Case Report

Gowers’ intrasyringeal hemorrhage associated with Chiari type I malformation in Noonan syndrome

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Abstract

Background: Idiopathic hemorrhage in a syrinx is a rare entity known as Gowers’ intrasyringeal hemorrhage. Bleeding confined to the syrinx cavity causes severe, sometimes acute, neurological deficits. We report a case of intrasyringeal hemorrhage into a preexisting lumbosacral syrinx associated with Chiari type I malformation.

Case Description: A 39-year-old female with Noonan syndrome underwent foramen magnum decompression and a cervical syrinx-subarachnoid shunt for Chiari type I malformation-associated syringomyelia 7 years ago. She presented progressive gait deterioration and acute urinary dysfunction, indicating conus medullaris syndrome. Initial magnetic resonance imaging revealed massive hemorrhage in the intrasyringeal cavity of the conus medullaris. The patient underwent surgical removal of the intrasyringeal hematoma and her neurological symptoms improved postoperatively.

Conclusion: Although Gowers’ intrasyringeal hemorrhage is rare, this entity should be taken into consideration in patients with syringomyelia showing acute neurological deterioration.

Key Words: Chiari type I malformation, intrasyringeal hemorrhage, Noonan syndrome, spinal disease, syringomyelia

INTRODUCTION

Intrasyringeal hemorrhage was first described by Gowers in 1904, and was named Gowers’ syringeal hemorrhage by Wilson in 1955.[1] The hemorrhages have mostly been observed in syringomyelic cavities, and patients present with a sudden onset, and/or acute and rapid worsening, of pain. Since increased intrasyringeal pressure caused by hemorrhage may result in irreversible neurological sequelae, it is important not to overlook the symptoms caused by intrasyringeal hemorrhage in patients with syringomyelia.

Here, we report a rare case of Noonan syndrome and spontaneous intrasyringeal hemorrhage associated with Chiari type I malformation (CMI). The patient underwent surgery for evacuating the hematoma. The patient has experienced neither recurrence nor clinical deterioration for 3 years in follow-up.

CASE REPORT

A 39-year-old female presented with progressive gait disturbance and urinary dysfunction of acute onset in 2010. She had been treated for tetralogy of Fallot
in childhood and had been diagnosed with Noonan syndrome on the basis of molecular genetic analysis. She had short stature, webbing of the neck, low-set ears, ocular hypertelorism, ptosis, and developmental delay. She also had multiple untreated cerebral aneurysms. She had CM1 with cerebral hydrocephalus and a large syringomyelia extending from C1 level to the conus [Figure 1]. Although she underwent foramen magnum decompression and a cervical syrinx-subarachnoid shunt in 2003, a large syringeal cavity remained throughout the spinal cord.

On admission in 2010, she experienced gait disturbance and dysesthesia of both the legs and perineal region, which indicated conus medullaris syndrome. Routine hematological, serological, and blood coagulation examinations were all normal. Magnetic resonance (MR) imaging showed a large syringomyelic cavity extending from the C1 level to the conus. Both T1- and T2-weighted images showed a heterogeneous mass on the caudal end of the syringomyelic cavity, suggesting hematomyelia [Figure 2]. Partial gadolinium enhancement was revealed at the centre of the hematomyelia.

Emergent laminectomy was performed from the T12 to L3 level because she developed complete urinary retention. A swollen spinal cord showed xanthochromic deposits because of hematomyelia. After midline myelotomy, dark-red hematoma flowed out, and was carefully removed with suction and CUSA®. During surgery, we could not detect the precise bleeding point. The cranial and caudal edge of the hematoma showed transition into yellowish and firm tissue. Since we suspected this tissue to be intramedullary neoplasm, several pieces of this neoplasm-like tissue were sampled. However, histological examination of the resected tissue showed gliotic spinal cord tissue with hemosiderin pigmentation. The surgical specimen revealed diffuse growth of spindled astrocytic cells with Rosenthal fibers that had no atypical features. We diagnosed spontaneous intrasyringeal hemorrhage associated with CM1, that is, Gowers’ intrasyringeal hemorrhage.

