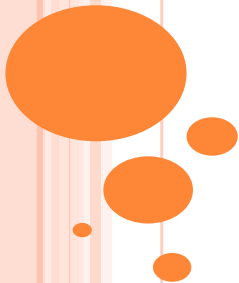




DIFFUSE ORBITAL LYMPHANGIOMA

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□ 10 years old girl presented by RT progressive proptosis 3 months ago.

O/E;

- RT dystopia
- Palpable cystic masses around the globe
- VA: 3/60
- Mild exposure keratopathy
- IOP: 27 mmhg
- Normal fundus apart from optic neuropathy



DD

- Lymphangioma
- Orbital Cellulitis : acute course ,mostly preceded by trauma or nearby infection , associated with fever , all these are not present in our case.
- Rhabdomyosarcoma : typical for age group , Rapidly progressive , firm in consistency , sometimes painful , common in the upper outer quadrant .
- Lymphoma :The typical presentation is low-grade proptosis with minimal pain, sometimes bilateral in systemic disease.

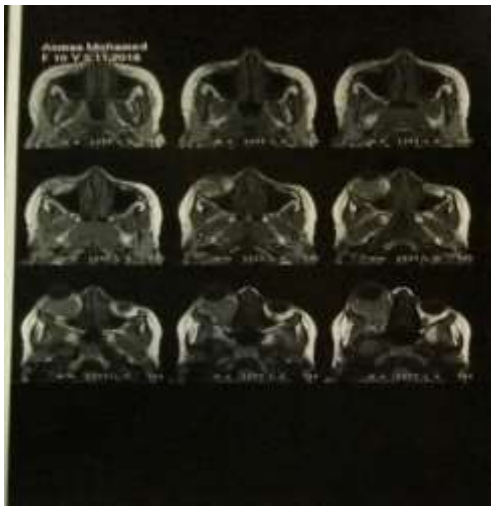


Typical for age

- Optic nerve glioma
- Metastatic tumours i.e. neuroblastoma
- Langerhans cell histiocytosis
- Traumatic retrobulbar hemorrhage

