



Orbital Lymphoma

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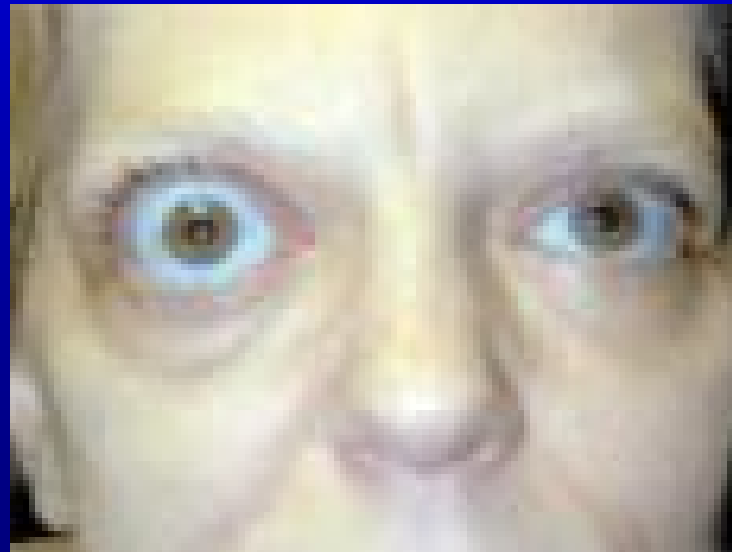
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Our Patient – Clinical Picture

- This topic will be presented in the context of our patient.
- She is a 71 year old female who presented with gradual painless right sided proptosis.





Imaging Options for the Orbits

To image the orbit, we can use:

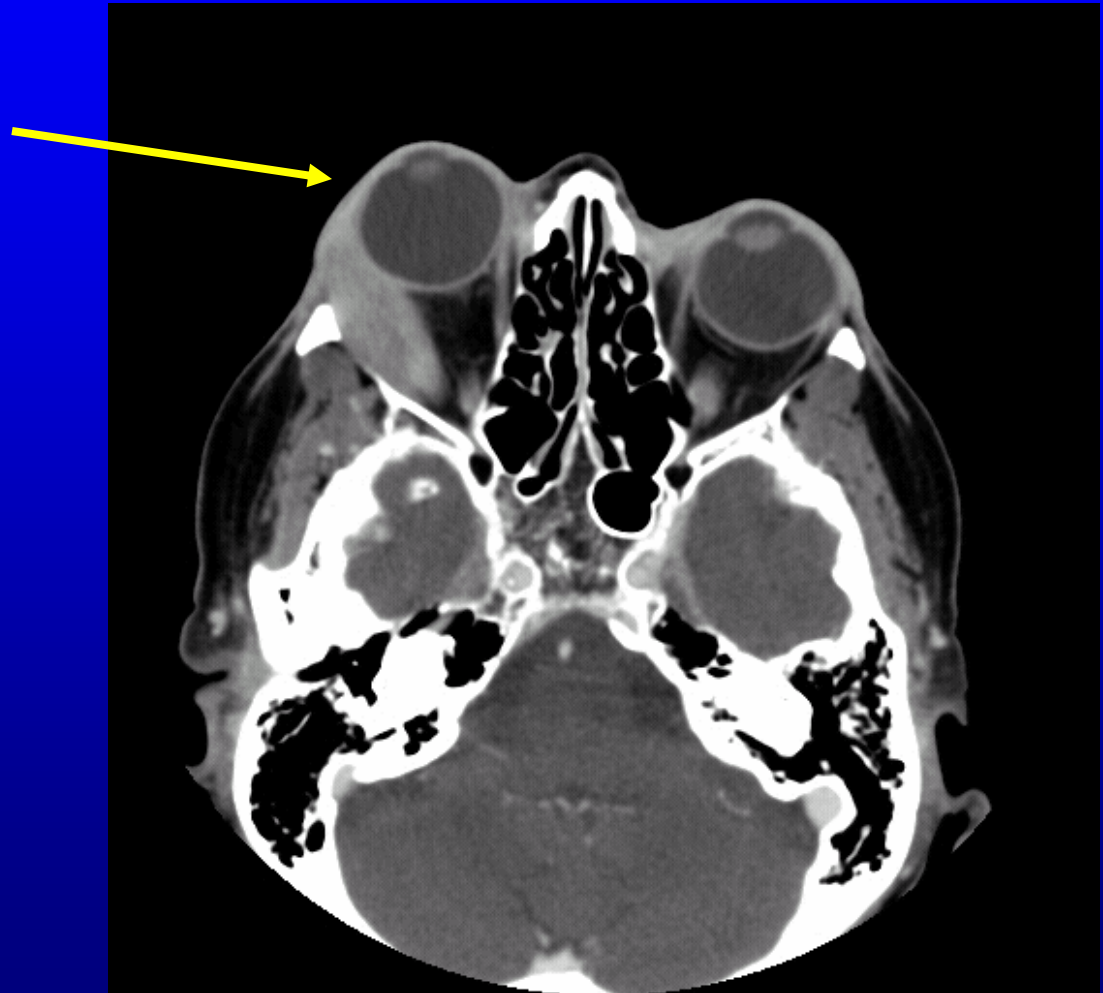
- **Plain Film**
 - Its main use is screening for metallic foreign bodies.
- **Ultrasound**
 - It is used to characterize abnormalities (e.g., masses).
- **CT**
 - For further characterization of abnormalities and bone detail (e.g., abnormal bone mineralization).
- **MRI**
 - It is chosen for its enhanced soft tissue contrast.
- **Angiogram**
 - It identifies aberrant blood vessels such as those created by tumors (not routinely used).



Our Patient's CT Scan of Orbits

(with contrast)

- Marked displacement of the globe anteriorly.
- A lateral mass, hyperdense to fat, is present in the right orbit:
 - It is homogeneous in its composition.
 - It measures 4x2cm.
 - It has intra-conal and extra-conal components.
 - There is no evident local bone erosion.

