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Case Report

When Fire Meets Shadow: A Rare Case of Tolosa-Hunt Syndrome Associated with Discoid Lupus Erythematosus

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Abstract

Background: Tolosa-Hunt Syndrome (THS) is a rare inflammatory disorder presenting with painful ophthalmoplegia due to granulomatous involvement of the cavernous sinus or superior orbital fissure. Though autoimmune diseases such as systemic and discoid lupus erythematosus (SLE/DLE) are known to overlap with other disorders, their association with THS remains poorly documented. DLE, a chronic photosensitive condition with scarring skin lesions, is particularly rare in conjunction with THS. This report explores a case of THS associated with DLE, highlighting diagnostic complexities and therapeutic strategies. **Case:** A 54-year-old woman presented with right-sided headache, diplopia, and ocular pain. Examination revealed right third cranial nerve palsy, hyperpigmented macular lesions, and alopecia areata. Imaging demonstrated cavernous sinus inflammation, and laboratory findings included elevated ESR, positive dsDNA titers, and a homogeneous immunofluorescence pattern. A biopsy confirmed DLE, aligning with clinical and imaging findings of THS. The patient was treated with corticosteroids and mycophenolate mofetil, resulting in sustained symptom resolution with no relapse during follow-up. **Conclusion:** This case underscores the need to consider autoimmune conditions like DLE in patients with THS, suggesting a possible shared autoimmune mechanism. Early recognition and timely initiation of immunosuppressive therapy with corticosteroids and mycophenolate mofetil were key to achieving remission, supporting their use as first-line treatment. This report adds to the limited literature on DLE-associated THS and highlights the importance of thorough diagnostic evaluation and long-term follow-up to monitor progression and prevent recurrence. Additional reports are needed to improve understanding of the pathophysiology, clinical features, and optimal management of these rare coexisting conditions.

Introduction

Tolosa-Hunt Syndrome is a rare condition characterized by painful ophthalmoplegia, involving the third, fourth, and/or sixth cranial nerves. It is caused by nonspecific inflammation in the cavernous sinus or superior orbital fissure, though the exact cause remains unknown.[1]

Autoimmune diseases, such as systemic lupus erythematosus (SLE) and discoid lupus erythematosus (DLE), present with a range of systemic and cutaneous manifestations, with some reports suggesting an overlap of lupus with THS. Discoid lupus erythematosus (DLE), the most common form of cutaneous lupus erythematosus, is a chronic, scarring, photosensitive autoimmune skin condition characterized by erythematous and scaly lesions.2 Although THS has been reported with other autoimmune conditions, its coexistence with DLE is rare and poorly documented in the literature. To date, only one published report has described an association between THS and DLE, further emphasizing the rarity of this presentation.

This case report details a unique presentation of THS associated with DLE, highlighting the diagnostic challenges and therapeutic approach. The report emphasizes the importance of thorough differential diagnosis, including autoimmune and granulomatous disorders, and underscores the role of immunosuppressive therapy in achieving symptomatic relief and preventing recurrence in such rare, complex cases. Additionally, this case

Highlights:

- Uncommon Association: Demonstrates the rare coexistence of Tolosa-Hunt Syndrome (THS) and Discoid Lupus Erythematosus (DLE), contributing to limited existing literature.
- Clinical Challenge: Highlights the complexity of diagnosing overlapping autoimmune and neuro-ophthalmic disorders, requiring careful evaluation and integration of clinical, imaging, and biopsy findings.
- Therapeutic Insights: Shows the efficacy of combining corticosteroids with mycophenolate mofetil, providing practical treatment guidance for similar cases.
- Broadens Understanding: Expands knowledge of DLE's systemic manifestations, specifically its involvement in cranial neuropathies and painful ophthalmoplegia.
- Educational Value: Offers clinicians a structured approach to managing rare presentations, emphasizing a multidisciplinary diagnostic and therapeutic strategy.
- This report is critical for advancing the understanding and management of rare autoimmune and inflammatory disorders.

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demonstrates the critical importance of a meticulous general examination in identifying subtle, systemic manifestations that may provide crucial diagnostic clues in complex presentations like this. Informed consent was obtained in written format from both the patient and the patient companion for publication.