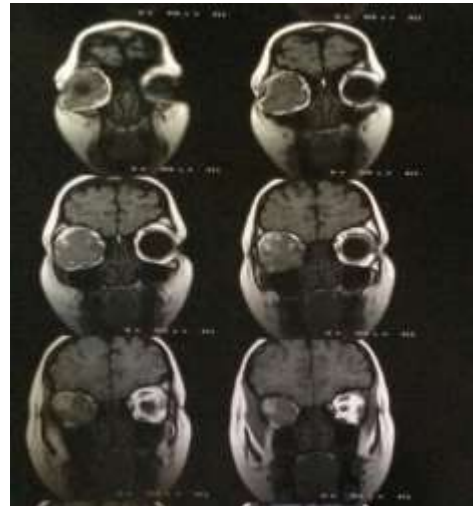
Not common at this age

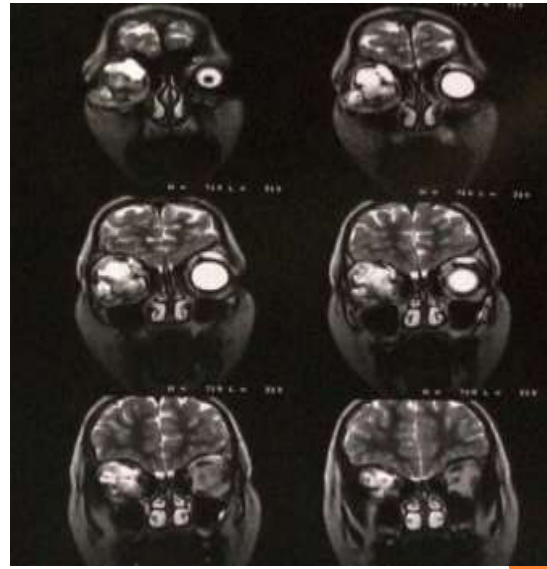
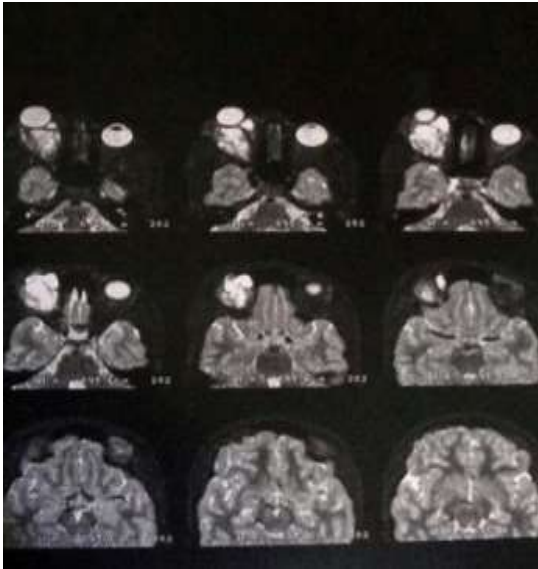
- Optic nerve sheath meningioma
- Hemangioma
- Orbital pseudotumor
- Foreign body
- Ruptured dermoid cyst
- Subperiosteal abscess
- Orbital myositis
- Thyroid orbitopathy



MRI

- Amultilobulated cystic mass
- Isointense to slightly hyperintense on T1
- Hyperintense in T2





❑ ***Lymphangioma*** is rare benign tumor, represents 1 to 3 % of all orbital masses.

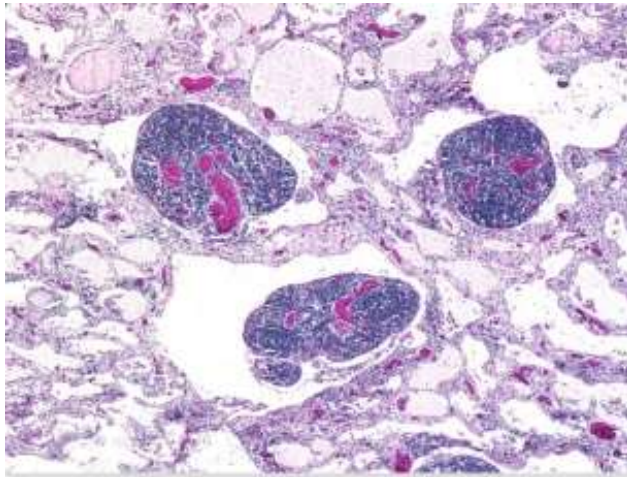
❑ Common presentation is either gradual proptosis , acute proptosis after trauma or upper respiratory infection

❑ Lymphangioma tends to start in the superior and nasal orbital quadrants.

❑ Typically, the lymphangioma bleeds into itself causing chocolate-cysts, If the cyst forms behind the eye, it pushes the eye forward.

❑ If the tumor in the eyelid or adnexa ,lymphangiectasias can be seen beneath the conjunctiva.

- Pathologic examination of a lymphangioma reveals a nonencapsulated lesion with variably sized cystic spaces lined by flattened endothelial cells. Pericytes and smooth muscles are absent from the vessel wall.
- Although the lumens are not continuous with the orbital circulation, there are small nutrient vessels within the fragile walls, which may be the source of hemorrhage.



The lymphatic spaces are filled with **proteinaceous fluid** containing **lymphocytes** and **red blood cells**. The cystic spaces are separated by **delicate stroma** which contains several **dense lymphoid aggregates**.