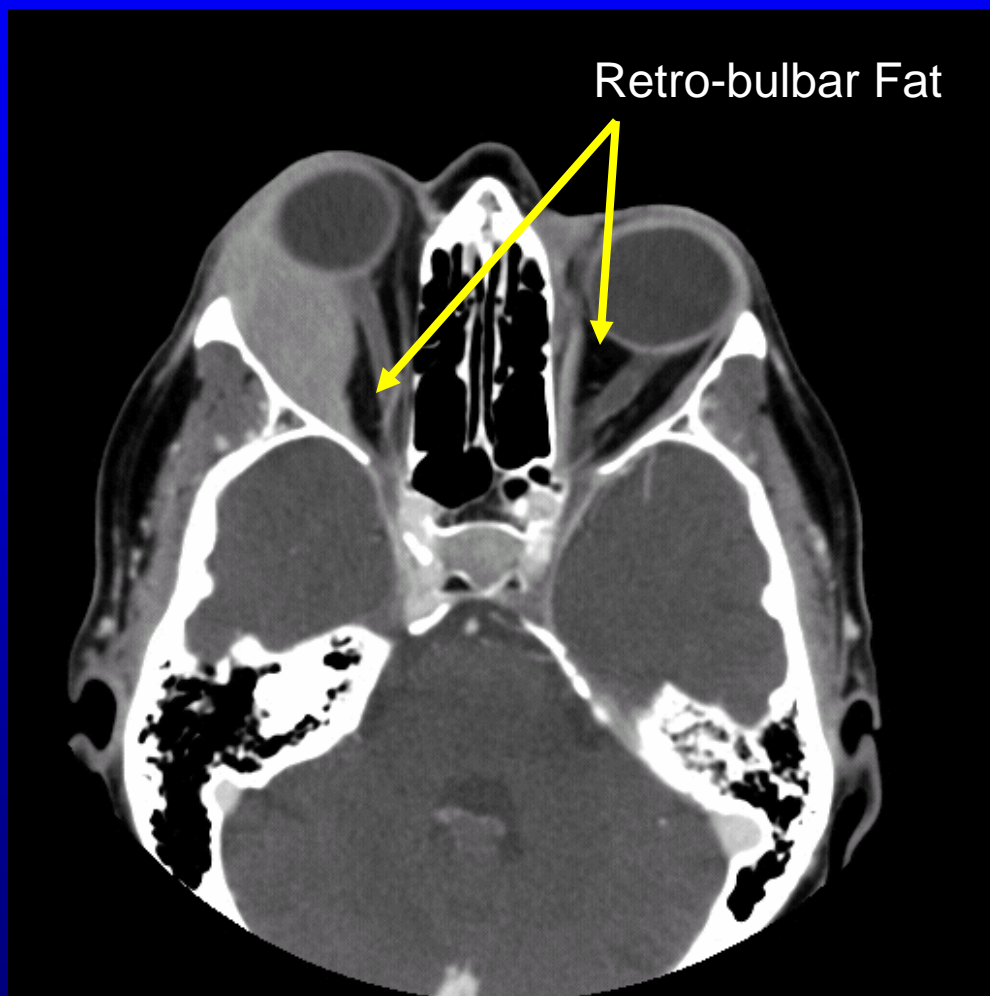




Our Patient's CT Scan of Orbits

(with contrast)

- The right lateral rectus muscle is not seen. The mass is either displacing, encasing or expanding it. A coronal view would aid its identification.
- There is no remodelling or indenting of the globe contour.
- There is no reticulation of retro-bulbar fat (unlike that seen with pseudotumor).

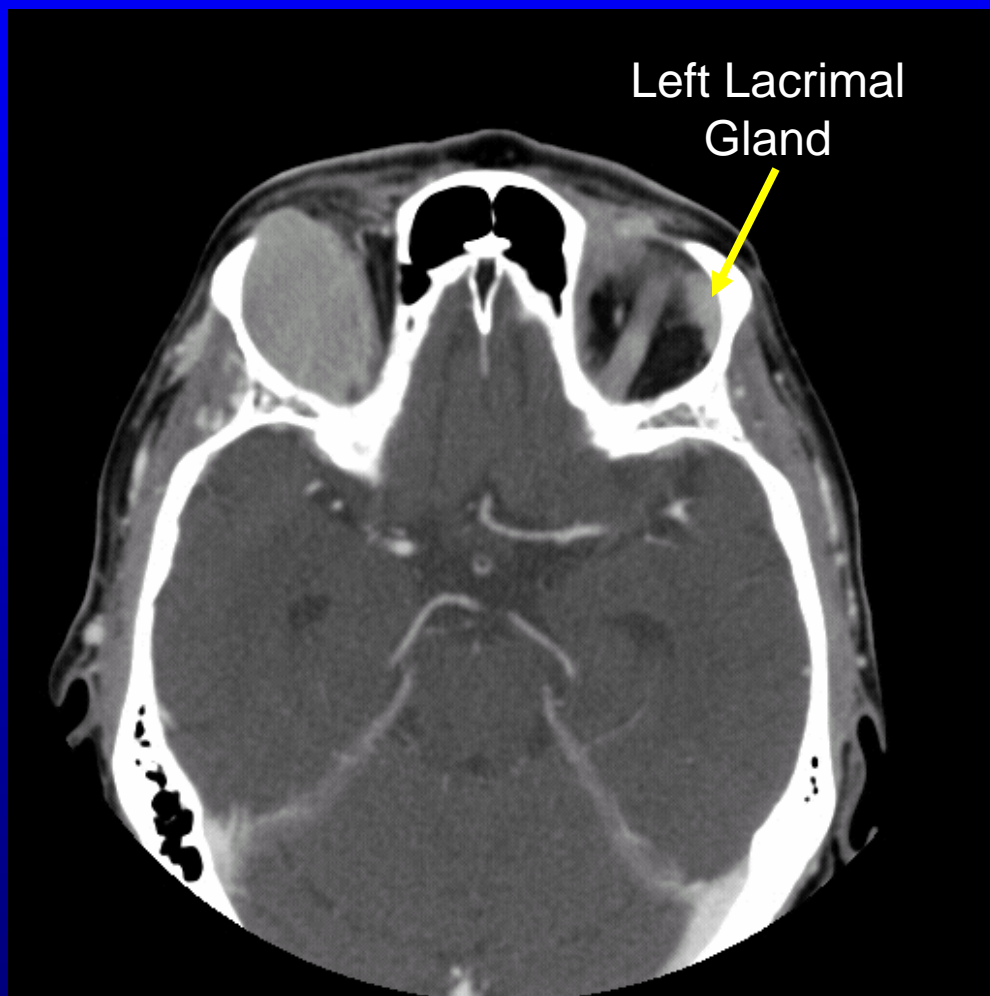




Our Patient's CT Scan of Orbits

(with contrast)

- This is the level of the superior aspect of the orbit.
- There is still no evident bone erosion.
- Right lacrimal gland involvement is likely since it is not visualized in the right orbit unlike in the left orbit.





