Indian Journal of Medical Case Reports ISSN: 2319–3832(Online)
An Open Access, Online International Journal Available at http://www.cibtech.org/jcr.htm
2014 Vol.3 (2) April-June, pp.33-35/Shankar et al.

Case Report

SYRINGOMYELIA – A RARE VARIANT

***J. Gauri Shankar, R. Tolstoy, Santini and Mohammed Zeya Ansari**
Department of Medicine, PSG Institute of Medical Sciences and Research, Peelamedu, Coimbatore, Tamil Nadu
**Author for Correspondence*

ABSTRACT
Syringomyelia is a well-known entity and it is due to formation of fluid filled cavity within the spinal cord. It usually involves the spinal cord and its extension into the medulla is also well reported leading onto cranial nerve palsies. Herewith we present a case who was diagnosed to have extension of the syrinx cavity upto the thalamus. To the best of our knowledge only ten such cases have been reported in English literature.

Keywords: *Syringocephalus, Syringocephalia, Syringoencephalomyelia, Syringomyelia*

INTRODUCTION
Syringomyelia - is a fluid-filled, gliosis-lined cavity within the spinal cord. Most lesions are between C2 and T9; however, they can descend further down or extend upward into the brainstem (syringobulbia). A syrinx can represent a focal dilation of the central canal, or it may lie separately, within the spinal cord parenchyma (Milhorat *et al.,* 2000). It is usually associated with an Chiari I malformation (Brickell *et al.,* 2006).
Herewith we report a case of syringomyelia which was diagnosed to have extension of the syrinx cavity upto the thalamus cranially and to the thoracic spine caudally with an Arnold Chiari I Malformation. To the best of the knowledge of the authors from various sources there have been only about 10 case reports of such instances in which the syrinx cavity extends higher above the brainstem and such extension is called as syringocephalia (Romero-Pinel *et al.,* 2006).

CASES
This 37 year old female presented to our hospital with sudden onset inability to walk and dysarthria following an episode of cough 15 days back. She also developed recurrent regurgitation episodes resulting in aspiration pneumonia. She was apparently normal 1 year ago when she developed insidious onset of left side numbness (including face) and diplopia on viewing objects on the right side.
On examination she was found to have pain and temperature loss on the left side of face (Lt V nerve / Rt thalamus), right side lateral rectus palsy (Rt VI nerve), right side palatal weakness and absent gag reflex (Rt IX and X nerve), right side tongue wasting (Rt XII nerve). Motor examination was apparently normal. Sensory examination revealed diminished posterior column sensation on the right side of the body and diminished anterolateral spinothalamic sensation on the left side of the body. Patient had stance ataxia with swaying to the right, in coordination on right side of the body (Rt cerebellum).
MRI brain and spinal cord shows a syrinx cavity that is extending from the thalamus to the thoracic spine with type I Arnold Chiari Malformation. The patient was operated and her symptoms improved following surgery.

DISCUSSION
The pathophysiology of syringomyelia development is not fully understood. Current prevailing theories suggest that increased pulse pressure in the subarachnoid space forces cerebrospinal fluid (CSF) through the spinal cord into the syrinx (Greitz *et al.*, 2013; Muthukumar *et al.*, 2012; Di Lorenzo *et al.*, 2005).
The common causes of syringomyelia are
1. Arnold Chiari malformation Type 1
2. Other congenital malformations (eg, Klippel-Feil syndrome, and tethered spinal cord) (See "Evaluation of neck stiffness in children", section on 'Congenital'.)

3. Postinfectious
4. Postinflammatory (transverse myelitis and multiple sclerosis)
5. Spinal neoplasms (especially ependymoma and hemangioblastoma) (See "Spinal cord tumors".)
6. Posttraumatic (See "Chronic complications of spinal cord injury", section on 'Syringomyelia'.)

This patient's MRI also showed features of Syringomyelia with Arnold chiari malformation

