

# POSTERIOR FOSSA ARACHNOID CYST EXTENDING TO FORAMEN MAGNUM INDUCES COMPRESSION OF CEREBELLUM, CERVICO MEDULLARY JUNCTION AND SYRINGOMYELIA- A CASE REPORT

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## Abstract

*It is very rare that a posterior fossa arachnoid cyst extending to foramen magnum induces compression of cerebellum, cervico medullary junction and syringomyelia. Currently there are a few treatment experiences available. Here we reported the case of a 18 years old male patient who admitted to the hospital with the complaints of insidious onset of weakness and decrease sensation of left upper limb for 1 year. MRI revealed posterior fossa arachnoid cyst associated with syringomyelia. Posterior fossa decompression, arachnoid cyst excision and establishment of CSF flow were performed. Post of MRI showed established CSF flow but syrinx still remained. Despite this, the symptoms of the patient were obviously improved compared to before surgery. Thus, for the treatment of posterior fossa arachnoid cyst with compression of cerebellum, cervico medullary junction and syringomyelia, is excision of arachnoid cyst as much as possible with complete decompression of the posterior fossa and establishment of CSF flow results in a satisfying outcome.*

**Keywords:** Arachnoid cyst, posterior fossa, cerebellum, foramen magnum, syringomyelia.

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## Introduction

The commonest type of posterior fossa lesion that causes compression of cervico medullary junction and development of syringomyelia is the Chiari malformation type I<sup>[1]</sup>. Other types of arachnoid cysts can occur as an occupied lesion in the posterior fossa and in Dandy-Walker syndrome<sup>2-10</sup>. Occasionally, a posterior fossa arachnoid cyst can induce compression of the spinal cord and development of syringomyelia<sup>11,12</sup>. Common features of these lesions are secondary cerebellar tonsillar herniation with syringomyelia due to mass effect, and the lesions cross most areas of the foramen magnum. It is very

rare that a posterior fossa arachnoid cyst extending to foramen magnum induces compression of cerebellum, cervico medullary junction and syringomyelia. Here we reported a rare case of such type. We performed surgery on this patient. Meanwhile, we undertook a literature review on this topic as well, in order to provide better understanding and relate our experience in the diagnosis and treatment of such type of arachnoid cyst.

## Case Report

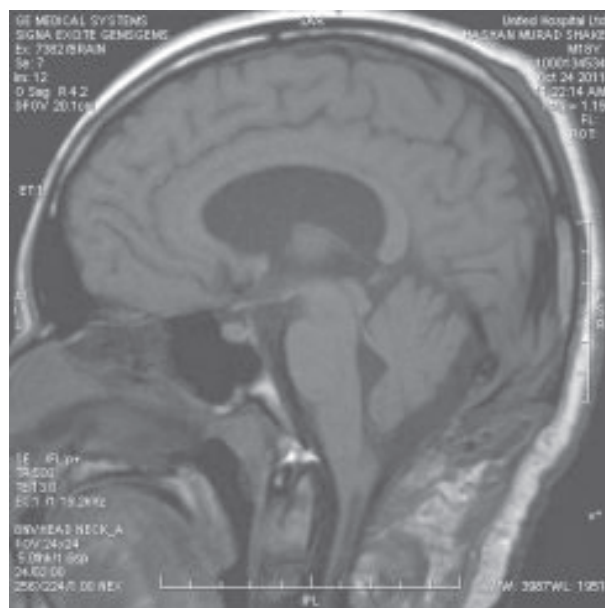
A male patient, 18 years old, was admitted under neurosurgery through OPD in United Hospital, Gulshan, Dhaka in October 2011 due to with the complaints of worsening weakness, and numbness in left upper limb for last 1 year. The patient had a history of fall from 20 feet height 2 years back. Physical examinations showed diminished superficial sensation in left C4,5,6 area, muscle wasting of left deltoid, trapezius and sternocleidomastoid (grade III muscle power). Reduced deep tendon reflex left upper limb, knee & ankle jerks was exaggerated, negative Babinski's sign. Magnetic resonance imaging (MRI) revealed cystic lesion in the posterior fossa extended to foramen magnum compressing the cerebellum & cervicomedullary junction. The lesion in T1WI imaging appeared as a low signal, and as a high signal in

