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Case Report

Giant posterior fossa arachnoid cyst causing tonsillar herniation and cervical syringomyelia

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Abstract

Acquired cerebellar tonsillar herniation and syringomyelia associated with posterior fossa mass lesions is an exception rather than the rule. In the present article, we describe the neuroimaging findings in a case of 28-year-old female patient presented with a history of paraesthesia involving right upper limb of 8-month duration. Magnetic resonance imaging showed a giant retrocerebellar arachnoid causing tonsillar herniation with cervical syringomyelia. The findings in the present case supports that the one of the primary mechanism for the development of syringomyelia may be the obstruction to the flow of cerebrospinal fluid causing alterations in the passage of extracellular fluid in the spinal cord and leading to syringomyelia.

Key words: Arachnoid cyst, Chiari malformation, hydrocephalus, posterior fossa, syringomyelia

INTRODUCTION

Acquired cerebellar tonsillar herniation and syringomyelia associated with posterior fossa mass lesions is an exception rather than the rule.[1-4] There are only few case reports where the Chiari malformation and syringomyelia were probably caused by an arachnoid cyst resulting in tonsillar herniation[2,5-7] either by retrocerebellar[8,9] or at the level of foramen magnum.[10] In the present article, we describe the neuroimaging findings in a case of giant retrocerebellar arachnoid causing tonsillar herniation with cervical syringomyelia.

CASE REPORT

This was a case report of a 28-year-old female patient presented with a history of paraesthesia involving right upper limb of 8 months duration. She had a spontaneous abortion 3 weeks ago. Following that, she developed sudden onset of weakness of both lower limbs (left side greater than the right side). There was a history of headache on and off increased by coughing. Her bowel and bladder functions were normal and also general and systemic examination was normal. Cranial nerves were normal. Fundus examination was normal. She had a weakness of all four limbs (lower limbs more than upper limbs). Deep tendon reflexes were exaggerated in lower limbs and sluggish in upper limbs. Planters were extensors. There was also dissociated sensory loss on her right arm and shoulder. Patient was investigated with magnetic resonance imaging (MRI) brain and cervical spine and it showed giant retrocerebellar arachnoid cyst extending into the left cerebellopontine angle causing descent of the cerebellar tonsils across the foramen magnum with a syringomyelic cavity extending from C1 down and associated ventriculomegaly [Figure 1]. The patient underwent midline suboccipital craniectomy, foramen magnum decompression and C1 arch excision with duroplasty. Part of the cyst wall was excised. Histopathological examination of the cyst wall showed a cuboidal layer of arachnoid cells suggestive of arachnoid cyst [Figure 2]. Follow-up images showed a reduction in the size of the cyst and reduction in the mass effect over cerebellum and brain stem structures [Figure 3]. At 10 months follow-up, patient made an uneventful recovery and doing well.

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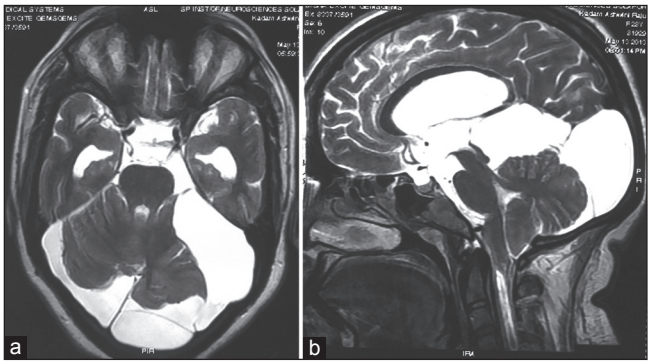

Figure 1: Magnetic resonance T2W images showing (a) a large posterior fossa arachnoid cyst more on left side extending into the left cerebellopontine angle with marked compression of the left cerebellar hemisphere and (b) there is caudal displacement of the cerebellar tonsils with obstruction of foramen magnum and associated syringomyelia

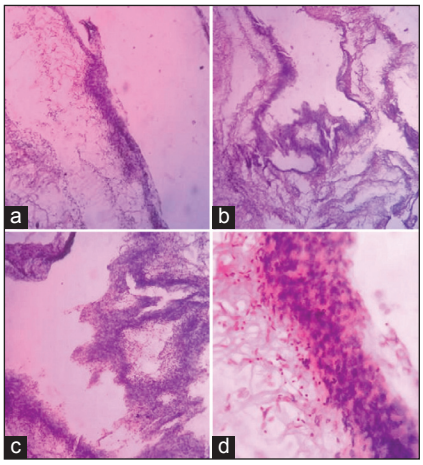

Figure 2: (a-c) Section showing cyst wall lined by meningeal cells showing stratification with subepithelial tissue showing acute inflammatory cells (H and E, ×100), (d) Section showing cyst wall lined by meningeal cells showing stratification with neutrophilic infiltrate and the subepithelial fibrocollagenous tissue shows acute inflammatory infiltrate (H and E, ×400)

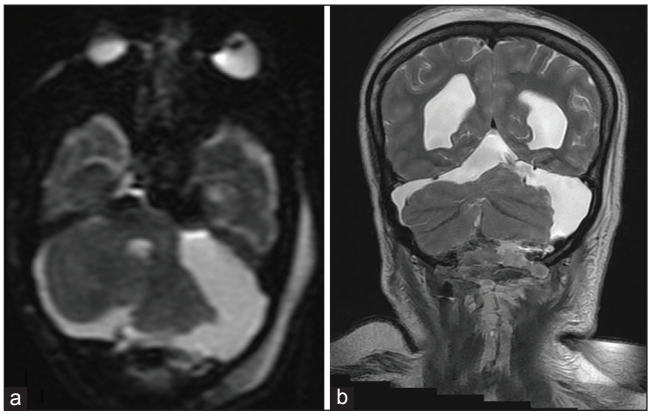

Figure 3: Following magnetic resonance images showing the reduction in the size of cyst and also reduction in the mass effect over cerebellum and brain stem structures

DISCUSSION

Arachnoid cysts accounting for approximately 1% of all intracranial mass lesions are benin collections of cerebrospinal fluid (CSF) developing as the result of an abnormal duplication of the arachnoid during the development of the brain.[11] With the advent of MRI many cases of large arachnoid cysts associated causing tonsillar herniation and syringomyelia have been recognized.[12-15] The primary mechanism for the development of syringomyelia is the obstruction to the flow of CSF causing alterations in the passage of extracellular fluid (ECF) in the spinal cord and leading to syringomyelia.[3-6,12,16,17] These patients with secondary cerebellar tonsillar herniation with syringomyelia usually present with the features due to mass effect of the primary lesion and associated syrinx.[8,10,16] Removal of the underlying pathology and establishing the normal CSF flow at the foramen magnum is the main stay of treatment for acquired Chiari and syringomyelia.[1-3,5,12] Arachnoid cysts can be managed with a variety of procedures including foramen magnum decompression, with or without cyst removal, cysto-peritoneal shunting or endoscopic cyst fenestration[15,18,19] As in the present case, posterior fossa decompression and cyst excision result in a good outcome.[1,10,20]

CONCLUSION

Based on the above study, it has been suggested that a simple blockage of CSF flow at the foramen magnum plays a vital role in the development of syrinx formation. The findings in the present case supports that the one of the primary mechanism for the development of syringomyelia may be the obstruction to the flow of CSF causing alterations in the passage of ECF in the spinal cord and leading to syringomyelia.

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