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## A Rare Case of Tolosa-Hunt Syndrome

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# A Rare Case of Tolosa-Hunt Syndrome

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## Case Description

27-year old female with no past medical history presented to the ED multiple times with intractable headaches and blurry vision. Her symptoms involved a right-sided throbbing pain for 6 weeks, for which she was seen by an outpatient neurologist, ophthalmologist, as well as a dentist. However, now she was experiencing recent onset of blurry vision with increasing right eye pressure. Her headache and visual symptoms improved dramatically in the ED with a dose of solumedrol. MRI/MRV revealed soft tissue thickening of the right cavernous sinus with right proptosis and edema/enhancement of the right superior and lateral rectus muscles as well as the right lacrimal gland (Figures 1, 2, and 3). During her hospitalization, her infectious and autoimmune workups were grossly negative as seen in Table 1. Ophthalmology performed a lacrimal gland biopsy, which demonstrated a vasculitic process with lymphocytic perivascular inflammation. She was discharged home with prednisone 60mg daily for 2 weeks with a planned steroid taper to a goal of 20mg once daily along with neurology follow-up.

## Discussion

Tolosa-Hunt syndrome represents an idiopathic disease process due to nonspecific inflammation in the cavernous sinus region that may also include the superior orbital fissure. Dr. Tolosa described it as “non-specific, chronic inflammation of the septa and wall of the cavernous sinus with the proliferation of fibroblasts an infiltration with lymphocytes and plasma cells.” Hunt et al. further delineated that the inflammation can “exert pressure upon the penetrating nerves,” including CN III, IV, VI, and superior branch of V.

The estimated yearly incidence of Tolosa-Hunt is approximately 1 per million. Average age of onset is 41 years old and sparsely seen in younger populations. There has been no reported predisposition based on gender, race, or geographic location

The International Headache Society guidelines summarizes the diagnostic criteria for Tolosa-Hunt as follows:

- Unilateral headache
- Must include both of the following:
  - Granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit either on MRI or biopsy
  - Palsy of at least one of the ocular nerves III, IV, or VI on the same side

## Conclusion

- Tolosa-Hunt is a rare, inflammatory disorder involving the cavernous sinus that can impinge on surrounding nerves, causing pain and visual disturbances.
- It is a diagnosis of exclusion and requires advanced imaging and laboratory investigations to corroborate the diagnostic suspicion.
- Tolosa-Hunt should be suspected in patients presenting with new-onset, recurrent headaches with or without painful ophthalmoplegia.
- The use of steroids for a patient with subacute headaches and concurrent periorbital inflammation may provide a diagnostic clue for the etiology.

## Differential Diagnosis

- Giant cell arteritis
- Migraine
- Cluster headache
- Pituitary adenoma
- SLE
- Sarcoidosis
- Wegener’s granulomatosis
- Lyme
- Herpes zoster
- EBV
- Intracranial aneurysm
- Metastasis
- Neuro-behcet’s syndrome

## Tables

| Lab test                   | Results   |
|----------------------------|---|
| ESR                        | 32  |
| CRP                        | <3.0  |
| ANA                        | Negative  |
| ds-DNA                     | Negative  |
| Lupus anticoagulant panel  | Negative  |
| Cardiolipin antibodies     | Absent  |
| ENA                        | Negative  |
| c-ANCA                     | <1:20   |
| p-ANCA                     | <1:20   |
| Sjogren’s antibodies       | Negative  |
| Complement 3               | 177 (90-230)  |
| Complement 4               | 42 (10-51)  |
| ACE (serum; CSF)           | 19 (8-52); <5   |
| IgG4 (serum; CSF)          | 31 (4-86);  |
| CSF cytology               | Lymphocytic pleocytosis; negative for malignant cells   |
| CSF count and differential | WBC 39<br>95% lymphocytes<br>Protein 30.7<br>Glucose 67 |
| HSV PCR (CSF)              | Negative  |
| VZV PCR (CSF)              | Not detected  |
| CMV PCR (CSF)              | Not detected  |
| EBV PCR (CSF)              | Not detected  |
| Lyme antibodies (CSF)      | 0.13 (<0.99)  |
| Bacterial culture (CSF)    | No growth   |
| Fungal culture (CSF)       | No growth   |
| AFB culture (CSF)          | No acid-fast bacilli seen                               |

