CLINICAL HISTORY

• 12 year-old female previously healthy
• Referral for evaluation of orbital mass “inflammatory pseudotumor”
• Symptoms started 3 years earlier
  • Right side proptosis
  • Visual changes
Orbital biopsies – Oct 2012
PATHOLOGY

• Battery of immunostains
  • CD163, CD68 (macrophages)
  • S100, CD34, CD1a and Alk-1
    • All negative
  • Orbital fibroadipose tissue with marked chronic inflammation

CLINICAL HISTORY

• Patient was treated with steroids
  • Good clinical and radiographic response
• Vision declined – tapering of steroids
• Infliximab – following 4 months
• Jun 2013 (6 m) – right 6th nerve palsy
• Jan 2015 - Right side hearing loss
Nasopharynx and Parapharyngeal mass Biopsies

Parapharyngeal mass biopsy

**PATHOLOGY**

- Extensive battery of immunostains
  - CD3, CD4, CD8, CD10, CD15, CD20, CD21, CD30, CD34, CD35, CD46, CD56, CD68, CD163, PAX-5, PGM1, ALK-1, calponin, cytokeratins (AE1/AE3 and 19), factor 13A, fascin, PGP9.5, Bcl-2, Kappa and Lambda, S100
- Lymphoma was ruled out
- Descriptive diagnosis – “pseudotumor”
CLINICAL HISTORY

- July 2015 - radiation therapy
  * 20 cGy
- Sept 2015 – left arm and leg weakness + sensory changes
- Brain and Spine MRI – new lesions

Brain MRI - FLAIR

October 2015

CLINICAL HISTORY

- Extensive systemic work-up – Neg
  * Infectious etiology
  * Autoimmune disease
- Steroid therapy
  * Improvement of symptoms
- Patient referred to Mayo for second opinion (Nov 2015)
CLINICAL HISTORY

2012
- Proptosis
- Visual changes
  June 13
  6th nerve palsy

2016
- April 15
  Parapharyngeal mass
- Nov 15
  Referred Mayo
  Steroids - Infliximab
  Radiation

- Jan 15
  Hearing loss
- Sep 15
  Arm & leg weakness

DISCUSSION

- Challenging case
- Patient of young age
- Unusual clinical course
- Extensive disease
- Diagnosis
- Management of disease
DISCUSSION

• Differential diagnosis
  1. Inflammatory process
     • Infectious etiology
  2. Lymphoma
  3. Neoplasm with exuberant inflammatory component
  4. Inflammatory process
     • Non infectious etiology

ORBITAL CELLULITIS

Differential Diagnosis

• Infection of soft tissues posterior to orbital septum
• Extension of sinuses infection
• More common in children than adults
  • Males > females
  • Median age (hospitalization) – 7-12 yrs

[References]

[Images]
Case of orbital cellulitis

[References]
ORBITAL LYMPHOMA
Differential Diagnosis

• Uncommon in children
  • Large series* – median age 69 yrs
• Burkitt lymphoma
• Extranodal marginal zone lymphoma
• Orbital involvement by leukemia
• Immunophenotyping is necessary

*Demirci H et al. Ophthalmol 2008; 115(9):1626-31
Rasmussen PK et al. JAMA Ophthalmol 2014; 132:851-8

NEOPLASMS WITH INFLAMMATION
Differential Diagnosis

• Neoplasms associated with marked inflammatory infiltrate
  • Rhabdomyosarcoma
  • Retinoblastoma extending to the orbit

Case of orbital Burkitt Lymphoma

Immunophenotype by flowcytometry

Clinical picture courtesy of Dr. James Garrity
RHABDOMYOSARCOMA

NON INFECTIOUS INFLAMMATORY ORBITAL LESIONS

- Lesions with specific features
  - Sarcoidosis
  - Granulomatosis with polyangiitis
  - Rosai-Dorfman disease
  - Orbital IgG4-related disease
- Lesion without specific features
  - Idiopathic orbital inflammation

Granulomatosis with Polyangiitis (Wegener’s)

21 years old woman
Bilateral proptosis

Rosai-Dorfman Disease

15 years old woman
Bilateral orbital masses
Histiocytic disorder
CD68+, S100+, CD1a-
**IgG4-related disease**

52 years old woman
Right blurred vision and diplopia
Enlarged right lacrimal gland

Consensus conf. criteria
- 100 IgG4+ cells/hph
- IgG4/IgG ratio >40%

Deshpande V et al. Mod Pathol 2012; 25:1181-1192

**NON INFECTIOUS INFLAMMATORY ORBITAL LESIONS**

- Lesions with specific features
  - Vasculitis
  - Sarcoidosis
  - Rosai-Dorfman disease
  - Orbital IgG4-related disease

- Lesion without specific features
  - Idiopathic orbital inflammation

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**Idiopathic Orbital Inflammation**

- Diagnosis of exclusion
- Cases with typical presentation
  - Middle age patient, pain
  - Steroid test
  - No biopsy
- Unusual presentations
  - Tissue biopsy
Idiopathic Orbital Inflammation

Sclerosing type

Idiopathic Orbital Inflammation

Children

- Uncommon diagnosis in pediatric population
  - 6-17% of all orbital lesions
- Largest series – Mottow and Jakobiec
  - 29 patients < 20 yrs
    - Systemic symptoms – 55%
    - Bilateral involvement – 45%
    - Eosinophilia – 28%


Idiopathic Orbital Inflammation

Extra-orbital extension

- Uncommon – < 50 cases reported
  - Middle cranial fossa, cavernous sinus
- Extension outside the orbit
  - Superior orbital fissure
  - Optic canal
  - Inferior orbital fissure
- Patients required additional therapy

Zborowska et al. Eye 2006; 20:107

PRESENT CASE

- Last follow-up – recent
  - Improvement of disease in orbit
  - Vision loss in right eye remains unchanged
  - Improvement of spine and brain lesions
PRESENT CASE
• Steroid discontinued
• Scheduled to return in 2 months

SUMMARY
• Case of idiopathic orbital inflammation
• Young patient (9 years at presentation)
• Extra-orbital extension
• Resistant to steroids
• Brain lesions of unknown etiology

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