

Dropped Head Syndrome Secondary to Parkinson's Disease: A Case Report

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Abstract

Dropped head syndrome occurs when the posterior musculature complex of the neck is weakened by different disorders, including Parkinson's disease. We present a 48 year-old female patient visiting our clinic because of neck weakness and difficulty in maintaining her horizontal gaze. Her past medical history revealed a Parkinson's disease discovered 6 months prior to admission, well controlled with oral medications. She then underwent a posterior cervical surgery including C2 to T3 instrumentation and correction in addition with C3 to C6 laminoplasty. After the operation, all radiological parameters returned to optimal values and the patient was satisfied with the result. Dropped head syndrome with failed conservative treatment can be successfully treated by surgical intervention with instrumentation from upper cervical to upper thoracic region and deformity correction.

Keywords: Dropped head syndrome; Parkinson's disease; Isolated neck extensor myopathy; Chin on chest deformity

Introduction

Dropped head syndrome (DHS) occurs when the posterior cervical muscles are weakened, resulting in a flexion deformity. The deformity is initially flexible and correctable, however with time it becomes rigid and difficult to deal with. DHS was described for the first time by Lange et al in 1986 [1] and until now there were only 129 cases fully reported with treatment offered in the English literature [3]. The etiology is not fully understood, and possible causes include amyotrophic lateral sclerosis, Parkinson's disease, myasthenia gravis, hypo or hyperthyroidism, Cushing syndrome, polymyositis, dermatomyositis... [2-5]. DHS can also be the consequence of a cervical myelopathy which compromises the activity of paraspinal muscles. When underlying condition is absent, the term isolated neck extensor myopathy (INEM) can be used alternatively. Conservative approach includes treatment of causative pathology (eg with Parkinson's disease, thyroid disorders) and immunosuppression (eg with myositis, polymyositis) [2-4], in parallel with bracing and physiotherapy. Surgical treatment comprising deformity correction and instrumentation can be considered when non operative solutions fail or concomitant cervical myelopathy exists [3]. Drain et al (2019) in their Meta-analysis stated that surgery alone or with immunosuppression might provide good outcome [3].

Case Report

A 48 year-old female patient came to the outpatient clinic of our hospital because of fatigue and weakness of the posterior cervical musculature complex that made her unable to maintain a horizontal gaze for a long duration. This condition began 3 years prior to admission with gradual evolution. 6 months before admission, she started to have resting tremors in both hands and has been diagnosed with Parkinson's disease. The conservative treatment included trihexyphenidyl and benzerazide that helped to control her tremors but not the cervical symptoms. Two weeks before admission, she developed radicular pain in both arms and the chin on chest deformity became permanent. During the entire history, she did not have fever, body or limb pain, ptosis, dermatologic lesions or any other indicative symptoms.

On physical examination, the prominent symptoms of the patient

is marked weakness of posterior paraspinal muscle of her neck with a power of 2/5. The flexion deformity (Figure 1) remained flexible with normal passive range of motion (Figure 2) and this corresponded to



Figure 1: Preoperative photo (A) with antecollis and postoperative photo (B) with restoration of cervical sagittal balance and horizontal gaze.

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DHS criteria of diagnosis). The muscle power of all four limbs was normal (5/5) and no sensory disturbance has been noticed when her head was held in neutral position with a hard collar. However she felt mild radicular pain when her neck was in full flexion or extension. Myelopathic tests including Hoffmann and Babinski were negative. Muscle reflexes were normal in all four limbs.

The EMG revealed a moderate chronic lesion of bilateral 5th, 6th

and 7th cervical nerve roots and no signs of motor-neuron disease have been detected. Needle EMG of the trapezius and splenius capitis showed minimal electrical activities with fibrillation potentials and some positive sharp waves whereas the stencleidomastoid's electrical pattern was normal (Figure 7).

Preop EOS film of our patient depicted severe sagittal imbalance (Figure 4A) with cSVA of 52.9mm, C2-C7 kyphotic angle of 18°, cervical



Figure 2: Lateral X-ray of patient's neck in (A) flexion, (B) neutral position and (C) extension. Note that the deformity is still flexible with normal range of motion.

