 cm), eliciting dense tracer uptake e' SUVmax of 4.1.
- Large tracer avid posterior pelvic retro-vescial soft tissue density mass lesion measuring ca. 5x4 cm, e' SUVmax of 4.4.
- Otherwise; no pathologically enlarged or dense tracer fixing LN.s could be seen all over the surveyed body LN basins.



- Multiple tracer avid osseous lesions seen predominately at humeral heads, proximal shafts of both humeri, sternal end of the clavicle, scattered lumbar vertebral bodies (namely L3 and L5 VB), sacrum and iliac bones, proximal femoral shaft bilaterally e' SUVmax up to 5.2.
- Hepatomegaly with homogenous texture showing no definite tracer avid focal lesions.
- Splenomegaly with diffuse tracer fixation eliciting SUVmax of 3.
- Mild tracer starved ascites is noted.
- Otherwise; no focal dense tracer fixation could be detected elsewhere in the body.
- Normal physiological tracer bio-distribution is seen over scanned portions of the brain, heart, bowel and urinary tracts.

IMPRESSION:

- The current study findings are of glucose avid anterior cranial based mass lesion, peri-diaphragmatic LN.s, posterior pelvic lesion and multiple osseous medullary based osseous lesions. C Active Tumor Biology, for correlation with histopathological data and the overall findings would suggest imaging Stage IV Disease.

Unfortunately, after one month of trials of chemotherapy and immunosuppression…

SHE passed away

BURKITT LYMPHOMA

- The most common childhood tumour in subSaharan Africa
- An aggressive form of Non-Hodgkin Lymphoma
- Typically affects the jaws and abdomen.
- Eye affection occurs in orbital or CNS disease

Case Reports Ann Trop Paediatr. 10 (3), 319-22 1990

Burkitt's Lymphoma Presenting With Blindness: A Case Report

M Ibrahim, W N Ogala ... E A Afolayan + expand

PMID: 1703753

Indian J Med Paediatr Oncol. 2013 Apr-Jun; 34(2): 99–100

doi: 10.4103/0971-5851.116188

PMCID: PMC3764753

PMID: 24049296

Acute visual loss as the initial presentation in a child with systemic Burkitt lymphoma

Maria Kourti, Elpidoforos Mantadakis,[1] Athanassia Anastasiou[2] and Dimitrios Kolialexis

J Med Case Rep. 2018 May 16;12(1):129. doi: 10.1186/s13256-018-1682-3.

Acute blindness as a presenting sign of childhood endemic Burkitt's lymphoma in Cameroon: a case report.

Vofo BN[1], Nnanjam GVP[2], Nowasiri CA[3], Atem JA[4], Aminde LN[5]

Int J Pediatr Otorhinolaryngol. 2012 May;76(5):740-1. doi: 10.1016/j.ijporl.2012.01.032. Epub 2012 Mar 6.

Acute bilateral blindness as a presenting symptom of Non-Hodgkin's lymphoma.

Emami N[1], Daniel SJ.

TAKE HOME MESSAGE

- Respect, respect and respect the patient's complain
- Don't always blame it on amblyopia, search for a cause
- Don't forget to examine the pupils
- If in doubt, don't think economically and order the investigation
- Lastly, pray for the child :'(

ACKNOWLEDGEMENT

Prof. Dr. Sahar El-Sheikh

Youssef A. Elmassry

In the memory of
Yara
11/2011-7/2018

Thank You