Table 1. Laboratory tests and corresponding patient results

## Advanced Imaging

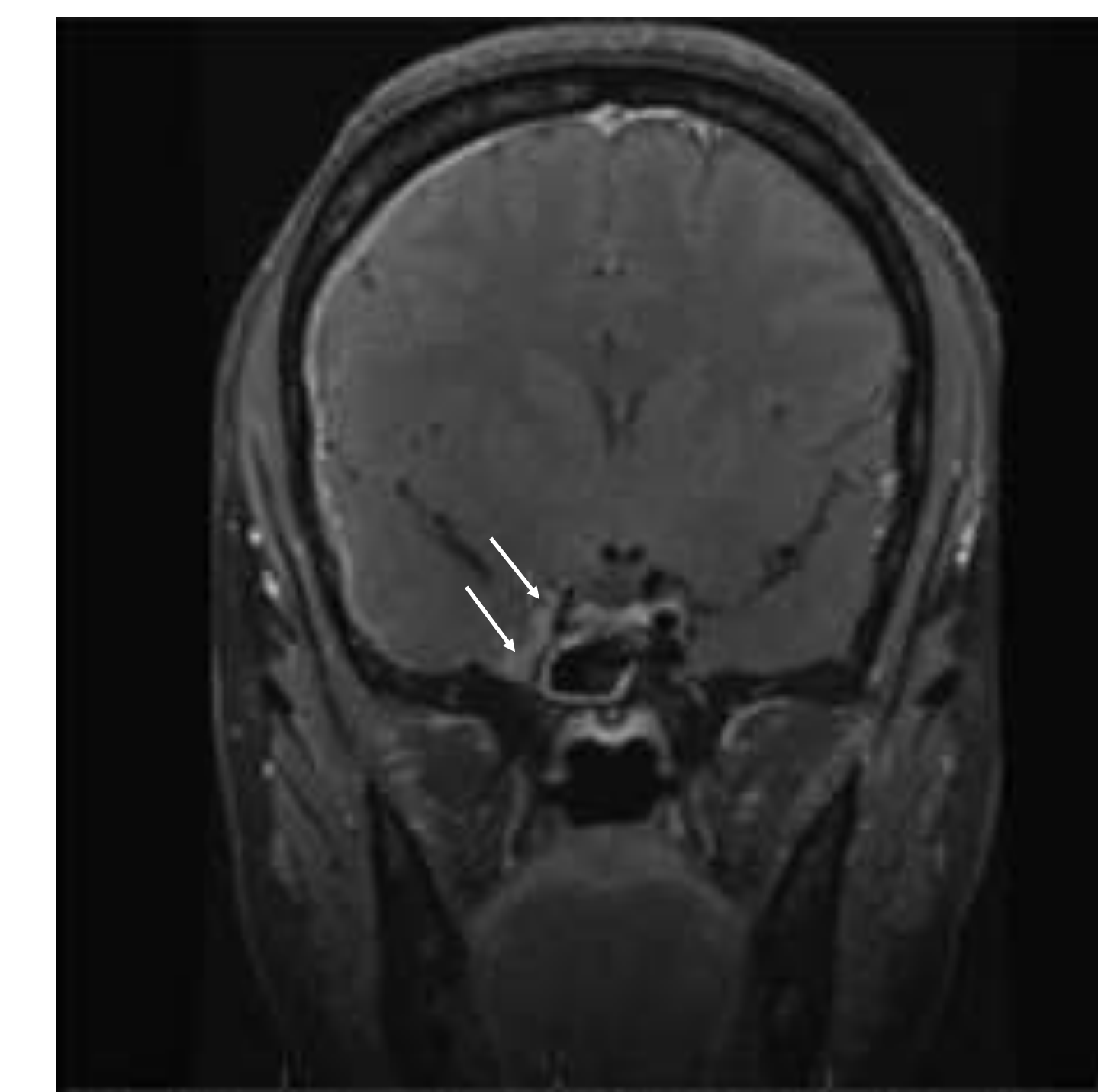


Figure 1. Inflamed cavernous sinus