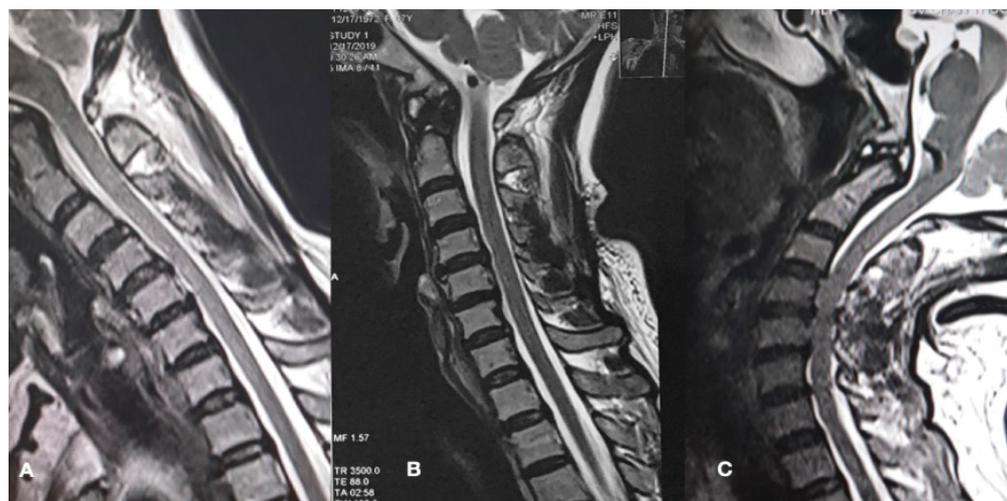


Figure 3: Mid-sagittal MRI scan of patient in flexion (A) with mild spinal cord compression at C5-6 and C6-7 levels, in neutral position (B) without compression and in extension (C) with severe compression at C4-5, C5-6 and C6-7 levels due to disc protrusion and yellow ligament buckling.

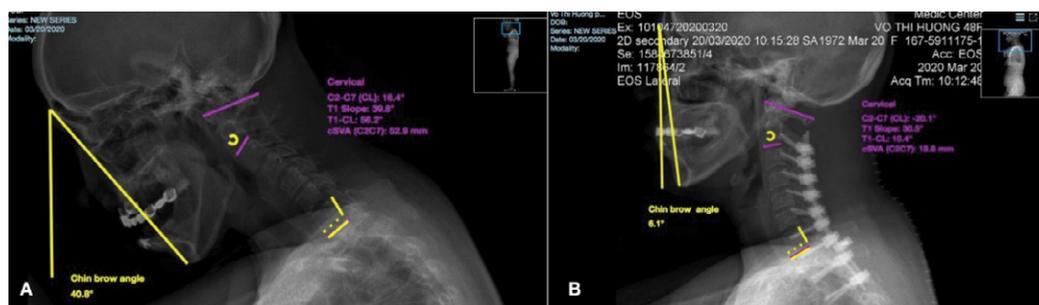


Figure 4: EOS scans of patient: Preoperative scan (A) revealed severe sagittal imbalance and postoperative scan (B) with optimal sagittal balance.

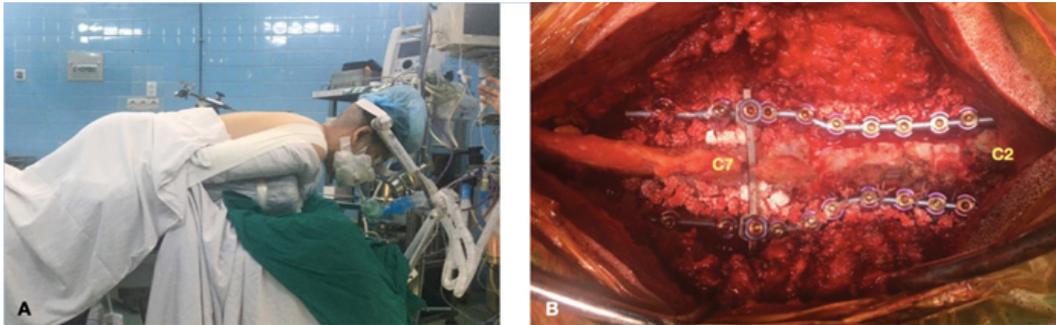


Figure 5: Positioning patient with Mayfield tong in prone position (A) and final construction prior to closure (B) (lateral mass screw fixation from C2 to C6, pedicle fixation from C3 to C7).