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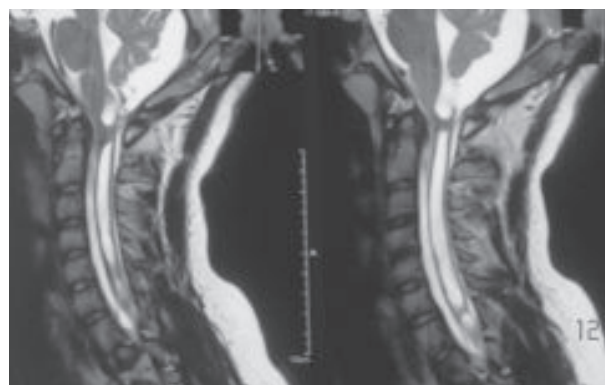
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T2WI imaging. The cerebellar tonsil was compressed upwards, the pons and cervical spinal cord appeared notch-like due to the compression of the cyst. A syrinx was seen in the spinal cord with a low T1W1 signal and a high T2WI signal down to D4. The size of the supratentorial ventricular system was mildly dilated. Based on history and physical and MRI examinations, we diagnosed the lesion as a arachnoid cyst with syringomyelia.

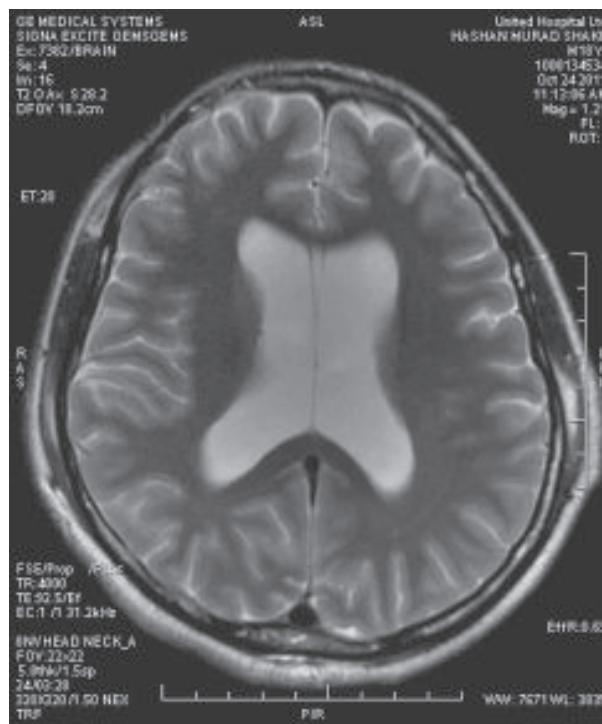
Presurgical MRI examinations. T1WI imaging showed the lesion as a lower signal; the cerebellar tonsil was compressed and moved upwards. The pons and cervical spinal cord anterior to the lesion appeared notch-like (Fig. 1). T2WI imaging showed the lesion as a high signal (Fig. 2). Head MRI revealing a dilated ventricle (Fig. 3). MRI showing a cystic lesion in posterior fossa.



