UNRAVELLING STROKE IN ERDHEIM-CHESTER DISEASE WITH OPTIC NERVE GLIOMA: A RARE CLINICAL CHALLENGE

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Background and Aim

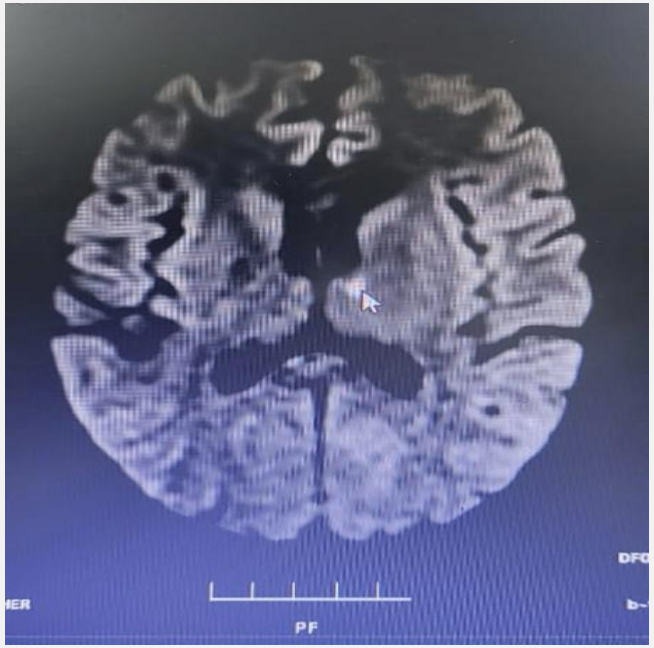
1. Erdheim-Chester Disease (ECD) is a rare non-Langerhans cell histiocytosis marked by xanthogranulomatous infiltration of organs by CD68+ and CD163+ histiocytes, often with surrounding fibrosis
2. CNS involvement is reported in up to 50% of cases, but ischemic stroke is extremely rare; cases with coexisting optic nerve glioma are exceptional.
3. The interplay of direct vascular infiltration and treatment-induced vasculopathy poses complex diagnostic and therapeutic challenges.
4. This case highlights a rare instance of ischemic stroke in ECD with optic nerve glioma, following chemotherapy.

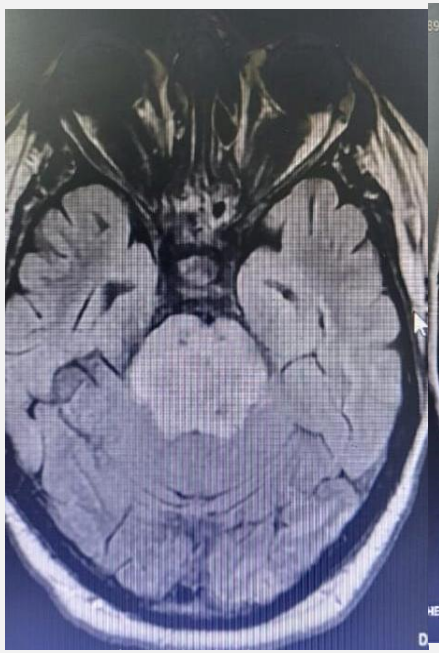
Methodology

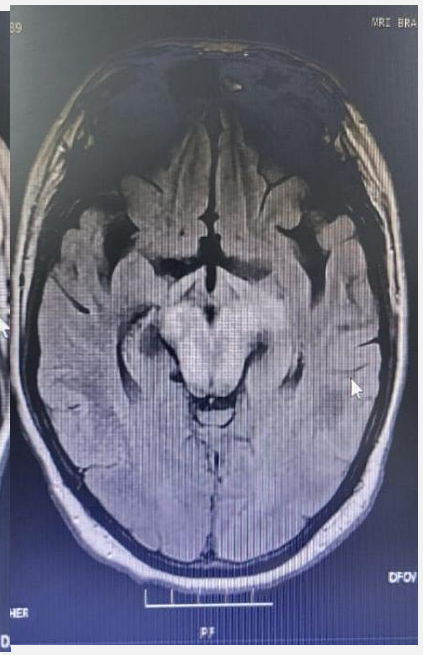
A middle-aged male with known ECD and optic nerve glioma, post-chemotherapy, presented with progressive right hemiparesis developing over one month.Clinical assessment revealed spastic quadriparesis.Systematic work-up included MRI brain, systemic imaging, and laboratory evaluation to rule out conventional stroke etiologies.