The Case

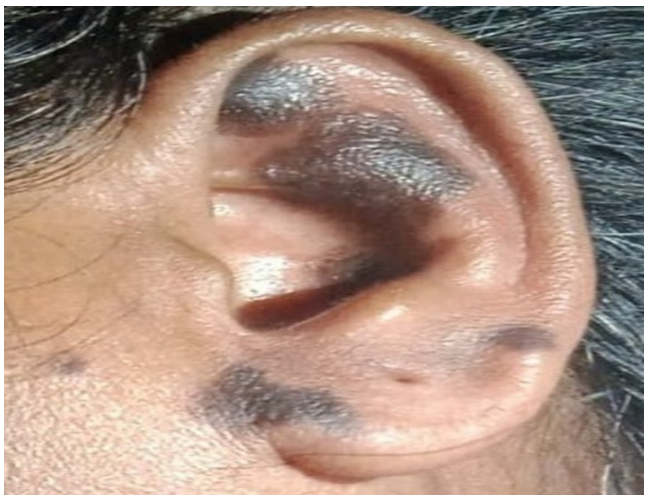
History

A 54-year-old female with no known comorbidities came to the neurology OP with chief complaints of sudden onset double vision for 2 days and pain in right eye on rightward gaze for 2 days with right sided unilateral headache for 1 week. The headache was continuous, right-sided, with a pin-pricking sensation and no identifiable triggers such as work exposure, talking, chewing, brushing of teeth, etc. It was not associated with nausea, vomiting, giddiness, or blurring of vision. The headache was predominantly present in the daytime thereby not affecting the sleep and the patient had no similar pattern of headache in the past. There were no complaints of vision loss, vomiting, nausea, fever, neck pain or stiffness, redness of eye, excessive tears, trauma and abnormal sweating of face.

Examination

On examination, the patient had pallor and macular lesions. These lesions were 5 in number and hyper pigmented in nature with irregular shape and margins having no associated pain. Alopecia areata was also noted. On cranial nerve examination, right 3rd cranial nerve palsy was observed with inability to adduct, elevate/depress right eye but intact 4th and 6th cranial nerve. The 5th cranial nerve was normal except for pain in the right V1 dermatome i.e. ophthalmic branch of the trigeminal nerve. Other neurological and systemic examinations yielded no significant findings.

Figure 1. Hyperpigmented Macular Lesions in the Earlobe and Concha.

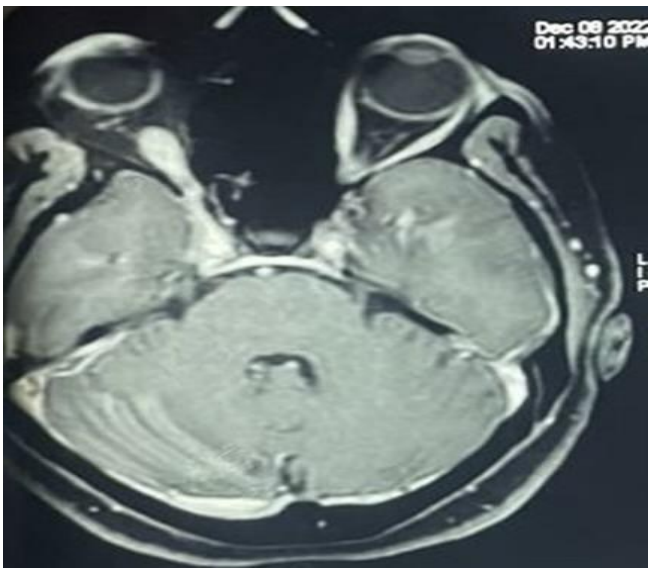


With this patient history and examination findings, the pathology can be localized to either the cavernous sinus, superior orbital fissure or orbital apex syndrome. As the colour vision and the visual field were normal, the optic nerve is found to be intact ruling out orbital apex syndrome. Some inflammatory etiologies that could be suspected are infectious (mucormycosis), granulomatous (Tuberculosis, sarcoidosis, Tolosa Hunt syndrome, IGG4 disease) and connective tissue disorders.

Investigations

For further management, the patient was admitted and necessary investigations were carried out. Complete blood count showed decreased hemoglobin and elevated erythrocyte sedimentation rate (ESR). High erythrocyte sedimentation rates (ESRs) and high titres of antinuclear antibodies (ANAs) are associated with progression to SLE in patients with DLE, and SLE patients with DLE3. Fasting and postprandial blood glucose levels were within the normal range. To identify if the pathology developed from the cavernous sinus or superior orbital fissure, MRI brain contrast was ordered which showed asymmetric thickening of the right cavernous sinus more than the left thereby confirming our localisation. The lesion also extended into the superior orbital fissure and orbit. Normal CSF analysis ruled out infectious causes(mucormycosis) and normal IgG4 and ACE levels ruled out granulomatous etiology. Indirect immunofluorescence showed a homogeneous pattern with 4+ positivity for antinuclear antibodies (ANA). The C3 and C4 levels were normal with an ENA panel showing 2+ titres for dsDNA. A biopsy of the skin lesion was taken which helped us confirm the diagnosis of Discoid Lupus Erythematosus (DLE) presenting with Tolosa Hunt Syndrome.

Figure 2. MRI Brain Contrast Showing Asymmetric Thickening and Enhancement of the Right Cavernous Sinus More than the Left.