Differential Diagnosis

Of an orbital mass involving the lacrimal gland:
(when using CT scans)

- **Abscess** – Shows as a fluid-filled mass with an enhancing rim.
- **Lymphoma** – It molds to its surrounding structures with usually no bone erosion.
- **Inflammatory Pseudotumor** – It also molds to its surrounding structures.
- **Sarcoidosis** – It is usually accompanied with extra-ocular manifestations, e.g., lung granulomas.
- **Dermoid** – It is located at suture lines and is fatty in composition and it erodes the local bone.
- **Metastases** – They are rare and they mainly originate from the breast, the lung or the skin. There is also a great variety in their appearance on imaging.



Evaluating the Differentials Using the patient's CT Scan of Orbits

- Abscess:
 - This is unlikely as the mass on the CT scan is homogeneous in its appearance and it has no enhancing rim.
- Lymphoma:
 - This is possible as the patient's mass is encasing the lateral rectus muscle and it is not eroding the local bone.
- Inflammatory Pseudotumor:
 - This is also possible for the same reasons as lymphoma but the patient's proptosis is painless and there is no reticulation of retrobulbar fat (these are usual for pseudotumor).



Evaluating the Differentials Using the CT Scan of Orbits

- Sarcoidosis:
 - It is an unlikely possibility so plain films could be checked for any signs of lung pathology.
- Dermoid:
 - This is unlikely as the mass is not located near the suture lines nor is it fatty in its composition nor is it eroding bone.
- Metastases:
 - It is a possibility but they are rare. Imaging of the breasts, lungs and skin could be done to check for any signs of tumor presence.

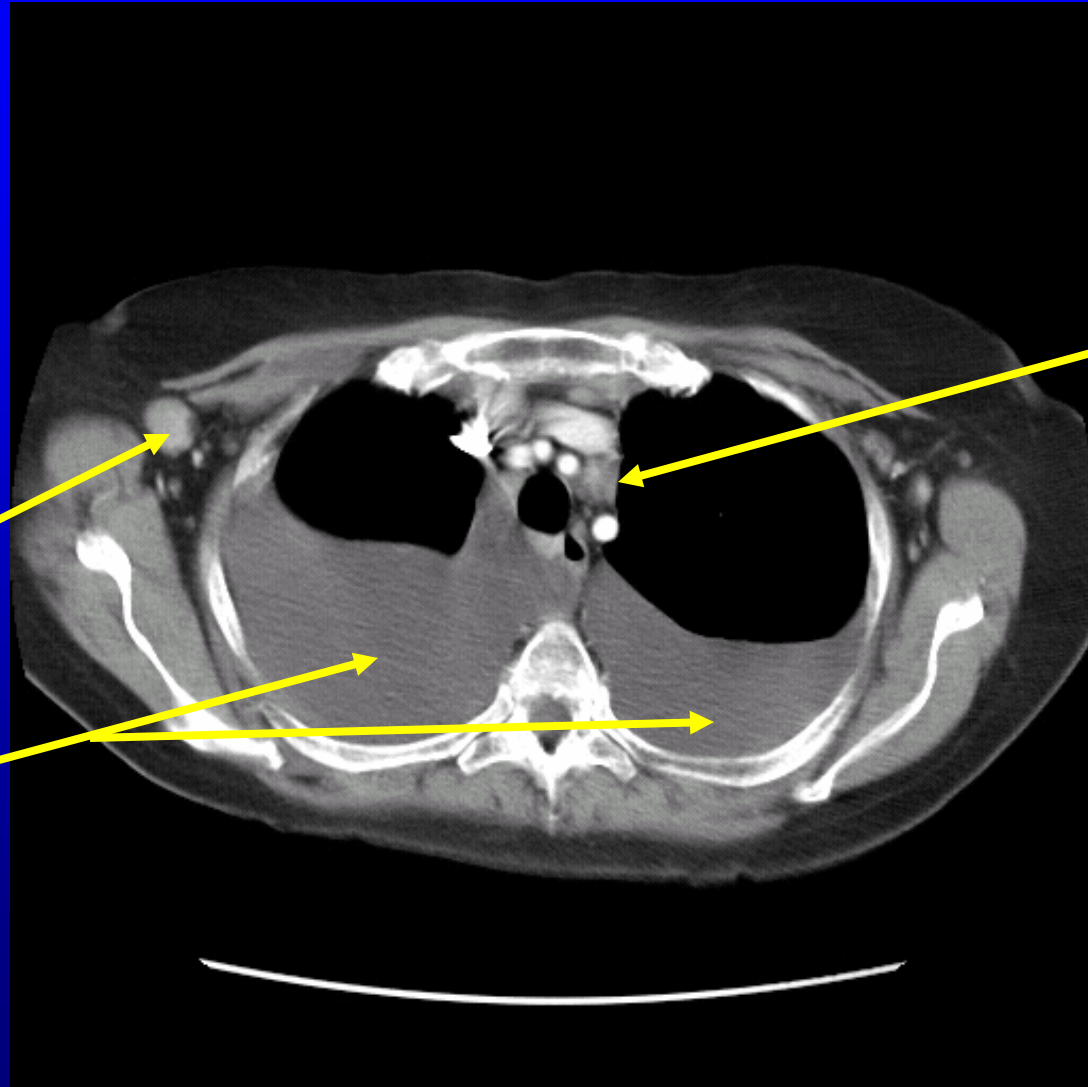


Our Patient's Past Medical History

- Our patient has a past medical history of widespread systemic lymphoma accompanied with left orbit involvement.
- Her Pathology results from her pleural effusion came back as:
 - **Grade I Follicular Center Cell Lymphoma**
(this is a type of Non-Hodgkin's Lymphoma)
- She then went on to have 6 cycles of chemotherapy (cytoxan, vincristine & prednisone) as well as left orbit irradiation which created a left sided cataract which has since been replaced with an intra-ocular lens.