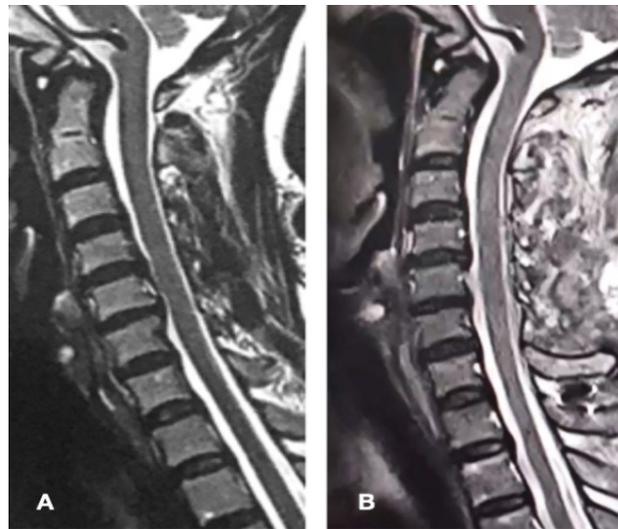


Figure 6: Mid-sagittal MRI scans of patient: preoperative scan (A) showed mild stenosis and postoperative scan (B) showed enlarged spinal canal after laminoplasty with good cervical lordosis

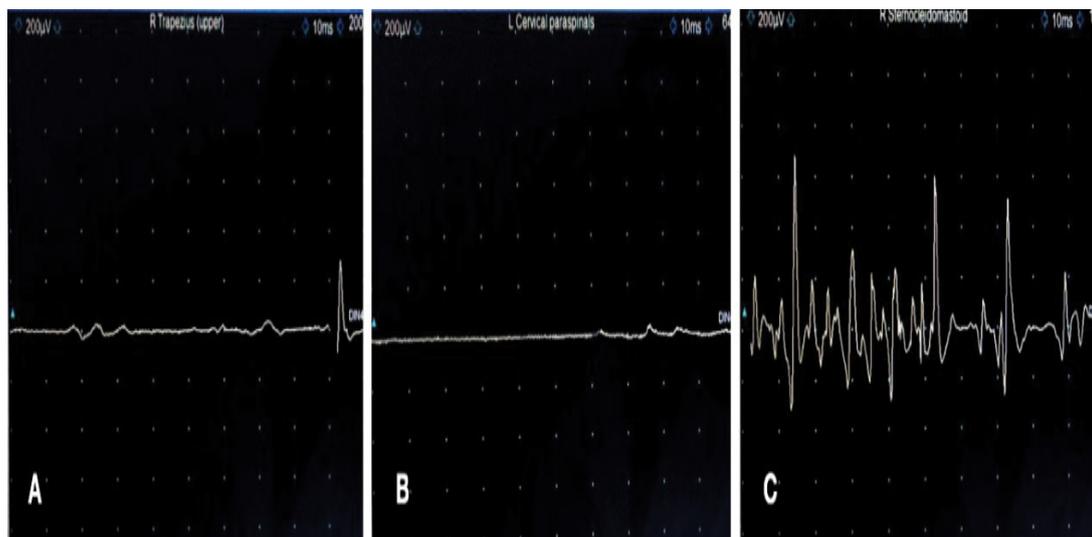


Figure 7: Needle EMG findings of the trapezius (A), splenius capitis (B) and sternocleidomastoid muscle (C) of patient. Note that the electrical activities of the posterior muscles are almost absent compared to normal potentials of the sternocleidomastoid muscle.

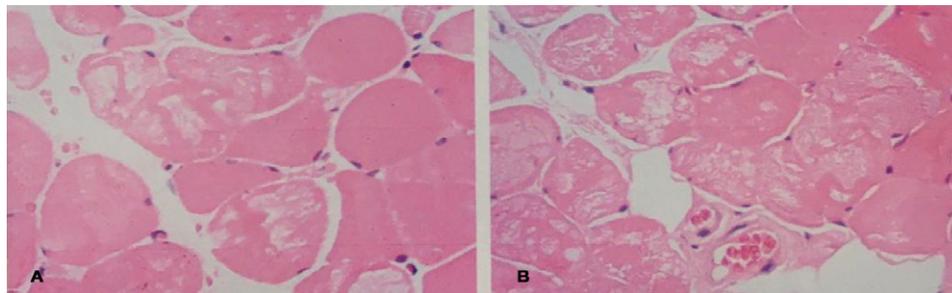


Figure 8: Trapezius specimen (A) and splenius capitis (B) showed focal necrosis with molt-eaten image among healthy fibers. Note the absence of inflammatory infiltration frequently seen in myositic conditions.

lordotic-T1 slope mismatch (CL-T1 slope) of 56° and chin brow angle of 40°. For the surgical planning, we applied the template method assisted by the Surgimap software (Nemaris, Inc) to calculate the amount of deformity correction necessary to restore sagittal balance. The result was -18° (cervical lordotic angle of 18°).

