Isolated Post-Traumatic Syringobulbia; Case Report and Review of the Literature

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Key­word­s: Syringobulbia; Traumatic brain injury; Syringomyelia; Post-traumatic; Syrinx; Cranial nerve palsy.

ABSTRACT

Syringobulbia is a very rare condition defined as slit-like fluid cavity in the brain stem. Several conditions have been reported to be associated with syringobulbia including neoplasms, spinal cord traumas or lesions such as tethered cord, hind-brain herniation, infections such as meningitis and in isolation. Although post-traumatic syringomyelia has been widely described previously, traumatic brain injury has not been reported as the mechanism and etiology of isolated syringobulbia. We herein report a 24-year old man with previous history of severe traumatic brain injury who presented with recent onset inability to walk or coordinate movements, ataxia, dysphonia, dysarthria, bilateral third nerve palsy with fixed dilated pupils and eyes deviated outward and downward. He was further diagnosed to have isolated syringobulbia extending to upper pons and lower midbrain. Isolated post-traumatic syringobulbia is extremely rare condition presenting insidiously with cranial nerve palsies, ataxia and dysarthria.

Keywords: Syringobulbia; Traumatic brain injury; Syringomyelia; Post-traumatic; Syrinx; Cranial nerve palsy.

Introduction

Syringobulbia is a very rare condition defined as slit-like fluid cavity in the brain stem. Several conditions have been reported to be associated with syringobulbia including neoplasms, spinal cord traumas or lesions such as tethered cord, hind-brain herniation, infections such as meningitis and in isolation. The clinical presentation depends on the site of the cavity and the involved structures which ranges from asymptomatic presentation to cranial nerve palsy, vocal cord paralysis and facial myokymia. Post-traumatic syringomyelia has been widely described previously with an incidence of 0.3-3.2% in those suffering from spinal cord trauma [10]. However, traumatic brain injury has not been reported as the mechanism and etiology of isolated syringobulbia. The current body of literature describing syringobulbia consists of case reports or small case series linked with syringomyelia. We herein report a patient with isolated syringobulbia following severe traumatic brain injury.

Case Report

A 24 year-old man presented to our neurosurgery
outpatient clinic with recent onset inability to walk or coordinate movements, ataxia, dysphonia and dysarthria. On physical examination he had hyperreflexia and bilateral third nerve palsy with fixed dilated pupils and eyes deviated outward and downward (Figure 1). His speech was apprehensible for his family, he was able to hear and understand voices and his gag reflex was normal and symmetric. He had normal symmetric muscle power in all extremities. In his past medical history, he was victim of motorcycle-vehicle accident 4 years earlier leading to deep brain contusions in basal ganglia and brain stem. He has been admitted with a GCS of 6 (M:4, V:1, E:1) and reactive pupils and was further diagnosed to suffer from diffuse axonal injury (Grade III). After ICU admission for 3 weeks and tracheostomy, he was discharged in vegetative state with severe disability. During the previous outpatient visits (1 year earlier) he had GCS: 13 (M:6, V:3, E:4) with reactive pupils, without any cranial nerve palsies and neurological deficit. Brain Magnetic resonance imaging (MRI) was requested which revealed syrinx formation in upper pons and lower midbrain (Figure 2). Cervical and thoracic MRI did not show any syringomyelia (Figure 3). No other pathology such as neoplasm, hind-brain stimulation or Chiariimal formation was found in MRI imaging of the patient. According to his previously healthy condition before suffering trauma, these clinical features of upper motor neuron involvement and cranial nerve palsies are attributable to formation of a syrinx within his brain stem.

**Fig. 1.** Bilateral 3rd nerve palsy with fixed dilated pupils and eyes deviated outward and downward in the patient with post-traumatic syringobulbia.

