Intention Tremor and Olivary Enlargement: Clinico-Radiological Study

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This study describes two patients who presented with intention tremor (IT) concomitant with olivary enlargement (OE), and discusses the relationship between these phenomena. Both patients had a vascular lesion in the brain stem, which subsequently caused IT and OE ipsilateral or contralateral to the site of the lesion, depending on the structures effected by the lesion. The results of our study suggest that damage involving the superior cerebellar tract, and central tegmental tract, or both, may result in IT and OE. However, IT and OE appear to have little physiological association, unlike the relation between OE and palatal myoclonus.

Key words: inferior olivary nucleus, superior cerebellar tract, central tegmental tract, vascular brain stem lesion

Introduction

Olivary enlargement (OE) is a swelling of the inferior olivary nucleus, particularly of the neurons and astrocytes. OE is a consequence of transneural degeneration that results from any pathological lesion interrupting the dentato-rubral (superior cerebellar peduncle; SCP) or rubro-olivary tracts (central tegmental tract; CTT) (1–8). Close association between OE and palatal myoclonus or palato-pharyngo-laryngo-oculodiaphragmatic myoclonus has been repeatedly emphasized (2, 3, 5, 6, 9–17). While OE is often asymptomatic and without myoclonic movement (1, 6, 8, 11, 18–21), it is rarely associated with other involuntary movements, such as athetosis (4) or intention tremor (IT) (4, 12, 22, 23). We report two patients with vascular brain stem lesions who presented with IT, in whom OE was detected by magnetic resonance imaging (MRI). Palatal myoclonus appeared much later in one patient and did not appear in the other patient during the observation period. In the present paper, we report a specific condition in which OE and IT coexist, and discuss the possible association between these two conditions.

Patient Report

Patient 1

A 55-year-old man suddenly lapsed into semicoma. Hemorrhage was detected in the right pontine tegmentum upon computed tomography (CT). After regaining consciousness, the patient showed transient right lateral rectus palsy, ataxic dysarthria, left hemiparesis with deep sensory disturbance and truncal ataxia. Nine months after the stroke, he noticed progressive tremulousness of the left arm when reaching for objects. Neurological examination 19 months after onset revealed neither disturbance of consciousness nor extraocular muscle palsy. The patient exhibited a trace of left hemiparesis with hyperreflexia, moderate disturbances in tactile sensations and proprioception on the left side, and mild truncal ataxia. Due to the patient’s involuntary movements, we could not determine whether the patient had cerebellar limb ataxia in the left arm; he was not ataxic in the other limbs. Myoclonus of the palate or the neighboring muscles was absent on admission. Bilateral palatal myoclonus, however, appeared 49 months after onset of hemorrhage. Frequency ranged from 110 to 130/min, and was asynchronous with the IT of the left arm.

The involuntary movements of the left arm were more apparent when the patient was standing or under mental stress, and were absent when he was asleep or at complete rest. Muscles participating in the movements were the supra- and infraspinatus, teres major, pectoralis major, deltoid, biceps, triceps and, less vigorously, the extensors and flexors of the wrist. The patient habitually held his left hand against his back, reminiscent of a dystonic posture, so that these movements would not disturb him. The left upper extremity exhibited reciprocal, rhythmic, tremulous movements that were more
prominent in the proximal portion of the arm, and were easily induced when the patient raised and held his left arm in the air unassisted or pointed his finger at an object. When he tried to touch his nose with his left index finger, as the finger came closer to the face, the whole arm showed repellent oscillations, which hindered the finger from touching or remaining in contact with his nose (Fig. 1). His finger often hit his face, or knocked off his eyeglasses.

