Tumefactive Demyelination

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Background

- Tumefactive demyelinating lesions (TDL) is an acute, inflammatory presentation of large acute demyelinating lesions (>2 cm) that can be associated with mass effect. TDLs are rare but can mimic brain neoplasms or abscess because of their associated edema within the cerebral hemisphere.
- TDLs are also known as tumefactive multiple sclerosis lesions, tumor-like demyelinating lesions, and demyelinating pseudotumor.
- They primarily affect females, ages 2 through 70 with the average age being 37.

Purpose

- Tumefactive demyelination is a rare neurological disorder that can present with multiple symptoms: cognitive, sensory, motor, visual and cerebellar.
- Patients with visual symptoms may initially present with sub-acute or acute vision loss but may not have any associated neurological symptoms at the time.
- This case will discuss the clinical quandaries of suspecting demyelinating changes.

Case Report

23-year-old white female was referred to the Neuro-ophthalmology clinic at Bascom Palmer Eye Institute.

Chief complaint:

- Mild superior field vision loss OS
- Severe vision loss OD

ROS:

- Neurological: Numbness of left side of face, index finger, thumb, left leg
- Constitutional: Imbalance

Medical history: None

Medications: None

Family history:

- Father: DM, Heart disease.
- Mother: Lung disease

Examination:

- Infrared Color Plates: 1/11 OD, 1/11 OS
- Anterior segment unremarkable

Imaging:

- [MRI images showing lesions and contrast]

Discussion

Vision loss, visual field loss, decreased color vision and pallor of optic nerves warrants an urgent work-up, both ophthalmic as well as systemic.

- ODX: optic neuritis/ optic neuropathy
  - ischemic, inflammatory, infectious, compressive, neoplastic or hereditary etiology.
- Inflammatory demyelinating disease spectrum
  - Multiple sclerosis (MS), neuromyelitis optica spectrum disorder (NMOSD), acute disseminated encephalomyelitis (ADEM)
  - Within the MS category - tumefactive demyelinating lesions
- Tumefactive lesions may be the initial presentation of relapsing MS seen in about 50-70%.
- TDLs occupy more space and can cause seizures, hemiparesis, neglect or encephalopathy.

Imaging:

- [MRI images of orbits and brain with and without contrast]

- Numerous FLAIR hyperintense lesions with incomplete peripheral rim of enhancement and confluent FLAIR signal.
- Asymmetric increased T2 signal and enhancement of the left optic nerve.
- MRI cervical spine with and without contrast:
  - Multiple cervical cord lesions

Labs of interest:

- ANA: positive
- Myelin Basic Protein 140: high
- CSF: protein 71 H, Glc 61, WBC 0, RBC 0
- Oligoclonal bands: Three (3) oligoclonal bands were observed in the CSF, which were not detected in the serum sample.

Conclusions

In this case of tumefactive demyelination, time may be a factor in getting the best visual outcome. MS can appear with ocular related symptoms early on.

- Clinical presentations can vary from mild symptoms to severe.
- Presentation can vary from typical MS cases.
- In some cases, asymptomatic patients remained untreated and were shown to improve on their own.
- Our patient reported her symptoms worsening over 2–3-month period, justifying treatment.
- Treatment: IV Solumedrol + Plasma Exchange (PLEX)
- It is imperative to have a clear history of patient’s presenting symptoms and associated review of systems.

Follow-Up x 2 weeks

VA 20/70+ 20/70-

References


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**ABSTRACT**

This is a case report of a patient with varicella-zoster infection with a secondary bilateral preseptal cellulitis. This report highlights secondary bacterial infections as one of the possible complications of a varicella infection.

**CASE HISTORY**

Demographics: 67-year-old Asian male

Chief Complaint: Periorbital pain and eyelid swelling OU that began 5 days prior.

Symptoms constant and severe

Ocular History: Unremarkable

Medical History: Unremarkable

Medications: Oral valacyclovir 1000 mg BID prescribed by primary care provider 3 days prior

**EXAM FINDINGS**

Gross Examination: Crusted necrotic vesicular scab-like lesions localized to the right forehead and right upper eyelid.

Adnexa: Bilateral periorbital edema and erythema OD>OS (Figure 1)

**DIFFERENTIAL DIAGNOSES**

VZV associated with orbital cellulitis, VZV associated with preseptal cellulitis, Severe post-herpetic neuralgia from VZV

**DISCUSSION**

Varicella-Zoster Virus (VZV)

**About**

- A human herpes virus that causes varicella (chickenpox) as a primary infection followed by latency in peripheral ganglion.
- The latent VZV may reactivate to cause herpes zoster (shingles).

**Classic Sign**

- Right or left-sided vesicular rash that follows a characteristic dermatomal distribution.

**Ocular Complications**

- Postherpetic neuralgia, stromal keratitis, giant cell arteritis, vasculopathy, and secondary bacterial infections especially in immunocompromised individuals.

**Treatment and Prevention**

- Oral antivirals and vaccinations

**Preseptal Cellulitis**

**About**

- Infection located anterior to the orbital septum involving periorbital region.
- Most commonly caused by trauma, insect bites or spread of a systemic infection.
- Most common organisms isolated are the Staphylococcus and Streptococcus species. Very rarely, associated with VZV (secondary bacterial infection).

**Demographics and Prognosis**

- More common in childhood and with good prognosis. However, if left untreated can cause severe complications including orbital cellulitis, orbital abscess, and cavernous sinus thrombosis.

**Signs**

- Periorbital erythema, edema, and eyelid swelling. Vision and pupils are usually normal. Typically, no proptosis or ophthalmoplegia that is seen in orbital infections.

**Treatment**

- Oral antibiotics in mild infections, hospitalization and intravenous antibiotics in severe cases.

**REFERENCES**