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What is your diagnosis?

Unilateral oral, pharyngeal and laryngeal vesicles

L. Alciato[a,*], P. Bonfils[a], F. Rubin[b]

[a] Hôpital Européen Georges-Pompidou, 20, rue Leblanc, 75015 Paris, France
[b] CS 71027, clinique Saint-Vincent, 8, rue de Paris, 97404 Saint-Denis cedex, Reunion

1. Description

A 28-year-old woman in complete remission from sacral myeloid sarcoma, treated by allogeneic stem cell transplantation and immunosuppressives, attended the emergency room with intense acute odynophagia associated with intractable hiccoughs and repeated vomiting. Physical examination revealed labile blood pressure and tachycardia. ENT examination revealed a non-contiguous oedematous vesicular rash covered by a whitish coating, affecting all of the right side of the oral cavity, oropharynx and larynx, while no lesions were observed on the left side. Soft palate and laryngeal mobility was preserved. There were no signs of other cranial nerve involvement. Examination of the neck did not reveal any rash, lymphadenopathy or mass.

2. Question

The oral and pharyngolaryngeal examinations are presented below (Fig. 1a,b).

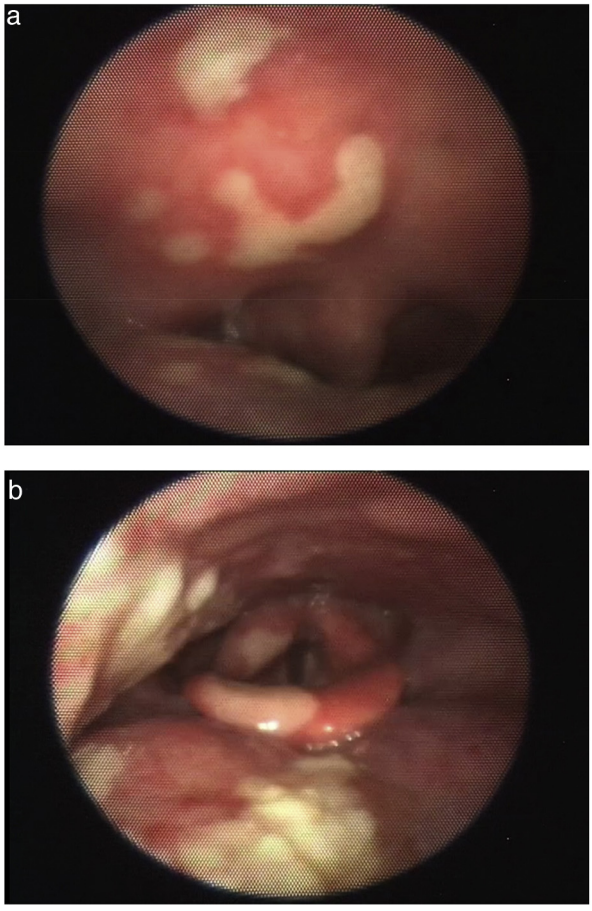

Fig. 1. a: fibroscopic view of the soft palate, uvula and tongue; b: fibroscopic view of the pharynx, larynx and base of the tongue.

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* Corresponding author.
E-mail address: lauranne.alciato@gmail.com (L. Alciato).

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3. Answer

The final diagnosis was that of isolated right glossopharyngeal nerve (IX) and vagus nerve (X) herpes zoster, confirmed by positive Varicella-Zoster Virus (VZV) PCR. MRI of cranial nerves was normal. The mucosal lesions resolved within 5 days and pain resolved after 7 days of treatment with aciclovir and anti-neuropathic analgesics, but hiccoughs persisted for about one month.

The glossopharyngeal nerve transmits sensory information from the palate and pharynx, and the base of the tongue, and also has a visceromotor function in the parotid gland. The vagus nerve has a sensorimotor function in the pharynx and larynx and a parasympathetic function in thoraco-abdominal organs, and is responsible for signs of dysautonomia [1].

Varicella-Zoster Virus (VZV) reactivation in adults is responsible for a painful vesicular rash limited to the territory of the sensory nerve root involved. Epidemiologically, the cranial nerves most frequently affected are the ophthalmic division of the trigeminal nerve (V1) and the facial nerve (VII) (Ramsay Hunt zone) [2]. Involvement of other cranial nerves is rare according to a recent review of the literature by Nisa et al., who reported only 57 cases [3]. Glossopharyngeal and vagus nerve herpes zoster is also usually (83% of cases) associated with involvement of other cranial nerves (especially VII and VIII) [3].

The diagnosis is rarely supported by imaging, as gadolinium-enhanced MRI thin-section T1-weighted sequences demonstrates enhancement over the course of the nerve involved in only one-third of cases, but CT is always normal [3]. This gadolinium enhancement appears to be particularly rare (only 4 cases have been reported in the literature) in the glossopharyngeal and vagus nerves [4]. Imaging is therefore not sufficiently sensitive to confirm the diagnosis of VZV neuritis, but remains necessary to exclude a tumour, especially when multiple cranial nerves are involved [5].

No consensus has been reached concerning optimal treatment. Antiviral agents are indicated when they can be initiated within 72 hours following onset of the rash in order to reduce pain and accelerate healing of skin lesions [3,6]. Conventional analgesics or tricyclic antidepressants can be effective to control herpes zoster pain and postherpetic neuralgia, while the use of corticosteroids remains controversial. The prognosis is related to long-term sequelae of IX or X lesions on voice and swallowing (dysphonia and dysphagia), with complete recovery observed in only 26% of patients [3].

Disclosure of interest

The authors declare that they have no competing interest.

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