A superficial electromyographic (EMG) study during the finger-to-nose test showed group discharges with a frequency of approximately 5 c/s and a duration of 13 to 15 ms that were reciprocal between the left flexor carpi ulnaris and extensor carpi radialis longus, or the biceps and triceps brachii (Fig. 2). This dyskinesia was compatible with IT (24, 25), especially with the rhythmic type (26). Defective proprioception seemed to play little part in these involuntary movements, because visual compensation was completely ineffective in reducing dyskinesia. The patient also exhibited a slower (2.7 c/s) tremor in the left wrist and fingers that was evident at rest, but overwhelmed by IT when the left arm was in motion. Although this slow tremor seemed analogous to a parkinsonian resting tremor, myorhythmia was also possible, which differs from parkinsonian tremor with regard to its slower rate (2 to 3 c/s) (15, 27, 28). IT in this patient was resistant to treatment, but administration of clonazepam (8 mg/day) and a motor point block of the left supraspinatus muscle were partially effective.

MRI eight days after pontine bleeding revealed a hematoma at the right pontine tegmentum (Fig. 3 left), extending longitudinally to the mesencephalo-pontine junction (Fig. 3 right). The lesion involved the right CTT, but appeared to be located too low to involve the right SCP, its decussation, or the red nucleus. The hematoma, however, might have functionally involved a more extensive area in the pontine basis than was depicted on MRI, because the patient had left hemiparesis and truncal ataxia. The medulla oblongata appeared to be normal during the acute phase. On MRI 10 months after the appearance of IT, the primary bleeding site was depicted at the right pontine tegmentum (Fig. 4a, c). The right inferior olivary nucleus was swollen, and showed a low signal on T1-weighted images and a high signal on T2-weighted images (T2WI) (Fig. 4b, c). No abnormal intensities were detected in the dentate nuclei. Anomalies on MRI remained essentially the same after the onset of palatal myoclonus.
Patient 2

A 59-year-old man was admitted to a local hospital because of sudden right oculomotor palsy, mild left hemiparesis, and swaying of the left arm upon attempted use. These symptoms were almost resolved by the fifth day, when the patient noticed gradually aggravating trembling movements of the right arm.

Neurological evaluation upon admission to our hospital 75 days after onset revealed residual right oculomotor palsy without pupillary involvement, symmetrical hyperreflexia, and bilateral extensor toe signs, but no motor paresis, sensory disturbance or ataxia was observed. We did not detect limb ataxia in the right arm when it was held in a specific position to minimize dyskinesia. The patient showed involuntary movements confined to the right shoulder and arm, which oscillated rhythmically and worsened progressively as the patient tried to touch his nose with a finger. The patient’s dyskinesia was diagnosed as typical IT similar to that in patient 1, and was effectively controlled by administration of 2 mg/day of clonazepam. Patient 2 did not have a resting tremor such as was observed in patient 1. Superficial EMG revealed rhythmic (4 c/s), reciprocal contractions of the agonist and antagonist muscles of the right arm. No palatal myoclonus was detected on repeated examinations during the next six months up until the time patient follow up was lost.

On MRI four months after onset, a T1-weighted axial view revealed a narrow attenuated signal at the right paramedian tegmentum of the midbrain extending toward the cerebral peduncle (Fig. 5a). The right red nucleus, oculomotor nerve, CTT, decussation of the SCP, and a portion of the substantia nigra might also have been involved. The right olive appeared mildly swollen and showed decreased signal intensity (Fig. 5b). A sagittal T2WI of the brain stem (Fig. 5c) depicted an ovoid high intensity area at the right ventral medulla oblongata. On MRI, no abnormalities were observed in the dentate nuclei.

Discussion

IT concomitant with OE is rare. Aside from the two patients presented in this paper, only four patients exhibiting this combination have been reported in the literature (4, 12, 22, 23). Due to the rarity of this specific combination, there has been little discussion of the pathophysiology of these two phenomena. Isolated IT usually results from damage to the SCP (24, 25).

