Letter to the Editor

An unusual case of Ramsay Hunt syndrome: The pathology yet unknown

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Sir,

The clinical presentation of Ramsay Hunt syndrome includes a vesicular rash in the ear (herpes zoster oticus) or in the oral mucosa, accompanied by acute peripheral facial nerve paralysis.

We present an atypical Ramsay Hunt syndrome case of a 45-year-old female without neuron facial palsy.

The 45-year-old female presented at the emergency department of dermatology complaining of severe pain in the right hemiface, described as electric discharges, with dermatologic lesions that appeared one week before.

She had a history of 20 years of smoking.

An examination indicated a vesicular rash and swollen papules affecting the right tragus, cheek, mental region, and ipsilateral hard palate (Figs. 1a–1c).

Initial treatment included 1000 mg valacyclovir three times a day for seven days.

Two days later, the patient expressed a progressing deterioration: dysphagia to solid foods, hearing loss, and headache without evident dropping eyes or erasing nasolabial folds.

A clinical diagnosis of Ramsay Hunt syndrome (RHS) was made.

The patient was started on treatment with prednisone 60 mg/day for seven days with tramadol 50 mg two times a day.

One week later, the patient showed significant improvement in the hearing loss and headache, and returned to eating foods as normal.

A four-month follow-up did not reveal residual symptoms.

Although Ramsay Hunt syndrome is traditionally defined as zoster oticus and lower motor neuron facial palsy, Hunt enumerated other common symptoms, such as tinnitus, hearing loss, nausea, vomiting, vertigo, and nystagmus, and explained these nervous symptoms by the proximity of the geniculate ganglion to the vestibulocochlear nerve within the bony facial canal.

Based on clinical presentations that indicated the involvement of more than one ganglion, Hunt surmised that the gasserian, geniculate, petrous, accessory, jugular, plexiform, and second and third cervical dorsal root ganglia comprise a chain in which inflammation of a single ganglion can extend to nearby ganglia. This hypothesis explains cases of unilateral facial palsy accompanied by contiguous cranial neuropathies associated with vesicles usually in the mouth, tongue, hard palate, and ears [1].

The largest retrospective Ramsay Hunt syndrome treatment study showed a statistically significant improvement in patients treated with prednisone and acyclovir within 3 days of onset [2].

Data from collective case reports and retrospective reviews suggest that both prednisone and acyclovir, if given early, improve the overall prognosis [3].

Patients who were diagnosed early and received appropriate treatment showed an improvement in the

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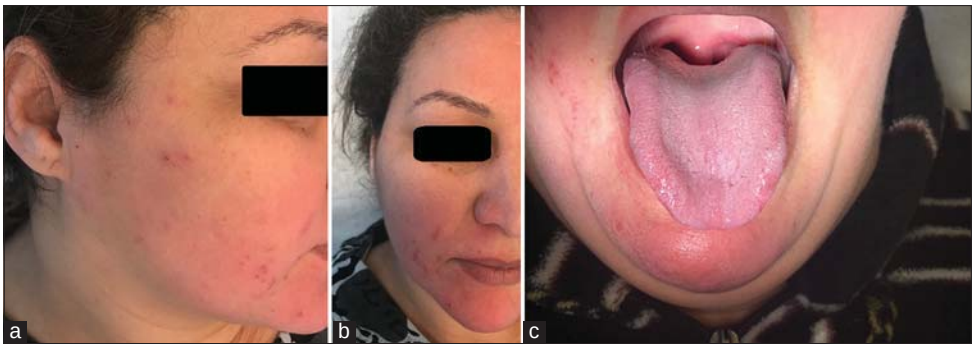

Figure 1: (a and b) Vesicular rash and swollen papules affecting the tragus, cheek, and mental region on the right side of the face. (c) Erythematous macules of the right hard palate.

damaged nerves and achieved a complete recovery of facial nerve function [3,4].

A diagnosis of Ramsay Hunt syndrome can be difficult to reach because the specific symptoms—otalgia, facial paralysis, and the distinctive rash—do not always develop simultaneously.

Our case is significant in that it establishes that early diagnosis leads to early treatment and prevents serious complications, such as permanent hearing loss and facial weakness, eye damage, and postherpetic neuralgia.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients have given consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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