**Fig. 2.** Axial T1- (A) and T2-weighted (B) brain MRI image showing syrinx formation in upper pons and lower midbrain of the patient.
**Discussion**

Syringobulbia is an uncommon pathology of central nervous system which is applied to formation of a cavity within brain stem. It can be accompanied by syringomyelia either communicating or non-communicating with it [11]. However isolated syringobulbia is an extremely rare condition and a few reports are available in the literature about this pathology(5). It can be encountered as a late complication of syringomyelia as the cavity may extend toward brain stem. Several mechanisms have been introduced for the formation of the syringobulbia concurrent syringomyelia. One possible mechanism is the rupture of the paracentral cervical syrinx rostrally resulting in syrinx formation in the brain stem [12]. The other mechanism is dilation and propagation of the spinal central canal to the medulla oblongata and brain stem leading to syringobulbia. One possible mechanism for isolated syringobulbia is the extravasation and entrance of the fluid from the perivascular spaces and cisterns into the brain stem. Encephalomalacia at the site of previous brain stem contusions could also be the origin of cavitation and syrinx formation. In the present case we assume that the latter two mechanisms could explain the formation of isolated syringobulbia following traumatic brain injury. The interesting point is that the syrinx in syringobulbia does not have ependymal lining and usually communicates with fourth ventricle [13]. To the best of our knowledge this is the first report of isolated post-traumatic syringobulbia.

The clinical presentation of syringobulbia depends on the size, location and extension of the lesion. As the lesion usually involves the brain stem, the clinical signs and symptoms are bulbar signs, cranial nerve palsies, corticospinal and corticobulbar signs [13]. Most often the onset of the sign and symptoms is insidious, with dysphagia, hiccup, or hoarseness of voice (8, 9). Later, as the lesion becomes more extensive, the subjective symptoms may ameliorate or disappear being replaced by objective signs. Our patient presented with recent onset inability to walk or coordinate movements, ataxia, dysphonia and dysarthria. Bilateral third nerve palsy with fixed dilated pupils and eyes deviated outward and downward were the prominent physical finding (Figure 1). Cranial nerve palsies are among the common manifestations of syringobulbia [11]. It has been reported that nystagmus is the most common neurologic finding of syringobulbia, together with trigeminal sensory loss [6]. Greenlee and co-workers [11] reported cranial nerve dysfunction in all of their patients; most of their patients had multiple nerves affected. The patient presented here suffered from bilateral 3rd nerve palsy along with fix dilated pupils. This could be explained due to extension of the syrinx to the upper parts of the pons and lower segments of the midbrain. The oculomotor nucleus is located in the ventral parts of the lower segment of the midbrain in the gray substance of the floor of the cerebral aqueduct and extends in front of the aqueduct for a short distance into the floor of the third ventricle. The nerve fibers project to the tegmentum, the red nucleus, and the medial part of the substantia nigra, forming a series of curves with a lateral convexity, and emerge from the oculomotor sulcus on the medial side of the cerebral peduncle. Edinger-Westphal nucleus which is responsible for the autonomic functions of the oculomotor nerve, including pupillary constriction.
and lens accommodation is also located in the dorsum of midbrain along with tegmentum [14]. The syrinx in our case involved all the aforementioned nuclei resulting in the corresponding nerve palsies.

Treatment options are limited for the isolated syringobulbia although several approaches have been reported for syringobulbia concurrent syringomyelia [11,13]. As only few case reports have addressed isolated syringobulbia, the treatment option is experimental models and evidence regarding this issue is extremely scarce in the literature. Greenlee et al., [11] showed that posterior fossa decompression is a safe and effective treatment. Other surgical options include drainage or decompression of the syrinx cavity with diversion of fluid to the subarachnoid space or peritoneal cavity [15]. As the data is scarce regarding the treatment of isolated syringobulbia, the results are variable and the treatment option should be chosen based on underlying etiology, to achieve the best outcome.

In conclusion, isolated post-traumatic syringobulbia is extremely rare condition presenting insidiously with cranial nerve palsies, ataxia and dysarthria. Although rare, it should be kept in mind in those suffering from severe traumatic brain injury.

Conflict of Interest: None declared.

References