Our patient underwent MRI scan of cervical spine in three different positions: flexion, neutral and extension in order to detect dynamic compression of the spinal cord. The result showed a mild compression at C5-6, C6-7 in flexion and severe compression at C4-5, C5-6, C6-7 in extension. This was the reason why we decided to perform open-door laminoplasty in addition to instrumentation and correction. Lateral mass screw fixation was applied from C2 to C6 and pedicle screw fixation for C7 to T3. Open-door laminoplasty was then performed from C3 to C6. We have also taken specimen of the trapezius and splenius capitis for biopsy. One week after the surgery, our patient could stand up and walk with soft collar and she was happy with immediate surgical outcome when her horizontal gaze in the upright position was restored (Figure 1B).

The trapezius and splenius capitis specimens (Figure 8) showed focal necrosis with molt-eaten pattern without inflammatory infiltration.

Postop EOS scan was satisfying with all cervical sagittal balance parameters returned to normal values (Figure 4B). Postop MRI scan showed enlarged spinal canal with optimal lordosis compared to preop scan (Figure 6).

Discussion

One of frequently studied etiologies of DHS is Parkinson's disease (PD) [6-14]. In contrast to the scarcity of DHS in general population, this syndrome is not uncommon in Parkinson's patients [10]. The reason why PD can lead to DHS condition remains unclear, however neuromuscular disorder secondary to neuron degeneration is one of main characteristics of PD [15]. Neuromyopathy can occur in any muscle groups, but the posterior cervical musculature complex seems to be affected the most and the imbalance of contraction force of anterior and posterior muscles alone can lead to DHS [5]. Some studies have concluded that by weakening the anterior muscles (sternocleidomastoid and scalene muscles) with Botulinum toxin can somehow improve the situation [11]. In our case, the function of posterior muscles was severely compromised without contracture of anterior muscles, therefore the botox therapy was not considered. The clinical findings was confirmed by needle EMG with minimal electrical activities of posterior muscles. Our EMG findings were in line with that of other researches [6,15]. The muscle biopsy of our patient also confirmed a neuromuscular disorder with focal necrosis and no sign of infiltration [8,9,12,17] which frequently existed in autoimmune myositic pathologies [18].

Kashihara et al in 2008 studied 252 Parkinson's patients in Japan and noted that DHS occurred in 6% of that population. Female individuals are predominantly affected with the sex ratio female:male of 4:1. The mean age of onset was 63 [10]. The evolution of DHS was not parallel with that of PD in some individuals. In our patient's situation, PD was diagnosed at early stage with only resting tremors as indicative symptoms and the condition was well controlled with oral medications meanwhile DHS aggravated over time. Moreover, according to some studies [10,13,14], dopaminergic antagonists could induce or aggravate DHS. That was not the case of our patient.

We decided to surgically correct the chin on chest deformity by using posterior approach alone as the deformity was still flexible. There were minimal degenerative changes of the cervical spine, especially at the cranio-cervical junction and upper cervical spine, therefore we decided to stop at C2 as the upper most level to preserve cervical mobility. For the lowest ended vertebra, we preferred to stop at T3 in order to avoid junction syndrome and implant failure [5,6]. Finally, open-door laminoplasty was performed thanks to the cervical MRI scan in extension revealing severe multilevel spinal stenosis. Both postop X-ray and MRI scans were satisfying [19-23].

Conclusion

The remaining issue of our plan was to calculate the optimal amount of correction to restore the sagittal balance not only of the cervical spine but the spine as a whole. Under or over-correction would result in persistent sagittal imbalance. The preop EOS scan demonstrated pronounced sagittal imbalance of the head. To be specific, the cSVA was 52.9mm (optimal value < 40mm), the C2-C7 kyphotic angle was 18°, CL-T1 slope mismatch was 56° (optimal value was 16.5°), the chin brow angle was 40° (optimal value < 10°) [20-23]. By using the template technique assisted by the Surgimap software we could calculate the optimal cervical lordotic angle of 18°. This could be achieved during the operation by adjusting the position of the patient's head with the Mayfield tong and image intensifier. Postop EOS scan showed optimal sagittal parameters as planned with cSVA of 18mm and chin brow angle of 6.1°. The postop radiological findings correlated with clinical feelings of patient when she was satisfied with the immediate surgical result despite a mild movement restriction due to instrumentation.

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