



Cephalalgia heterotopica: a case series of extratrigeminal cluster headache

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Introduction

Cluster headache is a very disabling neurological disorder that usually presents with unilateral severe headache associated with ipsilateral cranial autonomic symptoms. Even among the typical cases, there is a considerable diagnostic delay and most patients will have seen three general practitioners before being referred to neurology services, some having been to colleagues in dentistry or otorhinolaryngology. The extratrigeminal presentation of this condition poses a great obstacle to a proper diagnostic workup.

Objective

This consecutive case series study aimed to report clinical features, treatment, and outcome of 3 patients with extratrigeminal cluster headache.

Materials and methods

Case series of our Headache Clinic. Data disclosure was authorized by the patients through an informed consent form.

Results

Two males and one female were evaluated. The ages were 53, 64, and 71 years, respectively. The mean age of symptoms onset was 50, 60, and 66 years, respectively. Two patients described excruciating shoulder pain and one presents severe pain in the malar area. All patients reported prominent autonomic ipsilateral symptoms such as lacrimation, conjunctival injection, nasal symptoms, and restlessness accompanying pain attacks. Preventive treatment with verapamil and occipital nerve block was useful.

Conclusion

Extratrigeminal cluster presentation is a diagnostic challenge. Proper recognition of these cases prevents unnecessary examination and treatment trials and directly benefits the patients since effective treatment is already available. A proper case definition of this rare clinical presentation may provide new insight into our understanding of the cluster headache mechanisms.

Keywords: Cluster headache, Extratrigeminal headache, Shoulder pain.