Our Patient's Body CT Scan (With Contrast) Before Treatment



Mediastinal
Lymphadenopathy

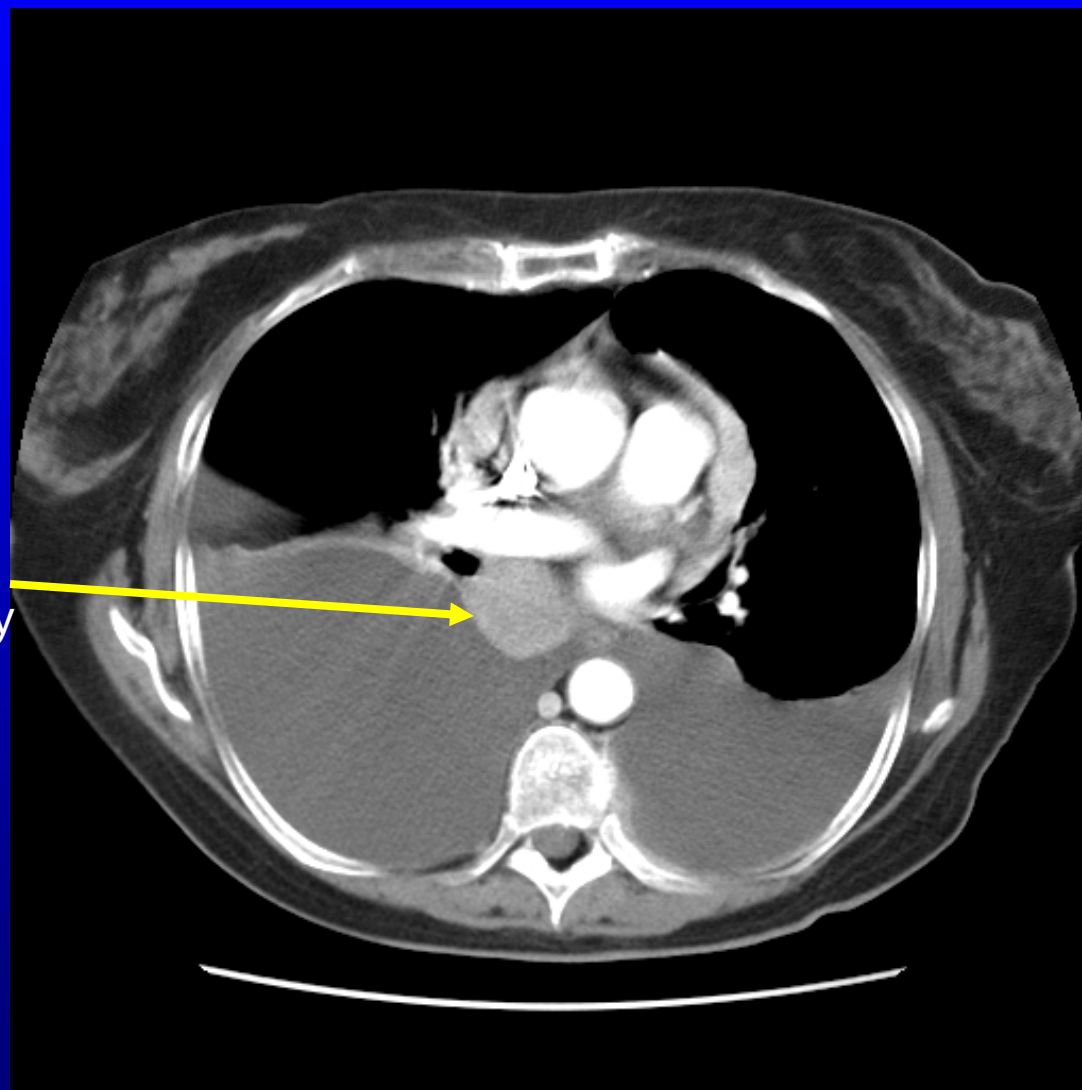
Enlarged Axillary
Lymph Node

Widespread
Pleural Effusion

PACS, BIDMC (Axial CT scan, C+)



