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Eti - Anatomico - Clinical Correlation of Dejerine - Roussy Syndrome with Choreoathetoid Movement and Ataxic - Hemiparesis in Localization of Stroke- A Case Report

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Abstract: ***Background: The Déjerine - Roussy syndrome is caused by lesions in the posterior Lateral nuclei of the thalamus, which usually have vascular aetiology. It has a prevalence of 17%–18%, after a stroke involving the inferior - lateral thalamus. This syndrome is characterized by, superficial hemianaesthesia, allodynia, severe and paroxysmal pain and choreoathetoid movements in the limbs on the paralyzed side. A posterior lateral thalamic lesion can present with ataxic hemiparesis contralateral to the side of the lesion. Methods: We have reported a case of a 65 - year - old diabetic and hypertensive male who presented with sudden onset of superficial hemianaesthesia, allodynia, severe and paroxysmal pain on the right side of the body with choreoathetoid movements in upper limbs along with slurred speech and unsteadiness while walking. Conclusion: We aimed to correlate aetiology and clinical features with the case of dejerine –roussay –syndrome with choreoathetoid and ataxic hemiparesis representing specific lesions of the central pathway. This case includes somatosensory landmarks along with tracts and pathways that are linked to the topography of the cerebrovascular lesions and therefore will help in suitable clinical evaluation and management of stroke.***

Keywords: Dejerine - Roussay Syndrome, Ataxia, Stroke, Thalamus.

1. Introduction

DejerineRoussy syndrome, a/k thalamic pain syndrome occurs after infarction of the ventro posterolateral thalamus. Joseph Jules Dejerine and Gustave Roussy first described it in 1906 in their paper titled *Le syndrome thalamique*. It is associated with inadequate blood supply from the posterior cerebral artery in the posterior lateral nuclei of the thalamus, which usually has vascular aetiology. [1, 2]It affects approximately 8% of patients after a stroke but is more common after strokes that involve the lateral medulla and inferior - lateral thalamus, with a prevalence of 25% and 17%–18%, respectively. [3, 4]This syndrome is characterized by transient mild hemiparesis, superficial hemianaesthesia, allodynia, mild hemiataxia, stereognosis, severe and paroxysmal pain on the hemiparetic side, and choreoathetoid movements in the limbs on the paralyzed side. The sensory disorder involves both superficial (touch, pain and temperature) and deep (position, vibration) modalities. The pain is continuous, with paroxysmal exacerbations, and it is not suppressed by conventional analgesic treatment.

Fisher and Cole 1965 coined the term ataxic hemiparesis after pathological studies of three cases which revealed infarct cavities in the basis pontis on the side contralateral to the pyramidal and cerebellar signs. Ataxic hemiparesis has also been described as a result of lesions in the midbrain, [5 6] posterior limb of the internal capsule [7 - 11] and corona radiate. [12]Cerebellar ataxia was most likely due to interruption of the dentate - rubro - thalamo - cortical fibres at the level of the injured ventrolateral nucleus. Hemiparesis was probably caused by local oedema compressing the corticospinal tract in the adjacent posterior limb of the internal capsule.

Isolated Dejerine –Roussy syndrome and Ataxic hemiparesis have been reported but Dejerine Roussy syndrome with choreoathetoid and ataxic hemiparesis is rarely reported. We aimed to correlate aetiology and clinical features with the case of Dejerine –Roussy –syndrome with choreoathetoid movement and ataxic hemiparesis representing specific lesions of the central pathway. These causes include somatosensory landmarks along with tracts and pathways that are linked to the topography of the cerebrovascular lesions and therefore will help in suitable clinical evaluation and management of stroke.

2. Case Report

A 65 - year - old right - handed man, known hypertensive and diabetic for 4 years, presented with sudden onset of superficial hemi - anaesthesia, allodynia, severe and paroxysmal pain on the right side of the body with choreoathetoid movements in upper limbs along with slurred speech and unsteadiness while walking. Soon after he noticed weakness and numbness in his right arm and leg such that while walking he would stagger and fall to the right. There was no headache, altered sensorium, vertigo or diplopia. On general examination revealed a BP of 170/100 and a regularpulse of 82 per minute. No bruit was heard in the neck. He was alert, oriented, and had mild dysarthria. Cranial nerves and fundi were normal. Power in the right upper and lower limb was 3+/5 and the left upper and lower limbs were 5/5. Right upper limb involuntary movement was noted. The Deep tendon reflexes were brisk. The plantar response was extensor on the right and flexor on the left. There was marked ataxia on the heel shin and finger - nose tests on the right but normal on the left side. Rapid alternate movements were impaired on the right and normal on the left. Touch, pain, and temperature sensations were absent on