When the damage is below the decussation, IT appears ipsilateral to the lesion, and when the damage is above the decussation, IT appears contralaterally (24, 25, 29, 30). A lesion in the CTT can also produce contralateral IT (4, 25, 31). IT in patient 1 can be ascribed to interruption of the right CTT by a hematoma at the mid-pontine level. In patient 2, the persistent IT of the right arm might be a consequence of the infarct that involved the right SCP below the decussation. However, we lack sufficient information about the initial tremulousness of the left arm to make any further analytical assessment.

Conversely, the interruption of the right CTT in patient 1 might have caused right OE, which developed during the 20 months between onset and the first MRI examination. In patient 2, it seems reasonable to attribute the right OE to ischemic injury of the right red nucleus or CTT.

A review of pathological findings in the literature may help to clarify the association between lesion site and laterality of IT.
and OE. A 50-year-old woman presented with left-sided IT and oculo-facio-palato-pharyngo-laryngeal myoclonus. Autopsy revealed a lacune involving the right CTT, demyelination in the right SCP, and right OE (12). An 18-year-old man presented with skeletal myoclonus and resultant IT of the left extremities. A lacune at the pontine tegmentum involved both the right SCP and right CTT, and OE was observed on the right side (22, 23). A 42-year-old woman presented with myoclonus on the left side and terminal swaying of the right finger that the authors denoted as atypical IT. Autopsy revealed an infarction involving the right SCP, secondary degeneration of the left red nucleus, and left OE (32). A 43-year-old woman who showed bilateral IT had an old hemorrhagic lesion interrupting the right CTT, gliosis of the left red nucleus, and bilateral OE (4).

In summary, it can be assumed that when mainly the CTT is involved, the lesion usually results in contralateral IT and ipsilateral OE relative to the lesion, as in patient 1 and other documented patients (12, 22, 23). Furthermore, when damage to the SCP is below the decussation, ipsilateral IT and contralateral OE may occur as in the patient reported by van Bogaert and Bertrand (32). Finally, when a lesion involves the red nucleus or the post-decussated SCP, contralateral IT and ipsilateral OE may result (4). This simplified schema may be helpful in untangling the relation between lesion site and
Figure 5. MRI in patient 2 four months after disease onset. T1-weighted axial view (TR 300 ms, TE 14 ms) showed a right paramedian infarction (a) and swelling of the right olive, which showed a slightly lowered signal intensity (arrow in b). The right olive appeared in the ventral medulla oblongata as a high intensity area on sagittal T2-weighted image (TR 3.000 ms, TE 100 ms) (arrow in c). A midbrain infarction was depicted as a high-signal lesion (arrowhead in c).

laterality of IT and OE.

We should keep in mind that exceptions exist, such as when both the CTT and SCP, and possibly the neighboring structures, are involved. For example, while we would expect to find left IT as a consequence of right CTT damage in patient 2, IT actually appeared on the right side, probably due to involvement of the right pre-decussated SCP. This framework also fails to explain the rarity and inconstancy of the combined occurrence of IT and OE, while tegmental lesions are common. Possible explanations include the fact that the probability of their concomitant occurrence is very low when each phenomenon does not necessarily result from damage to the CTT or SCP, and the possibility that this combination may remain undetected due to their different latent periods: three weeks (7) to a few months (4, 33) for OE and nine months to three years for IT (25).

Electrophysiological interaction between OE and IT has not been discussed previously. The relationship between OE and palatal myoclonus is well known (2, 3, 6, 9–17, 34). OE presumably induces palatal myoclonus by stimulating the ambiguus nucleus (3). If IT is closely related to palatal myoclonus, then we may hypothesize that OE influences IT electrophysiologically.

In patient 1, IT emerged 40 months prior to palatal myoclonus, and the conditions were not synchronous. In patient 2, IT was present even when palatal myoclonus was absent. These findings suggest that IT is physiologically unrelated to palatal myoclonus, and thus to OE. Therefore, based on the patients we reported in the present study, we assume that OE and IT have no electrophysiological relationship.

References
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