Our Patient's Body CT Scan (With Contrast) Before Treatment

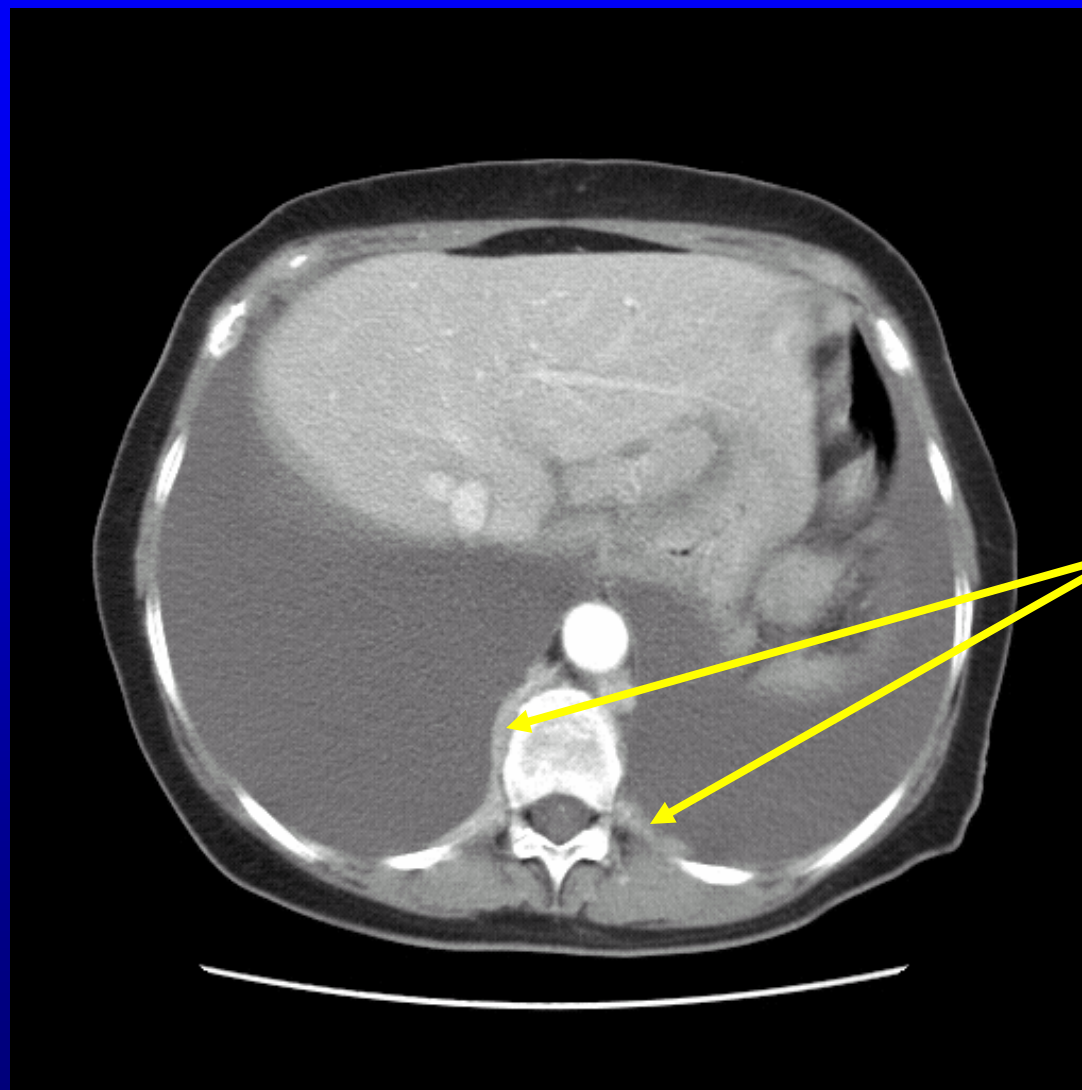


Sub-Carinal
Lymphadenopathy

PACS, BIDMC (Axial CT scan C+)



Our Patient's Body CT Scan (With Contrast) Before Treatment

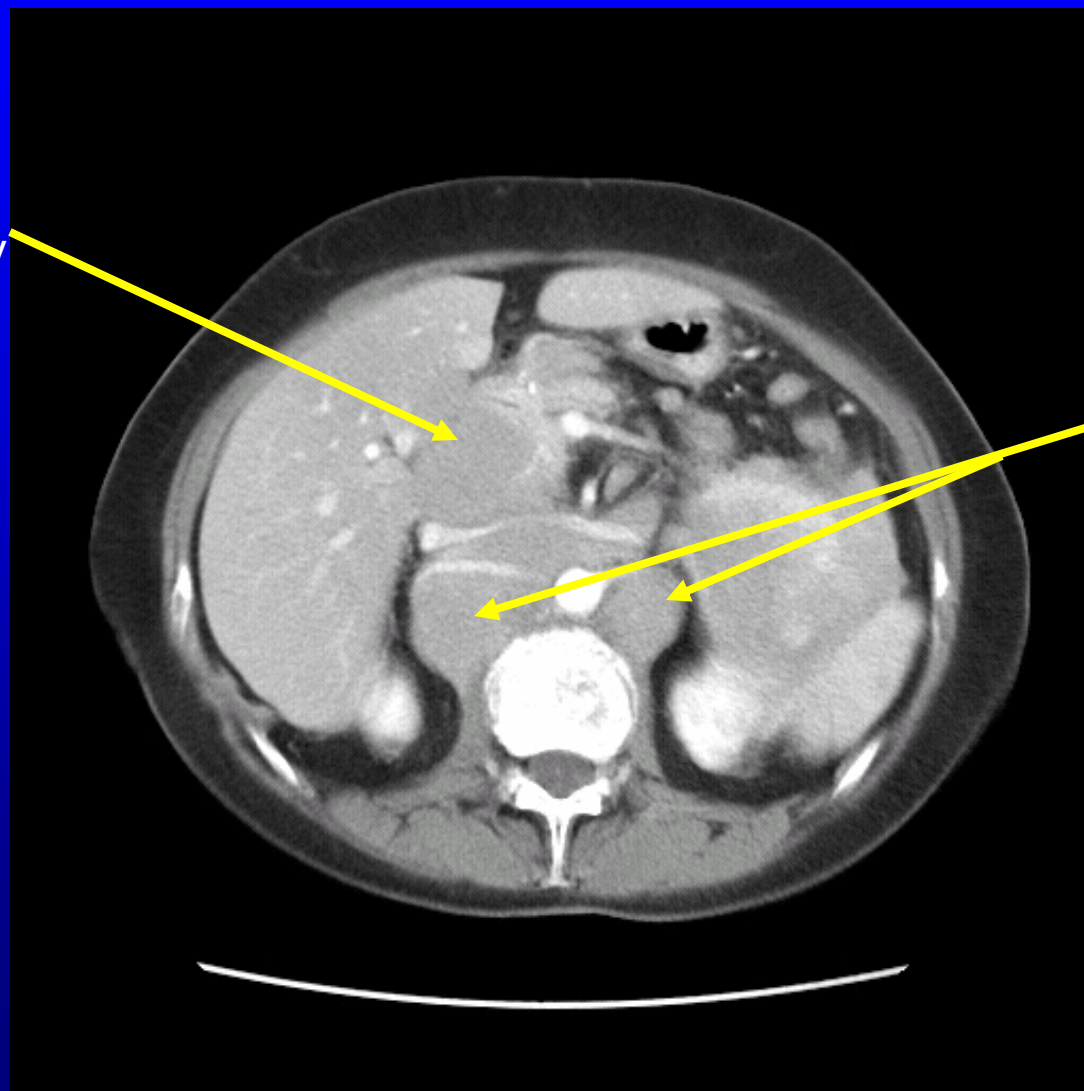


Bilateral Pleural
Thickening

PACS, BIDMC (Axial CT scan, C+)



Our Patient's Body CT Scan (With Contrast) Before Treatment



Peri-Portal
Lymphadenopathy

Para-aortic
Lymphadenopathy

PACS, BIDMC (Axial CT scan, C+)