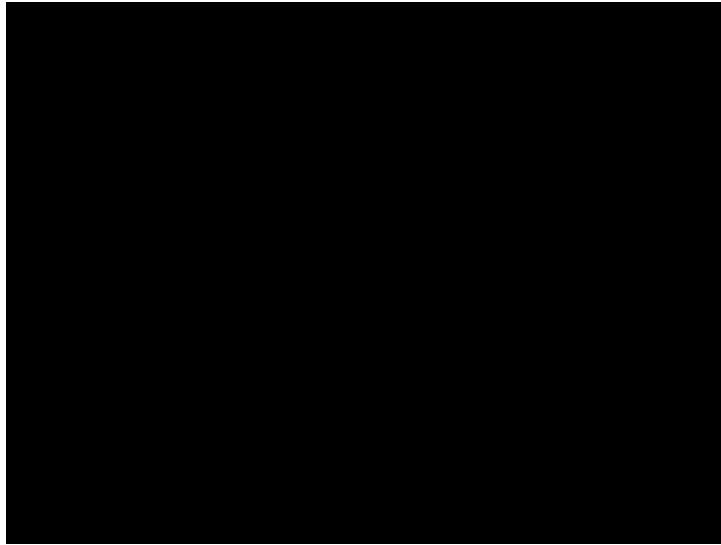
Indications of treatment

- Disfiguring proptosis
- Optic nerve compression
- Exposure keratitis
- Glaucoma

Options of treatment

1. Observation , if no indication of treatment
2. Medically by oral corticosteroids :Symptomatic improvement
3. Needle decompression of cysts with imaging guidance ,
Recurrence
4. Surgical excision , it is actually debulking
5. Intralesional injection of sclerosant material as bleomycin ,
ethanol 98%

Surgical excision



2 months
after sugery



Aspiration with bleomycin injection



What Is bleomycin ????

- Is a cytotoxic drug
- Used in the treatment of squamous cell cancers, melanoma, sarcoma, testicular and ovarian cancer, Hodgkin's, and non-Hodgkin's lymphoma, malignant pleural effusion
- Is mainly indicated for macrocystic lymphangioma
- Has been used since the 1970s for sclerosis of lymphangiomas to reduce the need for surgical excision (MacIntosh PW, Yoon MK, Fay A., Complications of intralesional bleomycin in the treatment of orbital lymphatic malformations., Semin Ophthalmol. 2014 Sep-Nov;29(5-6):450-5.

Side effects of bleomycin

- Fever, transient increase in size of the tumor, hemorrhage, leukocytosis, infection and pulmonary fibrosis.
- Pulmonary fibrosis has been associated with intravenous bleomycin administration exceeding the total cumulative dose of 400 mg. Bleomycin doses used in sclerotherapy are small in comparison, typically 1% to 5% of the lowest dose associated with possible pulmonary fibrosis. (V Kumar, P Kumar, I A Pandey, D K Gupta, R C Shukla, 2 S P Sharma, and A N Gangopadhyay , Intralesional Bleomycin in Lymphangioma: An Effective and Safe Non-Operative Modality of Treatment , J Cutan Aesthet Surg. 2012 Apr-Jun; 5(2): 133–136.)

Preparation

- ❑ Bleomycin for injection is available in vials containing 15 mg powder. It was diluted first with 5 ml distilled water to prepare the stock solution which can be stored for 60 days at 4–8°C.
- ❑ Maximum allowed dose in a single session is 0.3 to 0.5 mg/kg
- ❑ The maximum allowed cumulative dose is 5 mg/kg.

Z Rozman,¹ RR Thambidorai,² AM Zaleha,³ Z Zakaria,⁴ and MA Zulfiqar., Lymphangioma: Is intralesional bleomycin sclerotherapy effective?, Biomed Imaging Interv J. 2011 Jul-Sep; 7(3): e18. Published online 2011 Jul 1.

Mechanism of action

- Inhibition of DNA synthesis
- Sclerosing effect due to its direct action on the endothelial cells producing non-specific inflammatory reaction. Desired effect of sclerosis is achieved by local action of bleomycin, which depends on availability of drug per unit surface area of lesion.

Under the effect of sedation (oral chloral hydrate with IV pethidine added when necessary) and local anaesthetic and with strict aseptic precaution, the cystic components of the tumour were aspirated with 23G needle. While keeping the tip of aspiration needle within a cyst lumen, bleomycin aqueous solution was injected.



We injected 2 times with 4 weeks interval, we injected 8 mg in the first session, 6 mg in the second session, with no reported complications during injection or in the follow up period.





TODAY