Postoperatively, the patient’s symptoms disappeared except for the sensory disturbance of the lower limbs. Repeated MR imaging performed up to 3 years after the operation showed marked reduction of spinal cord diameter and shrinkage of the syringeal cavity [Figure 3]. There was peripheral hemosiderin deposition but no intramedullary neoplasms in the syringomyelic cavity.

**DISCUSSION**

Hematomyelia usually presents with an acute onset and rapid deterioration in neurological status and usually leads to an acute spinal cord syndrome. Intrasyringal hemorrhage may occur spontaneously in the course of acquired coagulation disorders, may result from trauma, or may be caused by surgical treatment of syringomyelia. Hamlet et al. reviewed 13 cases of Gowers’ intrasyringeal hemorrhage.
hemorrhage.\(^3\) Although there was no specific clinical picture, the most common characteristic features of intrasyringeal hemorrhage were acute onset of symptoms and/or rapid neurological deterioration.\(^1,3,7,9\) When bleeding occurs, it is confined to the preexisting syrinx cavity and causes increased intrasyringeal pressure. It has been suggested that the cause of bleeding might be a rupture of the blood vessels that have been observed on the syrinx cavity wall.\(^5,7\) Another theory is the existence of slow venous hemorrhage that may originate from a torn intraspinal vein deprived of its normal neural and glial support.\(^1,3,9\) At the same time, there are some reports that the symptoms of intrasyringeal hemorrhage chronically deteriorated for several months.\(^3,7\)

Syrinx cavities often coexist with primary intramedullary neoplasms, like glioma. Ependymoma might be the most likely glial tumor to present a diagnostic dilemma in cases of hematomyelia concomitant with intramedullary mass lesion because of its typical central location within the spinal cord, its complex structure with solid and cystic elements, and its definite propensity to hemorrhage.\(^6\) Partial gadolinium enhancement was revealed at the centre of the hematomyelia in our case, but the surgical specimen showed only gliotic spinal cord tissue without rosette formation. Hemangioblastoma contains enlarged arterial feeding vessels, a densely staining tumor nodule, and rapid shunting into a distended venous structure seen on an angiogram.\(^10\) A case of intrasyringeal hemorrhage associated with hemangioblastoma in the epiconus was previously reported,\(^10\) but MR imaging did not show the presence of dilated vessels in hemangioblastoma in our case. Although intraoperative findings suggested the coexistence of hemorrhagic spinal tumors, histopathological findings did not confirm the presence of neoplasm, and hence, we diagnosed our case as idiopathic hematomyelia in a preexisting syringomyelic cavity. No intramedullary neoplasms became evident in the 3-year follow-up period.

Noonan syndrome is a relatively common congenital genetic disorder with an estimated prevalence of 1 in 1000 to 1 in 2500 live births. Characteristic findings include distinctive facial features, short stature, chest deformity, and congenital heart disease. Until recently, diagnosis was based solely on clinical findings, but genetic mutations are identifiable in ~61% of the patients.\(^8\) There are a few reports of Noonan syndrome coexisting with CM1.\(^2,4,5\) Keh et al. reviewed seven cases of CM1 associated with Noonan syndrome, and suggested that other genetic mutations may affect the posterior cranium and create appropriate conditions for CM1 to develop. This is because the most common genetic mutation in Noonan syndrome tends to cause frontal and facial abnormalities, and the posterior fossa tends to be relatively spared.\(^1\) They said that a statistically significant association between Noonan syndrome and CM1 is difficult to obtain at present due to availability and logistical issues with scanning many asymptomatic patients.

