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Case reports

Unilateral laryngeal and hypoglossal paralysis (Tapia's syndrome) in a patient with an inflammatory pseudotumor of the neck

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1. Introduction

Tapia's syndrome (TS) is a rare condition thought to be caused by injury to the extracranial course of both recurrent laryngeal branch of the vagal nerve and hypoglossal nerve. First described in 1904, it occurs with unilateral paralysis of the vocal cord and tongue, with normal function of the soft palate. Commonly reported causes are direct trauma, neurofibromatosis of X and XII nerves, carotid artery dissection involving the ascending pharyngeal artery, and displacement of endotracheal tube during general anesthesia [1].

2. Case report

Here we report a patient with a non-traumatic Tapia's syndrome of the left side caused by a nasopharyngeal inflammatory pseudotumor.

A 42-year-old housekeeping woman was referred to our Neurology Ward with several months history of hoarseness of voice, dysphonia and a slight discomfort during chewing and swallowing. She had been in a good health, with no significant past medical history. A routine medical check-up had been performed one year and half earlier which was reported as negative.

General examination did not disclose abnormalities. The neurological examination revealed a marked left hemiatrophy of the tongue, which was deviated to the right side (Fig. 1A), a voice hoarseness and dysphonia. Of note, the patient was unaware of the tongue hemiatrophy. She was not significantly dysphagic, as the 100 ml water swallow test was within normal range. All other cranial nerves appeared undamaged.

An extensive biochemical and immunological work-up were negative (including blood cell counts and a search for onconeural and anti-ganglioside antibodies). Cytoplasmic circulating autoantibodies against neutrophils (c-ANCA) showed very low titer. Cerebrospinal fluid (CSF) analysis showed no abnormalities and absent oligoclonal bands. A serum and CSF search for antibodies to HIV, *Treponema pallidum* and *Mycobacterium tuberculosis* was negative.

Post-contrast chest and abdomen CT scans were negative.

A laryngeal optic fiber endoscopy documented the paralysis of the left vocal cord, which was lying in a medial position. Concentric needle EMG demonstrated an acute denervation of the left genioglossus muscle.

The patient underwent a MRI study that showed a fibroadipose degeneration of the left hemitongue (Fig. 1B and C) and a 1.9 cm ovoid mass in the skull base, close to the hypoglossal canal and the left carotid space. The lesion was hypointense in both T1- and T2-weighted sequences and showed a slight enhancement after i.v. gadolinium (Fig. 1D–F).

The radiological and clinical characteristics of the lesion were supporting the hypothesis of an inflammatory pseudotumor (IPT) causing a Tapia's syndrome [2,3]. However, as a malignant nasopharyngeal carcinoma could not be excluded, a diagnostic biopsy of the lesion was offered which the patient declined. A glucocorticoid therapy was therefore started, and the patient underwent

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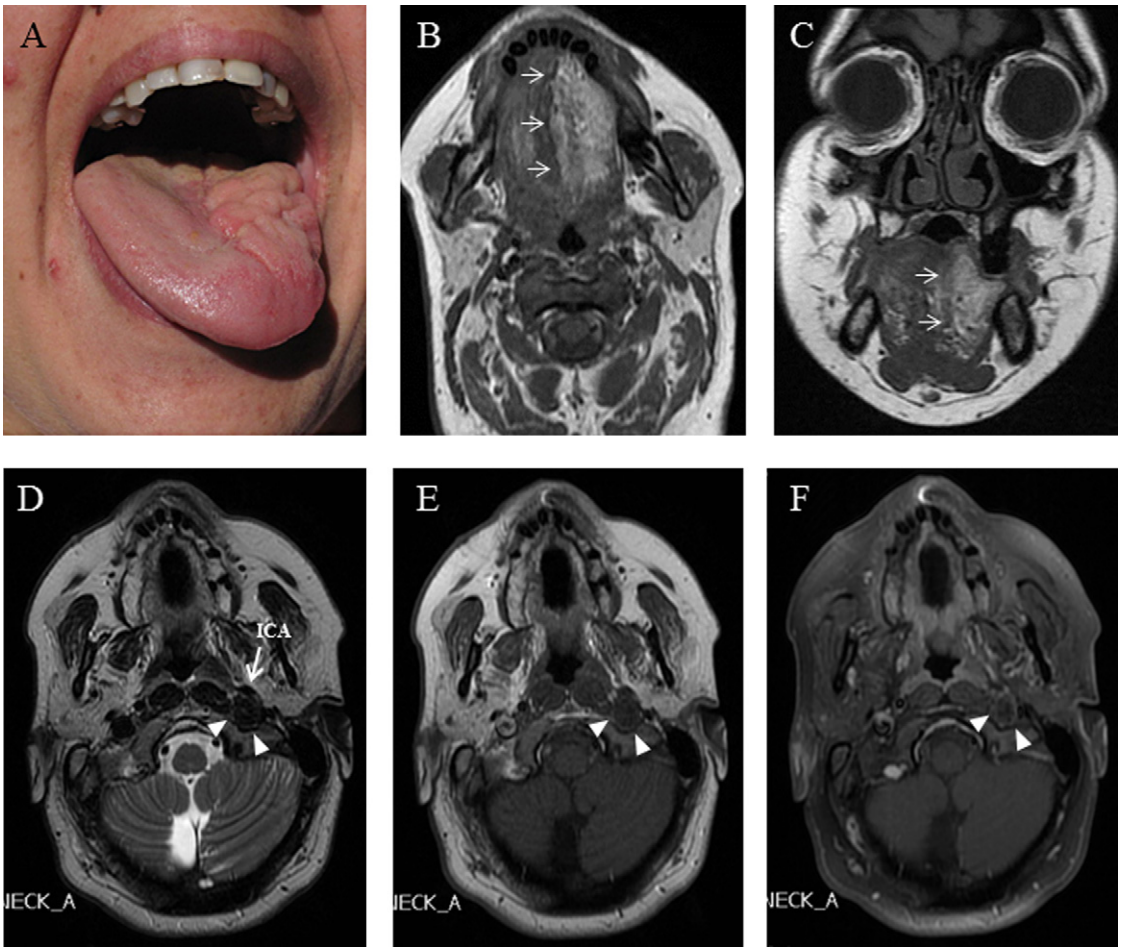