**Fig.-1: MRI of brain (T1W1)**



**Fig.-2: MRI of brain & cervical spine (T2W1)**



**Fig.-3: MRI of brain (T2W1)**

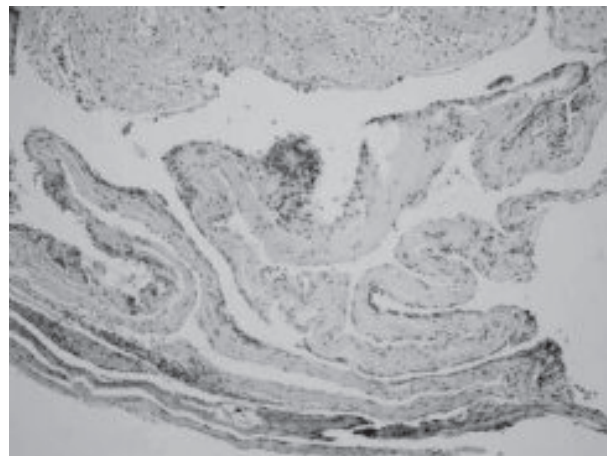
Decompression of the posterior fossa and excision of the arachnoid cyst were then surgically performed. A straight median incision was made from occiput to C4. Posterior fossa decompressive craniectomy, posterior edge of the foramen magnum and posterior arch of the atlas were then fully decompressed, followed by opening the dura mater. A cyst was seen to be located behind the cerebellum and posterior part of the cervicomedullary junction. The cerebellar tonsil was compressed and pushed upwards. After opening the cyst, it was seen that there were multiple compartments within the cyst. The cyst was tightly adhered to the cerebellum, and cervicomedullary junction. Wall of the cyst was excised. The tissue was sent for histopathological examination. Micro dissection was done & wall was separated from tonsil and cervicomedullary junction. Opening of foramen magnum & Luschka were done & CSF flow was established. A suture wound closure of the dura mater was performed using artificial mesh repair.

The postoperative symptoms were slightly improved compared to pre-surgery. The results of pathological examination confirmed arachnoid cyst (Fig. 5). During three months of follow-up, the condition of this patient continued to improve. Physical examinations showed that muscle power of left upper limb increased to grade IV. However, there was no improvement in muscle

wasting. MRI re-examination showed that the compression on the cerebellar tonsil, pons and cervical spinal cord was significantly reduced, the size of the syrinx was still the same as before surgery (Figure 4), but normal CSF flow is established.



**Fig.-4: Post surgical MRI of brain (T2W1)**



**Fig.-5: Histopathology**

## Discussion

There are two causes of compression of the spinal cord and syringomyelia induced by a foramen magnum lesion. One is a primary Chiari malformation type I, in which the cerebellar tonsil herniates into the foramen magnum and spinal canal to compress the spinal cord, consequently causing blockage of the spinal canal and the stoppage of cerebral spinal flow, leading to syringomyelia<sup>13,14</sup>. The other is an occupied lesion in the posterior fossa pushing the cerebellar tonsil

downwards to develop a malformation, which is similar to Chiari malformation type I. Examples of foramen magnum lesions reported in the literature include Klekamp et al.<sup>2</sup> who reported 3 cases of posterior fossa tumor in 1995, Bhatoe et al.<sup>3</sup> reported one case of meningioma in the cerebellar tentorium in 2004, Muzumdar et al.<sup>4</sup> in 2006 and Wu et al.<sup>7</sup> in 2010 each reported one case of pilocytic astrocytoma in the posterior fossa, El Hassani et al.<sup>5</sup> reported one case of cerebellar vermis medulloblastoma in 2009, and Suyama et al.<sup>6</sup> reported one case of a dermoid tumor in the cerebellum in 2009. These cases were all due to secondary cerebellar tonsillar herniation associated with syringomyelia, induced by an occupied lesion in the posterior fossa. The only large scale case study has been done by Tachibana et al.<sup>8</sup>, who in 1995 showed that in 164 cases of posterior fossa tumor, twenty-four (14.6%) had secondary cerebellar tonsillar herniation. Of these, only 5 cases (20.8%) were complicated with syringomyelia. Apart from the tumors mentioned above, some arachnoid cysts in the posterior fossa also cause similar changes to that in Dandy-Walker syndrome.<sup>9,10,15,16</sup>

Most posterior fossa arachnoid cysts result in cerebellar tonsillar herniation, consequently leading to compression of the spinal cord and syringomyelia due to the effect of the mass<sup>15-18</sup>. It is extremely rare for the posterior fosa arachnoid cyst to directly compress the spinal cord and develop syringomyelia. In 2000, Jain et al.<sup>19</sup> reported one case of a giant posterior fossa arachnoid cyst extending into the spinal canal to compress the spinal cord and develop syringomyelia; Kiran et al.<sup>20</sup> in 2010 also reported such a case. Although the case we reported here had similar features to these two cases, differences exist. The cyst in our case occupy most areas of the posterior fossa as these two cases did, instead it extended across the foramen magnum into the spinal canal at the level of the atlas. Thus, the lesion in our case was extremely rare.