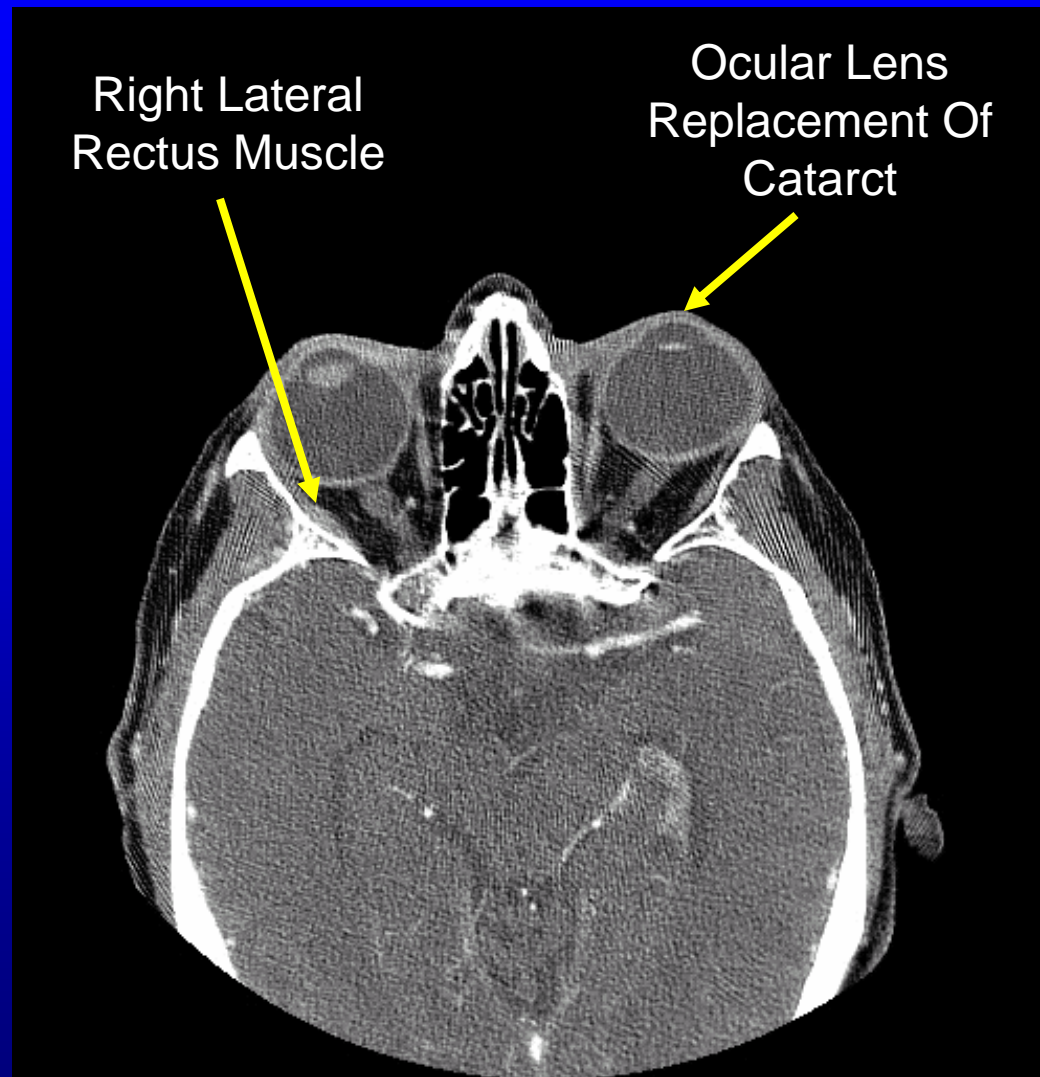
Course Of Our Patient

- Given our patient's past medical history of lymphoma with left orbit involvement, it was decided as highly likely that the right orbital mass was most likely to be a recurrence of her lymphoma.
- A biopsy was not taken after considering its risk of complications.
- To treat her right orbital lymphoma she went on to have 3 further cycles of chemotherapy (fludarabine and cyclophosphamide).
- Following treatment, her right orbital lymphoma went into complete remission!
- She is now on 'Rituximab' maintenance therapy and is now symptom-free enjoying 3 rounds of 18-hole golf a week!



Our Patient's CT Scan of Orbits After Treatment (with contrast)

- There is no evidence of any remaining tumor in the right or left orbits
- There is no evidence of any bone erosion in the right or left orbits.
- The right globe is no longer being displaced.
- The lateral rectus muscle can be clearly seen in both orbits in contrast to its incasement before treatment.
- An intra-ocular lens replacement can be seen in the left eye.





Our Patient's CT Scan of Orbits After Treatment (with contrast)

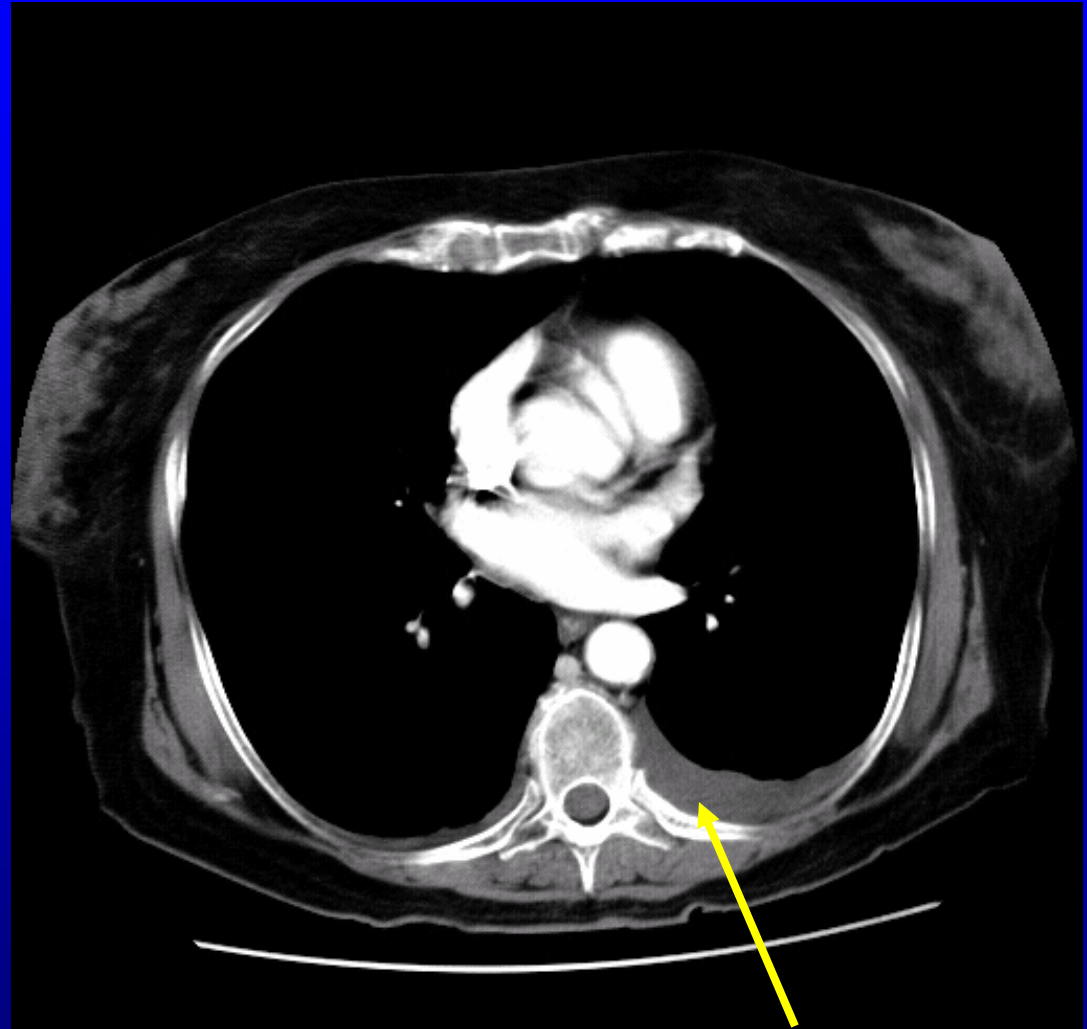
- No tumor is visible in the superior aspect of the orbit.
- The right lacrimal gland is no longer involved in any pathology.