Fig. 1. Tongue hemiatrophy and MR images of the patient with Tapia's syndrome caused by an inflammatory pseudotumor of the skull base. The left hemiatrophy and deviation of the tongue was consistent with a lesion in cranial nerve XII (A). T1-weighted axial (B) and coronal (C) MR images showing hyperintense fat infiltration in the left hemitongue (arrows). The 1.9 cm-wide rounded lesion was hypointense in both T1- (D) and T2- (E) weighted axial MR images (arrowheads), and it was encasing the left internal carotid artery (ICA, arrow in D). After gadolinium injection, the lesion showed a slight ring-like post-contrast enhancement (F).

strict monitoring with serial MRI and clinical observation. At 6-months follow-up the patient was clinically unchanged and the lesion appeared stable on MRI. The glucocorticoid therapy was gradually discontinued. At one-year follow-up, a new MRI further showed that the ovoid lesion was actually unmodified (data not shown). Based on the lack of pathological and clinical progression of the lesion and given the relative specific MRI characteristics, the small mass in the skull base of our patient was identified as inflammatory pseudotumor.

3. Discussion

Tapia's syndrome, which involves the recurrent laryngeal and hypoglossal nerves, has been often described as a complication of airway management under general anesthesia. The syndrome has also occasionally been associated to neurolemmoma or neurofibroma [1].

Our patient had a very rare non-traumatic Tapia's syndrome, most likely caused by an inflammatory pseudotumor (IPT) of the left carotid space. The term *pseudotumor* is an ambiguous designation and it is used to define a broad category of proliferating masses believed to be reactive rather than truly neoplastic.

Although the patient's refusal to undergo a biopsy prevented us to proceed to an pathologic diagnosis, several lines of evidence support the clinical diagnosis of IPT in our case. First, the serial MRI studies showed that the mass was hypointense in both T1- and T2-weighted images when compared to the gray matter of the brainstem (Fig. 1D and E). This has been suggested to be highly specific for IPT [2,3]. Second, the fact that lesion was showed a weak gadolinium enhancement (Fig. 1F), made it unlikely the hypothesis of a infiltrating nasopharyngeal carcinoma, a lymphoma, a schwannoma or a meningioma. It rather suggests that the IPT lesion in our patient had a spontaneous fibrotic involution, an hypothesis indirectly supported by the stability of the lesion at one-year follow-up and by the lack of a significant therapeutic response to a prolonged corticosteroid therapy [2]. A negative titer for c-ANCA allowed us to exclude a Wegener granulomatosis. Other pathological conditions potentially responsible for chronic inflammatory lesions were excluded.

The pathogenesis of IPT is unknown, but there is evidence that it may be related to an autoimmune reaction to virus infection [4]. Our patient was however in a relative good health until presentation, and in her medical history we could not find traces of a former infection of autoimmune reaction. Thus, the etiology of IPT in our case remain unexplained and no inciting factor could be determined, as reported by others [5]. The IPT in our case was most likely of a relative long duration, as we could document a fibroadipose involution of the left hemitongue already at presentation (Fig. 1A–C).

We report a rare non-traumatic Tapia's syndrome associated to a fibrosing IPT of the skull base. The distinctive MRI feature of the lesion was a marked hypointensity in T1- and T2 weighted images associated to feeble gadolinium enhancement. Our case shows that,

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in the absence of an histological study, an appropriate clinical diagnosis of IPT can be obtained after a careful follow-up and after considering a wide spectrum of differential diagnoses, requiring a thorough laboratory and imaging work-up.

Conflicts of interest

The authors declare that they have no conflict of interest.

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