the right half including the face. Joint position and sense of vibration were absent on the right half of the body. MRI brain done revealed a parenchymal bleed in the left thalamus with perilesional oedema and mass effect (Figure 1). {{Figure 1}} He was managed with anti - osmotic antihypertensive, neuropathic pain modulator steroids, physiotherapy, and gait training. A repeat NCCT head done on the fourth day of hospitalisation showed decreased focal parenchymal bleed in the left thalamus with perilesional oedema and mass effect (Figure 2). {{Figure 2}} On day 10 he was discharged with persistent numbness, but pyramidal signs disappeared and the ataxia resolved completely.

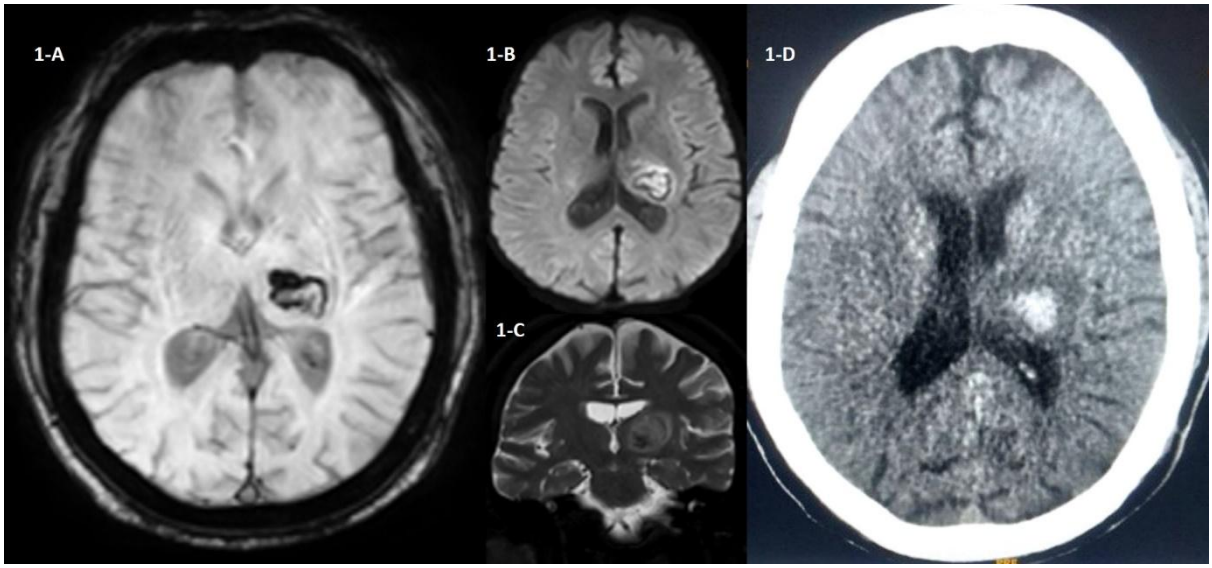

Figure 1: MRI Brain of the patient showing left Ventro - Postero - Lateral (VPL) thalamic bleed (Figure 1 A, B and C). Figure 1D depicts a repeat NCCT head of the patient done on the fourth day of hospitalization demonstrating decreased focal parenchymal bleed in the left thalamus with perilesional oedema and mass effect

3. Discussion

The symptoms and signs resulting from thalamic disease [13] may be categorized into disturbances of sensation, motility, and vegetative and mental functions. Speech abnormalities have been described in thalamic lesions of the dominant hemisphere. [14 - 15] Further topographic syndromes — such as posterolateral, anterolateral, and medial thalamic syndromes arising from a more discrete lesion have also been mentioned (Figure 2). [13] {{Figure 2}}