Our Patient's Body CT Scan After Treatment

- There has been an extensive reduction in chest pathology. Virtually all the pleural effusion has resolved.
- The sub-corinal and mediastinal lymphadenopathy have resolved.

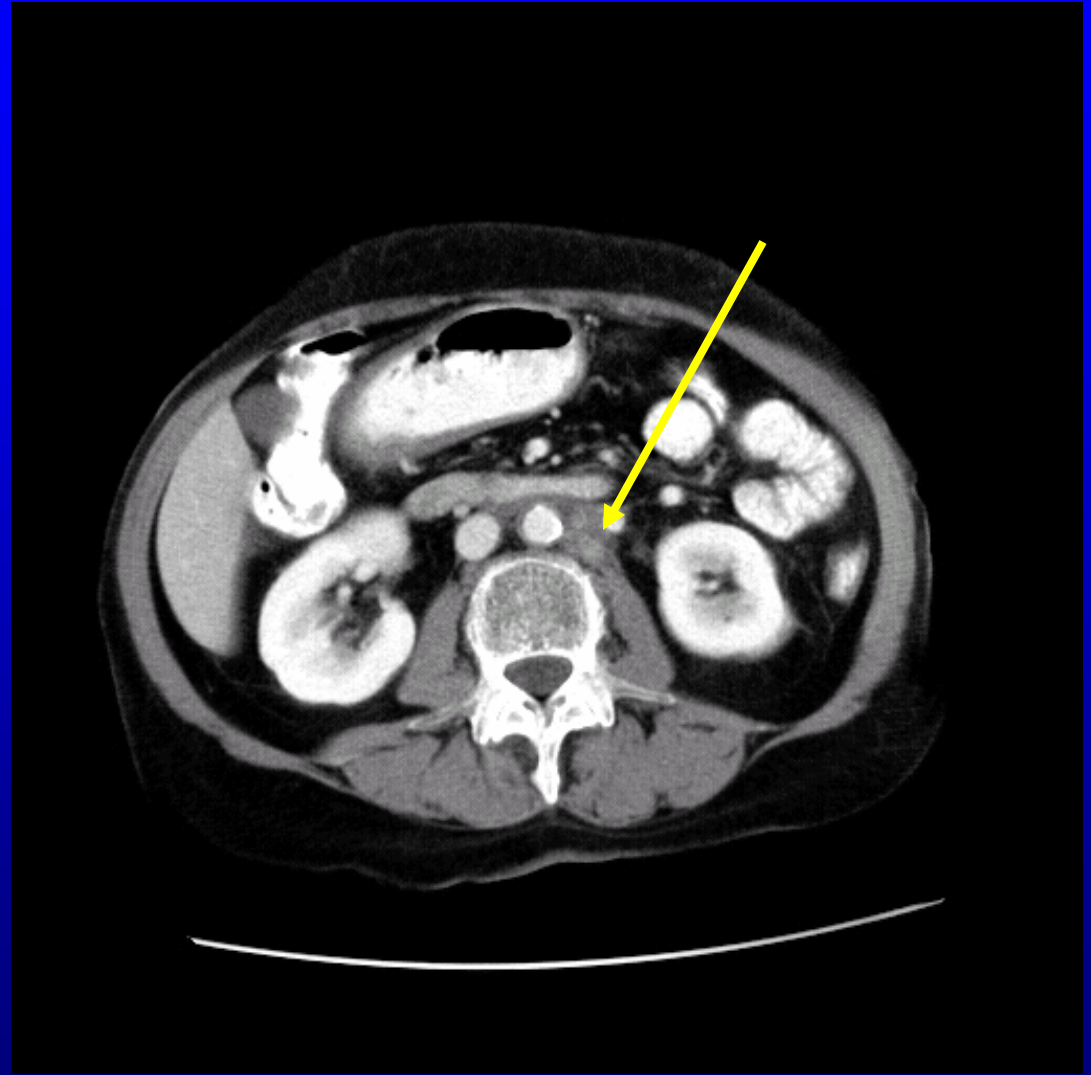


Pleural Fluid



Our Patient's Body CT Scan After Treatment

- Some para-aortic lymphadenopathy remains.
- The majority of the prior lymphadenopathy has resolved however.
- This is a good demonstration of how effective chemotherapy can be in treating lymphoma.





Notes On Orbital Lymphoma

- Incidence:
 - 75% of patients with orbital involvement have systemic lymphoma¹, therefore image other sites, e.g., the neck, chest and abdomen, for any lymphoma.
 - The incidence increases with advancing age².
 - There is no sex predilection².
- Classifications:
 - Subconjunctival involving
 - Lacrimal gland involving
 - Orbital involving (usually presents with proptosis)
 - Eyelid involving (usually presents with ptosis)
 - (our patient had both lacrimal gland and orbital involving)



Notes On Orbital Lymphoma

- Location:
 - Intra-conal (within the cone created by the extra ocular muscles, EOMs).
 - Extra-conal (outside the cone of EOMs).
 - Orbital lymphomas are mostly located in the anterior and superior aspects of the orbit.
- Clinical Symptoms:
 - Insidious onset
 - Diplopia (double vision)
 - Proptosis (bulging of the eyeball out of the socket)
 - Painless presentation (pain is more common with pseudotumor)



Notes On Orbital Lymphoma

- Imaging Characteristics of Orbital Lymphomas:
 - Usually no osseous destruction
 - except rarely for some malignant tumors
 - instead it molds to surrounding structures
 - Orbital lymphomas are:
 - Hyperdense relative to fat on CT³.
 - Hypointense on T1-weighted MRI³.
 - Isointense to extra ocular muscle on T1 & T2-weighted MRI³.
 - Orbital lymphomas are Gallium and FDG avid.