With the report of this case we also did a literature review in order to have a better understanding of arachnoid cysts. Currently, the noncongenital causes of arachnoid cysts are unclear. It has been hypothesized that infection, trauma, circulation of the cerebrospinal fluid (CSF) and/or changes in CSF pressure contribute to the formation of arachnoid cysts. It is generally accepted that fossa arachnoid cyst may be a congenital malformation due to the



dynamic CSF pressure changes during development, leading to tearing of the arachnoid mater<sup>21-23</sup>. The patient we reported here had a history of trauma at 16 years of age; he recovered after treatment. Although arachnoid cyst associated with trauma is uncommon, such cases have been reported. Because it is very difficult to know whether the cyst is congenital or acquired, it is unclear whether trauma was the cause of the posterior arachnoid cyst formation. Nevertheless, whatever the cause the patient had 1 year of clinical presentation and his condition had worsened in the past year. MRI revealed that the arachnoid cyst extended across the foramen magnum to compress the spinal cord, and thus surgical treatment was considered. Surgical indications should be considered when an arachnoid cyst becomes progressively enlarged and compresses surrounding blood vessels, leading to corresponding symptoms gradually worsening<sup>24-26</sup>. The features of the present case was considered a suitable standard for surgical indication. Thus, surgical treatment was performed in this case.

There are several types of treatment for arachnoid cyst, including cyst fenestration, cysto peritoneal shunting and complete or partial excision. The most effective treatment is excision of the whole wall of the cyst to effectively prevent recurrence, particularly posterior fossa tumor<sup>24,27,28</sup>. Posterior fossa arachnoid cyst usually occupies the cerebellopontine angle, a condition from which most experience in its treatment has been obtained. For example, Samii et al.<sup>29</sup> in 1999 reported 12 cases of posterior fossa arachnoid cyst which extended into the cerebellopontine angle. Prognosis in most cases were good after excision of the cysts. However, the location of the cyst in the case we reported here was special and although simple excision can effectively prevent reoccurrence, it was difficult to release the pressure on the spinal cord and the establishment of CSF flow or relieve syringomyelia due to the pathological changes similar to Chiari malformation type I. Based on the standard treatment of Chiari malformation type I, we thought that sufficient decompression of the posterior fossa and dural suture closure would have a better treatment effect<sup>30,33</sup>. It has been shown that decompression is certainly effective in patients with Chiari malformation type I associated with syringomyelia. Aghakhani et al.<sup>30</sup> reported 157 cases of treatment of Chiari malformation type I. Clinical improvement occurred in 63.06% of these cases, and the percentage of

reduction in syrinxes was 90%. Wetjen et al.<sup>31</sup> in 2008 reported 29 cases in which 94% of the patients had improved symptoms. During 3-6 months of follow-up, MRIs revealed a decrease in syrinx size to a varied extent. Heiss et al.<sup>32</sup> in 2010 reported 16 cases of Chiari malformation type I, and syrinxes decreased in 15 patients (94%) after decompression.

The patient we reported here had markedly improved symptoms after decompressive treatment. However, MRI demonstrated no reduction in the size of syrinx 3 months after surgery. We consider this to be related to the short follow-up period.

### Conclusion

Thus, we think, for the cases of posterior fossa arachnoid cyst with compression of the spinal cord and syringomyelia, even if the arachnoid cyst could not be completely excised, excision should be performed as much as possible with complete decompression of the posterior fossa, which may result in a satisfying outcome.

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