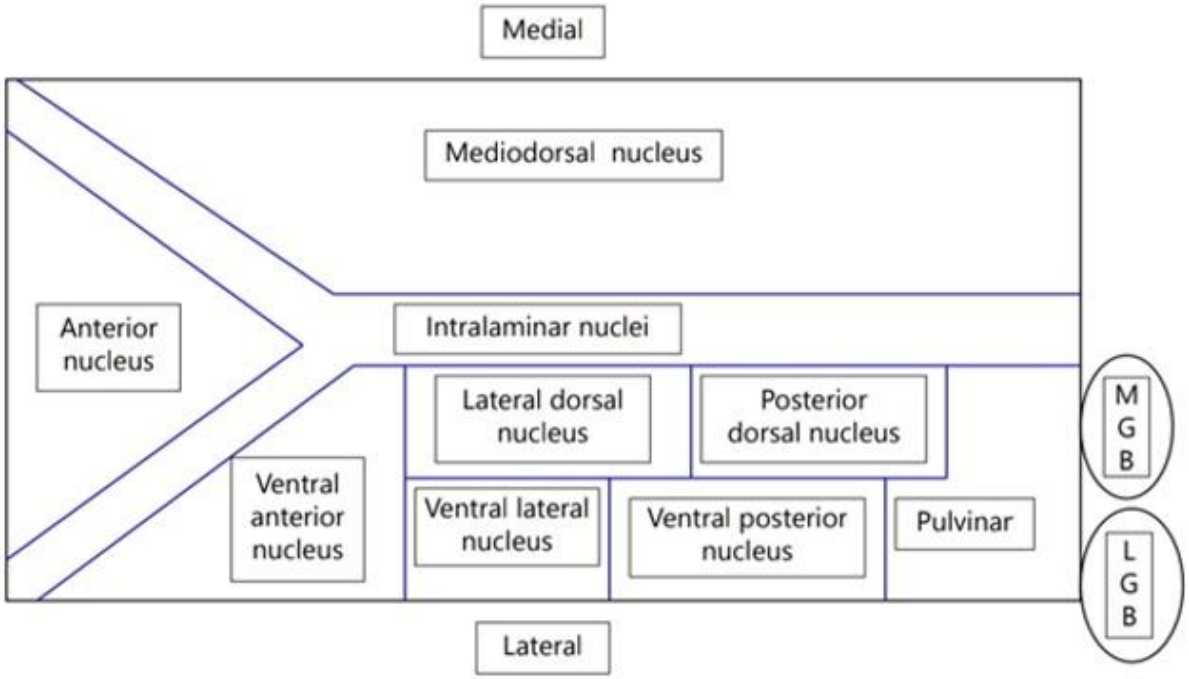

Figure 2: Line diagram showing different nuclei of the thalamus. LGB, lateral geniculate body; MGB, medial geniculate body. [16 - 17]

There are 4 major thalamic vascular territories, each with a predilection for supplying particular groups of nuclei (Figure 3). In the nomenclature used here, these are the (1) tuberothalamic, (2) inferolateral, (3) paramedian, and (4) posterior choroidal vessels. [18] {{Figure 3}}

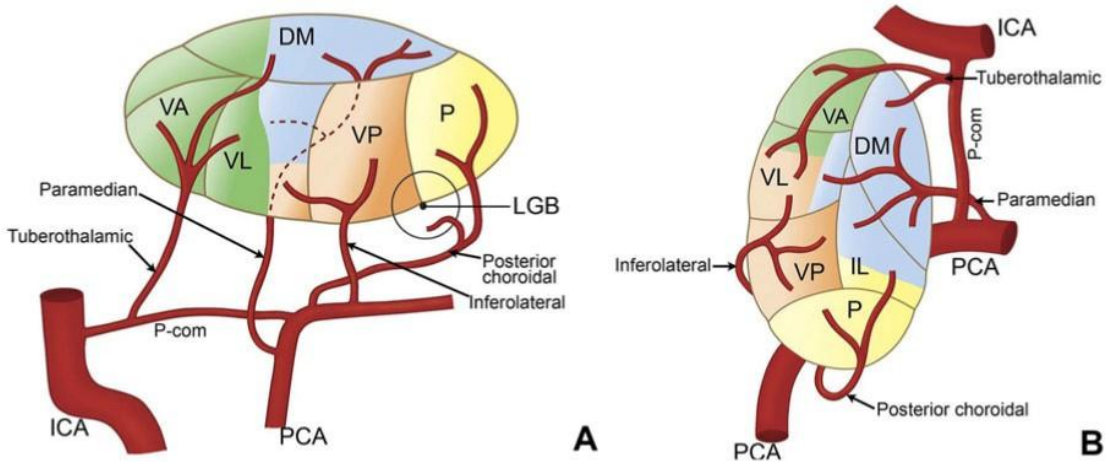

Figure 3: Schematic diagram of lateral view (A) and dorsal view (B) of four major thalamic arteries and the nuclei they irrigate. VA = ventral anterior; VL = ventral lateral; DM = dorsomedial; IL = intralaminar nuclear complex; VP = ventral posterior; P = pulvinar; LGB = lateral geniculate body; PCA = posterior cerebral artery; ICA = internal carotid artery; P - com = posterior communicating artery [18 - 19]

