Trigeminal trophic syndrome following anterior inferior cerebellar artery infarction

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DESCRIPTION

A 70-year-old woman with hypertension presented about 3 months ago with complaints of dysarthria, left lower motor neuron facial palsy with preserved Bell's phenomenon, sensory loss over the left side of her face (V1, V2, V3) along with the left half of the tongue, absent left corneal and conjunctival reflexes, and gait ataxia. MRI of the brain showed an acute left anterior inferior cerebellar artery infarction (figure 1). She was treated with antiplatelets, statins and antihypertensives.

At present, she complained of 1-month history of multiple non-healing painless ulcers over the left side of her face below the nostril, and on the left side of the nose and forehead. Her left eye had undergone keratosis (figure 2). She had consulted local doctors, who prescribed her topical antibiotics and steroids, and eye-drops, but there was no improvement. There were no vesicles or peripheral nerve thickening. Her vitals were stable. Neurologically, she had left lower motor neuron facial palsy, difficulty in closing her left eye, sensory loss over the trigeminal area, diminished corneal and conjunctival reflexes, and left sensorineural hearing loss. She also had left-sided incoordination. Other systemic examinations were normal. Her blood investigations such as complete blood count, renal and liver functions, electrolytes, thyroid-stimulating hormone and haemoglobin A1c were normal. Her lipid profile showed mild dyslipidaemia. Viral markers (HIV, HBsAg, anti-HCV), VDRL and ANA profile were negative. Skin smear for bacteria and acid-fast bacilli were also negative.

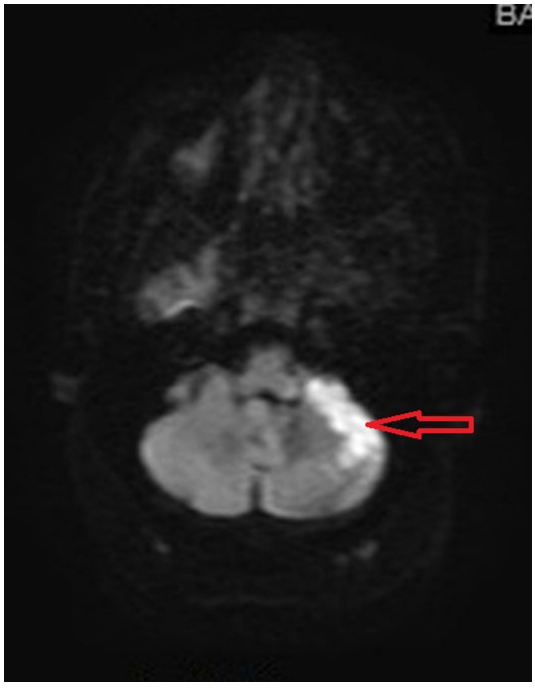

Figure 1 MRI of the brain showing an acute infarct in the left anterior inferior cerebellar territory.

On further probing, she revealed repeated scratching of the left side of her face due to an itching sensation following the stroke event. From the history and clinical findings, a diagnosis of trigeminal trophic syndrome was considered. She was treated with topical antibiotics (clindamycin) and saline dressings, and antibiotic eye-drops (ciprofloxacin). Antiplatelets (aspirin 75 mg and clopidogrel 75 mg once daily), antihypertensives (telmisartan 40 mg once daily) and statins (rosuvastatin 5 mg once daily) were continued. She was counselled regarding scratching of the left side of her face. She was also advised left eye keratoplasty, but was not willing. Botox injection was given as a therapeutic treatment for the partial ptosis of the left eyelid. She was asked to return after 1 week, but was lost to follow-up.

Cerebrovascular accidents involving the anterior inferior cerebellar artery are characterised by ipsilateral cerebellar ataxia, Horner's syndrome, involvement of V, VII and VIII cranial nerves, and contralateral sensory disturbance of temperature sensation. An occlusion of the internal auditory artery can result in peripheral vestibular syndrome.[1–4] Trigeminal trophic syndrome is a rare condition that occurs following damage to the trigeminal nerve or its central sensory connection. It was first described by Loveman[5] and McKenzie[6] in 1933, and is characterised by painless persistent ulceration of the ala nasi. These lesions may also occur in the corners of the eyes, scalp or the inside of the mouth. The tip of the nose is spared as it derives sensation from the medial branch of the anterior ethmoidal nerve. It is more common among women. It may be idiopathic or may follow trigeminal nerve ablation, Wallenberg syndrome, amyloid deposition in the central nervous system and trigeminal nerve, trauma, craniotomy, herpes zoster, herpes simplex, leprosy, syphilis, birth trauma, or as postencephalitic sequelae.[7 8] The manifestations of trigeminal trophic syndrome begin weeks to years after the injury to the trigeminal nerve, with an average of 2 years. These patients usually complain of picking, rubbing or scratching sensations over the affected areas, and the ulceration is unknowingly self-induced due to repeated touching of the face and

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Figure 2 Left-sided facial ulceration and left eye keratosis.

self-manipulation because of these sensations.[9] The diagnosis is made on the basis of the presence of facial paraesthesia, long history of repeated physical manipulation of the affected areas, facial ulceration with sparing of the nose tip and history suggestive of trigeminal nerve damage.[10] The treatment of trigeminal trophic syndrome is often ineffective. Patients are counselled to avoid self-manipulation of the wound. Medications such as diazepam, amitriptyline, carbamazepine and chlorpromazine are used to reduce paraesthesia.[11–13] Occlusive dressings may be applied over the affected area to reduce trauma. Transcutaneous electrical stimulation, stellate ganglionectomy, ipsilateral cervical sympathectomy, radiotherapy and iontophoresis with nerve blockade have been tried in treatment-resistant cases.[5 6 14 15] Surgical reconstruction with skin grafting followed by pulsed radiofrequency has been tried to improve paraesthesias.[9 16 17] The differential diagnosis includes vasculitis (Wegener's granulomatosis, giant-cell arteritis, temporal arteritis), infections (varicella zoster, herpes simplex, leprosy, syphilis, leishmaniasis, cutaneous tuberculosis, blastomycosis, paracoccidioidomycosis), carcinoma (basal cell and squamous cell), destructive lethal midline granuloma, pyoderma gangrenosum and factitial dermatitis.[9]

Learning points

- Trigeminal trophic syndrome is a rare condition that occurs due to damage to the trigeminal nerve.
- It is characterised by persistent non-healing unilateral facial ulceration, typically involving the ala nasi with sparing of the tip of the nose.
- Anterior inferior cerebellar artery infarction is an uncommon cause of trigeminal trophic syndrome.

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