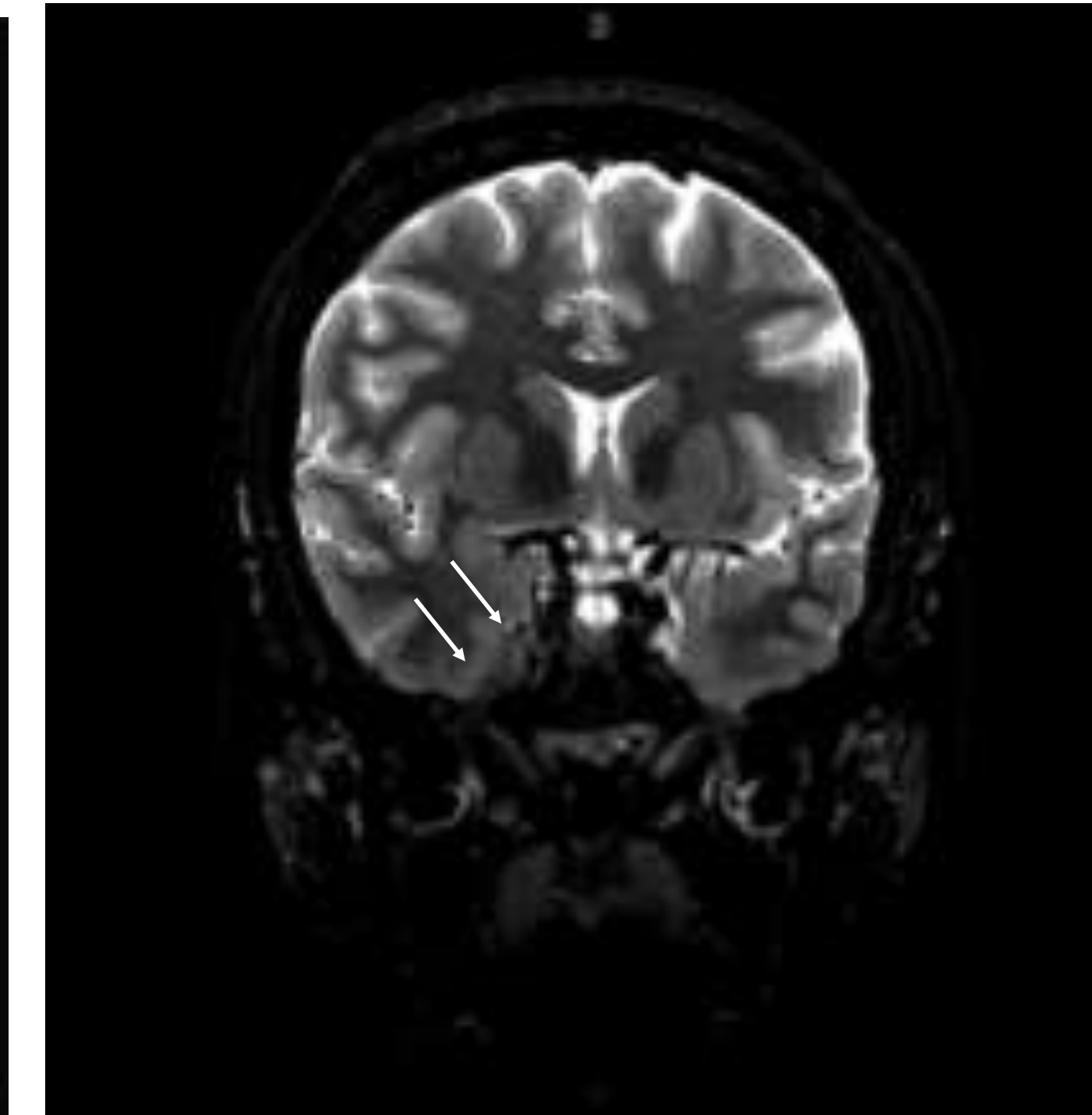


Figure 2. Redemonstration of inflamed cavernous sinus

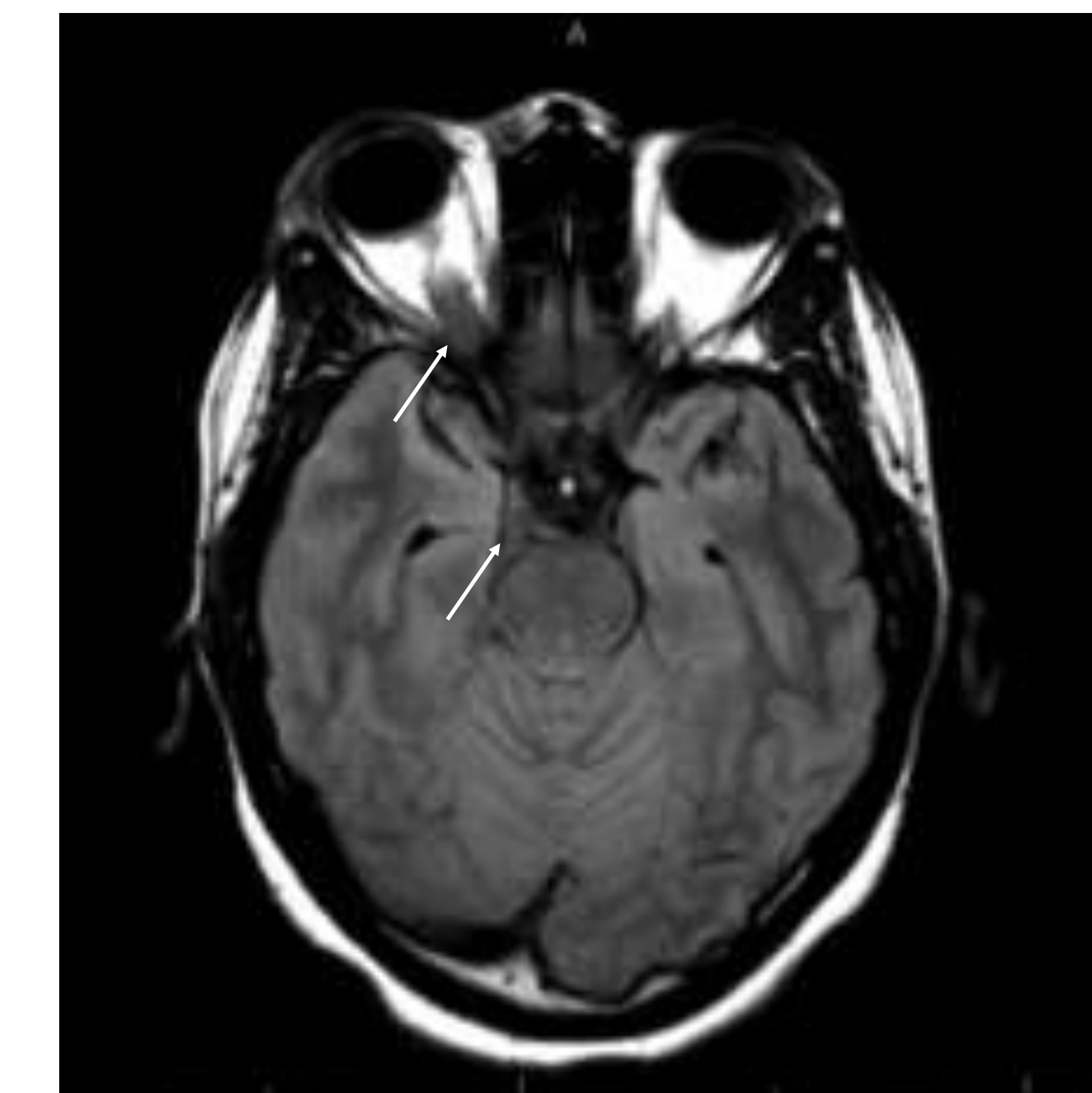


Figure 3. Superior and lateral rectus edema with inflamed cavernous sinus

## Citations

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