Our patient had superficial hemianaesthesia, allodynia, severe and paroxysmal pain on the right side of the body with choreoathetoid movements in the upper limbs because somatosensory modalities are processed in the ventral posterior nucleus of the thalamus contralateral to the side of the body. so, Posterolateral thalamic lesion, represents the classical thalamic syndrome of Dejerine and Roussy (Figure 4). [20] {{Figure 4}}

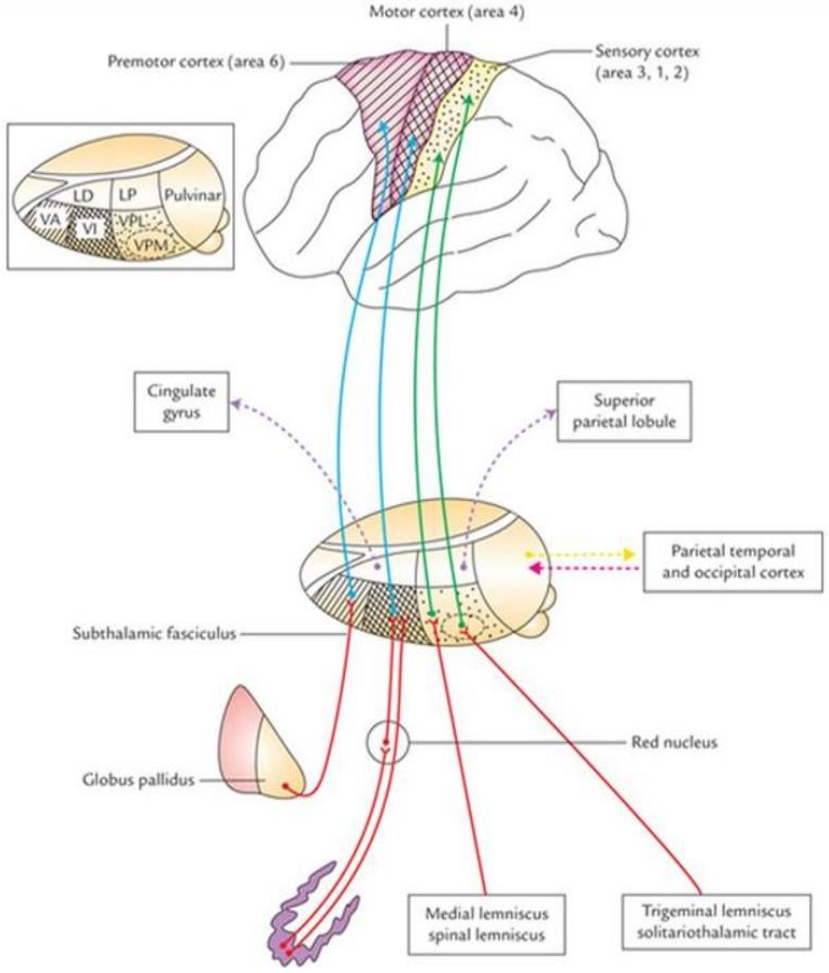

Figure 4: Scheme to show connections of ventral and lateral groups of thalamic nuclei. Dorsolateral view of the thalamus and its major subdivisions. (LD = lateral dorsal nucleus, LP = lateral posterior nucleus, VA = ventral anterior nucleus, VI = ventral intermediate N, VPL = ventral posterolateral n. VPM = ventral posteromedial n.) [21]

Choreoathetoid movements on the involved side in our case can be explained by the fact that The ventral tier nuclei (ventral anterior nucleus and ventral lateral nucleus) have important connections with basal ganglia and cerebellum. [22] So, Damage to these nuclei and their connections can give rise to abnormal involuntary movements. [23] The hemiparesis described in Dejerine and Roussy syndrome was because of local oedema compressing the corticospinal tract in the adjacent posterior limb of the internal capsule. Cerebellar ataxia was most likely due to interruption of the dentate - rubro - thalamo - cortical fibres at the level of the injured ventrolateral nucleus. Thus we have correlated aetiology and clinical features with its anatomical correlation which will help in suitable clinical evaluation and management of stroke.

4. Conclusion

1) It is evident from the above - mentioned scenario that Dejerine –Roussay –Syndrome may present with ataxic hemiparesis, superficial hemianaesthesia, allodynia, severe paroxysmal pain on the hemiparetic side, and choreoathetoid movements in the limbs on the paralytic side.
2) In view of the anatomical complexities and varied clinical presentations, clinicians must be aware of this clinical entity.

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