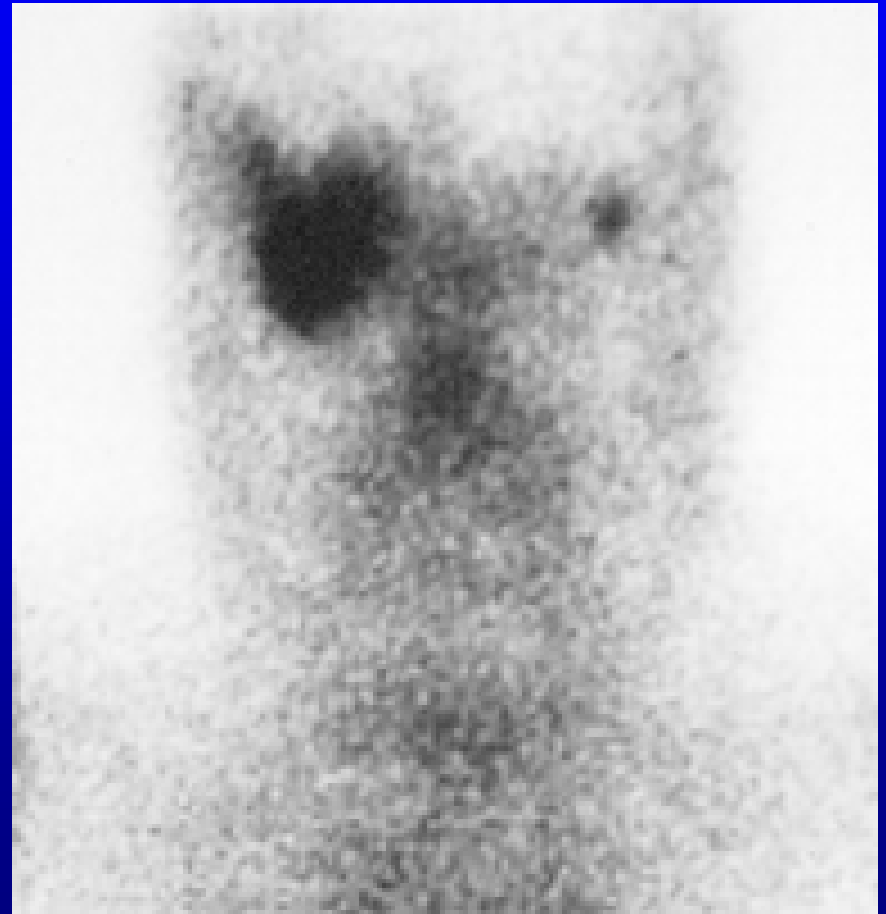
Notes On Orbital Lymphoma

- Treatment:
 - Orbital lymphoma responds well to conventional chemotherapy (using radiation if an adjunct is required but note that its propensity to create cataracts)
- Miscellaneous Facts:
 - Orbital lymphoma types range from benign lymphoid hyperplasia to malignant lymphoma confirmed by a biopsy.
 - Imaging cannot differentiate well between orbital lymphoma and inflammatory pseudotumor however empirical steroid treatment will often be employed followed by a biopsy if the mass does not resolve.



Planar Image of the Head & Neck (Gallium-67 Scintigraphy Scan)

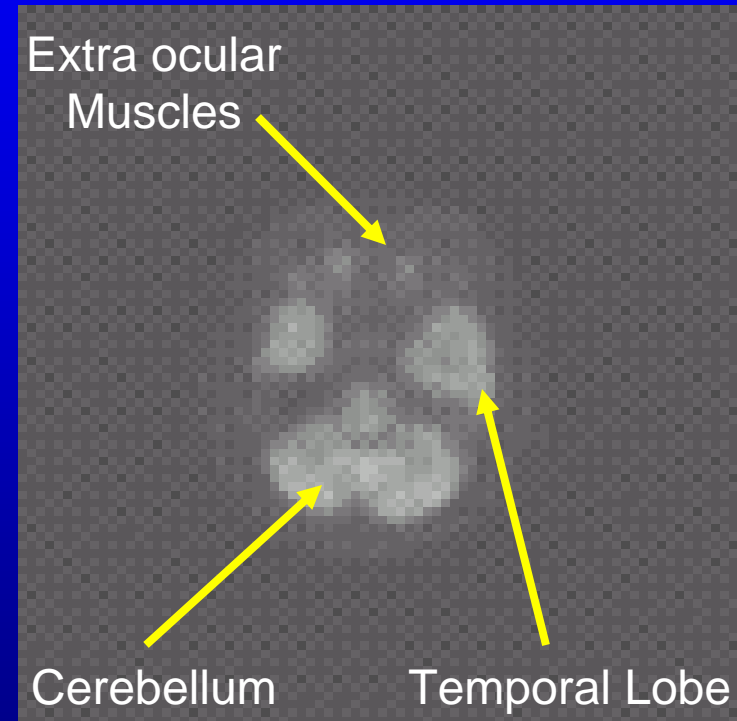
- Obtained from another patient who also had right sided orbital lymphoma
- Shows increased uptake of Gallium-67 limited to the right orbit because of the tumor's preferential uptake.
- This scan is indicated to search the body for any metastases and to monitor for tumor presence.





Our Patient's PET Scan After Treatment (With FDG, Fluorodeoxyglucose)

- This scan of Positron Emission Tomography uses Fluorodeoxyglucose which highlights metabolically active tissue.
- The cerebellum and temporal lobes are very active during the scanning procedure and hence are lighting up.
- The extra-ocular muscles are also active and also light up. The scan confirms no residual tumor remains.



PACS, BIDMC



Summary

- On a CT scan, orbital lymphoma is seen to mold to its surrounding structures with usually no bone destruction.
- 75% of patients with lymphoma in the orbits will also have lymphoma at other sites so these must be imaged following presentation (e.g., the neck/chest/abdomen).
- Imaging cannot differentiate well between orbital lymphoma and inflammatory pseudotumor however empirical steroid treatment will often be employed.
- Orbital lymphomas are very responsive to conventional chemotherapy treatment, which is shown by our patient who is symptom free and continues to lead a happy life.



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