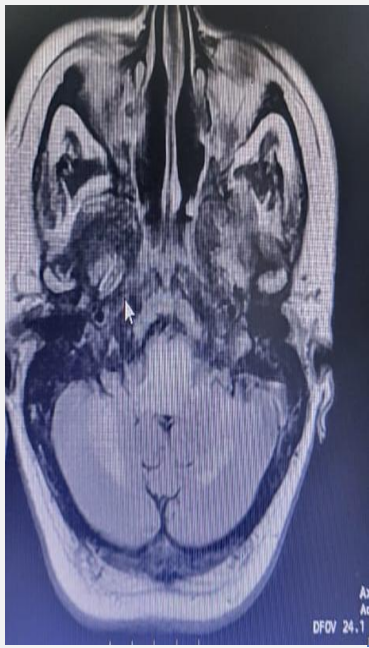
No conventional vascular risk factors identified.Etiology attributed to CNS vascular infiltration by histiocytes and possible chemotherapy-related endothelial injury.

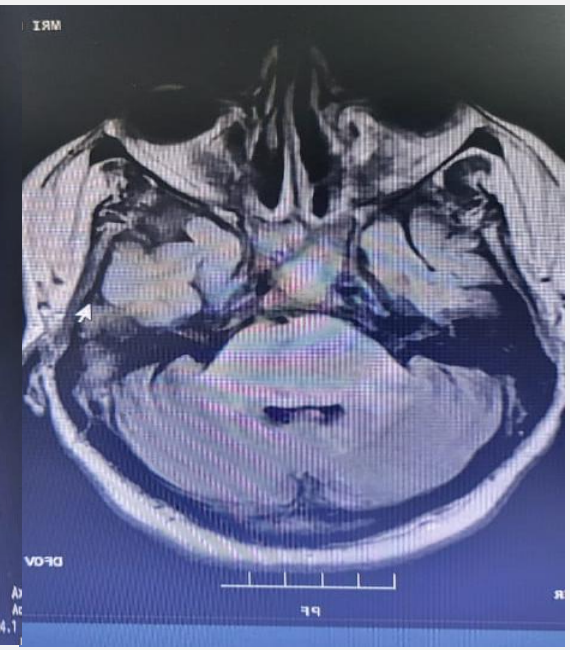
IMAGING & RESULTS











DWI/ADC and FLAIR sequences illustrating areas of acute ischemia (e.g., left temporo-occipital lobe, thalamus, hypothalamus), corresponding to the stroke event.

- T2/FLAIR MRI (Parenchymal and Chiasmal Changes)Axial or coronal sections showing T2/FLAIR hyperintensities in the optic chiasm, thalamocapsular region, optic radiations, and pons, demonstrating both infiltrative and ischemic changes in ECD.
- Orbital MRI (Optic Nerve/Chiasm Glioma)High-resolution orbital/coronal MRI slice showing thickened optic nerves, chiasm, and retrobulbar tissues—features suggestive of optic pathway glioma with surrounding infiltration.
- Gadolinium-Enhanced MRI (Pituitary Involvement)Sagittal section with post-contrast enhancement highlighting thickening of the pituitary stalk and adjacent hypothalamus (a classic site for ECD involvement)

Management:

Treated with intravenous corticosteroids, antiplatelet therapy, and low molecular weight heparin (enoxaparin).

Multimodal imaging and multidisciplinary management were crucial, with close monitoring for further neurological and ophthalmological sequelae.

Discussion:

Stroke in ECD, particularly with concomitant optic pathway glioma, is exceedingly rare but reported.

Mechanisms include perivascular infiltration, adventitial fibrosis, and endothelial injury from systemic therapy

Optic nerve and chiasmal involvement further complicates clinical presentation, imaging, and management

Prompt recognition and a combination of immunosuppressive and anticoagulant therapy are essential for stabilizing outcomes.

Conclusion:

Ischemic stroke is a rare but serious complication of ECD in the setting of optic nerve glioma and systemic treatment.

Early multimodal imaging and a multidisciplinary therapeutic approach are vital for improved outcomes in these complex cases.

Key References:

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- Haque A, Pérez CA, Reddy TA, Gupta RK. Erdheim–Chester Disease with Isolated CNS Involvement: A Systematic Review of the Literature. Neurol Int. 2022 Sep 5;14(3):716-726.