Treatment

With this diagnosis in mind, the patient was treated with pulse steroid therapy for 3 days followed by oral steroids. As the patient was progressing to Systemic Lupus Erythematosus, Mycophenolate mofetil was added after consulting a

Table 1. Summary

Clinical Presentation	Right-sided headache, diplopia, right eye pain, hyperpigmented skin lesions, alopecia areata
Differential Diagnoses	- Infectious pathology (e.g., mucormycosis) - Granulomatous diseases (e.g., tuberculosis, sarcoidosis, IgG4-related disease) - Tolosa-Hunt Syndrome - Connective tissue disorders
Key Investigations	- CBC: Anemia, elevated ESR - Blood glucose: Normal - MRI Brain (Contrast): Right cavernous sinus thickening and enhancement - CSF: Normal - IgG4 and ACE levels: Normal - ANA: 4+ homogeneous pattern - dsDNA: Positive - Skin Biopsy: Confirmed Discoid Lupus Erythematosus
Final Diagnosis Reasoning	- Cavernous sinus localization supported by MRI - Normal CSF and granulomatous markers ruled out infection and sarcoidosis/IgG4 disease - Positive autoimmune markers and skin biopsy confirmed DLE - Cavernous sinus inflammation with autoimmune profile favored Tolosa-Hunt Syndrome secondary to autoimmune process
Management	- Pulse corticosteroid therapy - Oral steroids tapering - Mycophenolate mofetil added for immunosuppression
Outcome	Symptomatic improvement, no relapse during 5-month follow-up

rheumatologist, as a precautionary measure. Patient improved symptomatically with no events of relapse.

Discussion

Tolosa-Hunt syndrome is a nonspecific granulomatous inflammation marked by the infiltration of lymphocytes and plasma cells, primarily affecting the cavernous sinus and occasionally extending into the superior orbital fissure or orbital apex.1

This case presents a unique combination of Discoid Lupus Erythematosus (DLE) with THS, an association scarcely documented in the literature. Discoid lupus erythematosus (DLE) is the most common type of cutaneous lupus erythematosus. It is a chronic, scarring, photosensitive autoimmune skin condition characterized by red, scaly lesions.[4] The pigmented macular variant is a lesser-known morphological form that should not be missed by clinicians while diagnosing DLE.[5] The patient's hyperpigmented skin lesions and positive immunological markers (notably, dsDNA positivity and homogeneous immunofluorescence pattern, positive skin biopsy) aligned with DLE diagnosis, while cranial nerve findings and MRI supported THS due to cavernous sinus inflammation. Alopecia areata and Hyperpigmented macular lesions in the earlobe and concha are highly suggestive of DLE and calls for further investigation to confirm the diagnosis.

The coexistence of DLE with THS may suggest a shared autoimmune mechanism, as noted in other autoimmune disorders linked with THS. It has been proposed that inflammatory lesions in autoimmune conditions can target structures within the cavernous sinus, manifesting as ophthalmoplegia.[6] While the exact autoimmune cause of THS remains unclear, it has been observed in connection with various systemic and autoimmune inflammatory diseases such as SLE, sarcoidosis and immune mediated vasculitis such as Wegner's granulomatosis. In certain instances, THS can be the presenting symptom of these conditions.[7,8]

A thorough review of literature yielded only one other similar case that was reported by Amy W Yu. et al. This further emphasizes the rarity of such a presentation in a patient. Amy W Yu. et al discusses THS's etiology and diagnostic challenges, which focuses on the need for thorough differential diagnosis and comprehensive immunological testing, especially in atypical presentations like this one. Both patients presented with ipsilateral headache followed by painful ophthalmoplegia due to isolated third nerve palsy, with imaging confirming cavernous sinus and superior orbital fissure involvement, consistent with THS. In both cases, DLE was diagnosed via skin biopsy, with characteristic facial lesions. However, our patient had positive autoimmune markers (ANA, dsDNA, 4+ homogeneous immunofluorescence) and no comorbidities, while the case report had negative autoimmune labs but a background of multiple systemic illnesses. Both had normal CSF and responded well to steroids, confirming the diagnosis of THS, likely secondary to autoimmune inflammation.

In this case, early and aggressive immunosuppressive therapy with steroids and mycophenolate mofetil led to symptom resolution, demonstrating the importance of prompt treatment to prevent relapse. Use of other immunosuppressants such as azathioprine and methotrexate are restricted to corticosteroid resistant cases. Injectable cyclophosphamide has been sparsely used with only one documented case suggesting successful treatment.[9] An initial regimen of pulse steroid followed by oral therapy aids in effective and rapid symptom amelioration.[10,11] This aligns with standard THS management, where corticosteroids are the primary treatment, and steroid-sparing agents are considered for recurrent or refractory cases. The patient's prognosis is favorable, given her positive response to steroid therapy and

immunosuppressive treatment, with no observed relapses. Long-term follow-up for a period of 5 months was carried out in the patient as it is essential to monitor for potential progression to SLE or recurrence of THS symptoms. This is in concordance with current management guidelines which advise a minimum follow up period of 6 months to ensure that the cranial nerve palsies resolve and to adequately taper the steroid dosage. Periodic MRI and serology is done to monitor for improvement. Continued monitoring and maintenance of immunosuppressive therapy will be critical in managing her condition and preventing future exacerbations.

Current literature lacks sufficient evidence to suggest a strong link between THS and autoimmune diseases. However, reported cases suggest that THS may coexist with other autoimmune disorders, such as SLE and Hashimoto's thyroiditis.[12,13]

This report thus underscores the importance of considering THS in patients with lupus presenting with cranial neuropathies and supports immunosuppression as an effective therapeutic strategy.

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