In our case, the patient was diagnosed with Noonan syndrome in childhood and had some related disorders. We think that the Noonan syndrome was associated with CM1, but have no idea about association between Noonan syndrome and intrasyringeal hemorrhage. At 7 years before hemorrhage, our case had been operated twice, foramen magnum decompression and a cervical syrinx-subarachnoid shunt. The shunt catheter was not thought to cause the hemorrhage because the catheter was stable in cervical potion. There is a tendency to generally overlook the deterioration of clinical signs and symptoms in such patients, but it must be emphasized that neuroimaging should be performed if acute or sub-acute neurological deterioration occurs in patients with syringomyelia.

**CONCLUSION**

Gowers’ intrasyringeal hemorrhage is a rare entity characterized by insidious onset in patients with syringomyelia. Radiographically, it mimics imaging features of some intramedullary neoplasms. We have to emphasize intrasyringeal hemorrhage as a differential diagnosis in hemorrhagic spinal lesion, and timely neuroimaging for early recognition and therapeutic management.

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Commentary

Idiopathic hemorrhage in a Syrinx (also known as Gowers’ intrasyringeal hemorrhage) is a rare event in the overall clinical history of a Syringomyelia (SM) cavity.\[1,3\]

It generally occurs within large Syrinx cavities, presents with a fast and phasic clinical deterioration, and has a peculiar heterogeneous aspect on magnetic resonance imaging (MRI) imaging.

Nonidiopathic causes of intrasyringeal hemorrhages are iatrogenic, traumatic (including barotrauma), dyscrasic (coagulation disorders), and neoplastic (ependymoma and hemangioblastoma).\[4\]

The association of a Noonan syndrome in the case described in this article was likely to be true, true, and unrelated to the underlying intrasyringeal hemorrhage.

In both idiopathic and nonidiopathic cases, the size (girth more than length) and the intraluminal pressure of the SM cavity are important pathophysiologic factors in the development of this rare complication.

There are two corollaries to the case presented here:
1. An asymptomatic but large Syrinx is never totally safe for the patient
2. A persistently large Syrinx after a posterior fossa decompression is not an acceptable anatomical outcome.

In our center, since the late 90s, the presence of an asymptomatic but large SM (defined as ≥75% of the transverse diameter of the cord) secondary to an asymptomatic (or oligosymptomatic) Chiari I Malformation (CMI) has been an indication for posterior fossa decompression, because not only does a large syrinx expose the patient to the rare risk of an intrasyringeal hemorrhage, but it invariably leads to a progressive atrophy of the surrounding spinal cord parenchyma over the course of several decades, via an accelerated mechanism of wear and tear related to expansile compression and transmural ischemia, with negative effects on the functional reserve of the spinal cord.

For the same reasons, the lack of deflation of an asymptomatic but large SM cavity after a posterior fossa decompression cannot and should not be considered an acceptable postoperative anatomical result, and should warrant a more aggressive surgical revision of the posterior fossa.

After some initial resistance, the concept and the importance of posterior fossa revisions have gained a widespread appreciation within the neurosurgical community,\[2,5\] due to a number of factors, including the proliferation of specialized centers focused on these disorders, an increasing body of literature on the topic, and the presence of a large patient base accumulated in the MRI era.

Unlike the field of spine, in which surgical revisions have a quite predictable and uniform pattern for a number of given scenarios, surgical revisions in CMI/SM do not have a codified playbook, besides the general concept that “more” needs to be done.

Some surgeons buy extra space “on the outside”, with larger bone work and/or different kinds of duraplasties. Other surgeons buy extra space “on the inside” with different techniques of tonsillar reduction.

The exploration of the Foramen of Magendie can sometimes lead to the discovery of a Velum, an embryological remnant made up of nervous tissue, totally or partially obliterating the Foramen itself, and hence explaining the persistence of a large SM after an initial decompression.

Other times, arachnoid scarring from a former surgery is found to be responsible for an obliterated Foramen of Magendie.

Unlike the strategies of the past, the modern thinking recommends that SM shunting should be considered in CMI/SM only after an aggressive posterior fossa revision has failed to effectively deflate the lesion.

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