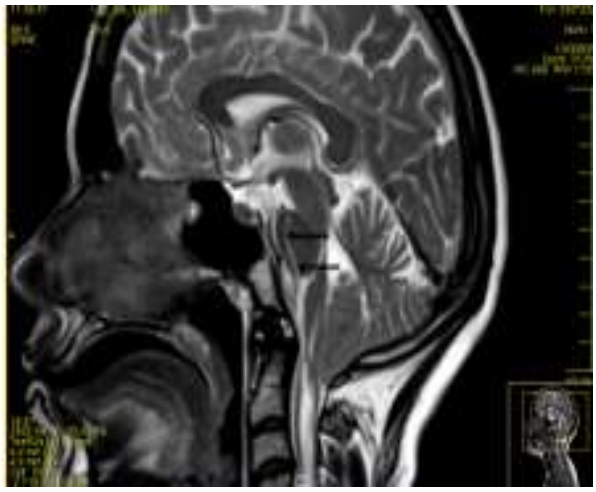

Figure 1: Sagittal T2 image

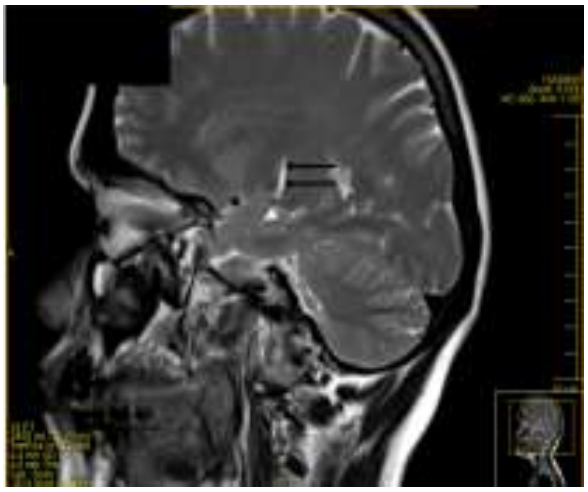

Figure 2: Sagittal T2 at internal capsule

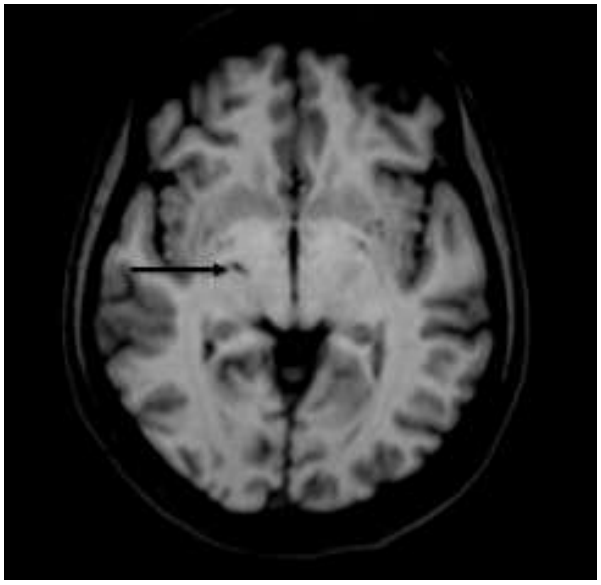

Figure 3: Axial T1 at internal capsule

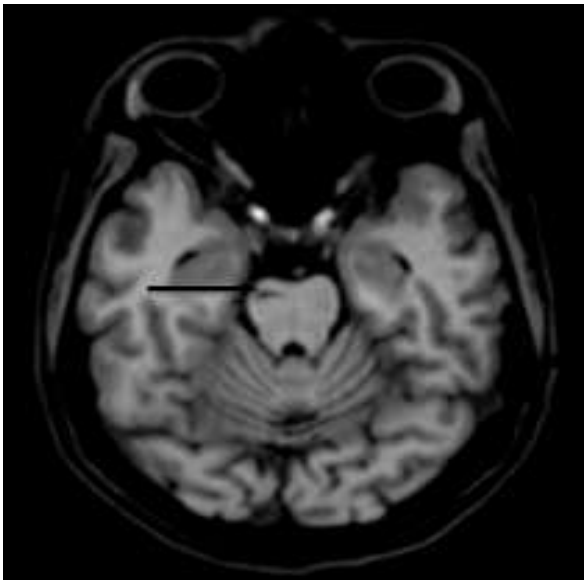

Figure 3: Axial T1 at pons

Figure 1-4: MRI of the patient : A linear tract is extending from the right internal capsule to the cerebral peducle which appears hyperintense in T2 sequences and hypointense in T1 and FLAIR sequences.

The typical clinical finding in patients with syrinx is the presence of dissociated sensory loss (loss of pain and temperature due to the involvement of the decussating spinothalamic fibres) which was found in this patient. Painless ulcers and Charcot's joint may be present (Nacir *et al.*, 2010). Syringomyelia may also present with lower motor signs due to involvement of the anterior horn cells. Autonomic signs are also known to occur. As syringomyelia progresses, reduction in sensation and increased spasticity may be seen (Caroll *et al.*, 2005). Progression is usually slow in most patients, with the clinical picture remaining

static for many years (Mariani *et al.,* 1991). When the syrinx extends into the medulla the syndrome is characterized by dysphagia, nystagmus, pharyngeal and palatal weakness, asymmetric weakness and atrophy of the tongue, and sensory loss involving primarily pain and temperature senses in the distribution of the trigeminal nerve. This extension into medulla is called as syringobulbia (Seki *et al.,* 2003; Tubbs *et al.,* 2009; Viswanatha *et al.,* 2009; Morgan *et al.,* 1992). It is a well-known entity.

In this patient, though all the lesions could be explained by a syringobulbia/myelia the loss of pain and temperature sense on the left side of face with contralateral (right side) loss of pain and temperature can only be explained by the involvement of Right thalamus. It is likely that the involvement of thalamus led to the loss of pain and temperature on the same side of the body.

This case was diagnosed to have extension of the syrinx cavity by MRI upto the thalamus. The involvement of the there have been only about ten case reports of such extension above the brainstem (Lee *et al.,* 2001). Such extension is also termed as Syringocephalus or syringoencephalomyelia (Berry *et al.,* 1981). This case is being presented for its rarity and MRI finding.

REFERENCES

Berry RG, Chambers RA and Lublin FD (1981). Syringoencephalomyelia (syringocephalus). *Journal of Neuropathology & Experimental Neurology* **40**(6) 633–44.

Brickell KL, Anderson NE, Charleston AJ, Hope JKA, Bok APL and Barber PA (2006). Ethnic differences in syringomyelia in New Zealand. *Journal of Neurology, Neurosurgery and Psychiatry* **77**(8) 989-91.

Carroll AM and Brackenridge P (2005). Post-traumatic syringomyelia: a review of the cases presenting in a regional spinal injuries unit in the north east of England over a 5-year period. *Spine* **30**(10) 1206–10.

Di Lorenzo N and Cacciola F (2005). Adult syringomielia. Classification, pathogenesis and therapeutic approaches. *Journal of Neurosurgical Sciences* **49**(3) 65–72.

Greitz D (2006). Unraveling the riddle of syringomyelia. *Neurosurgical Review* **29**(4) 251–64.

Lee JH, Chung CK and Kim HJ (2001). Cerebral dissection from syringomyelia demonstrated using cine magnetic resonance imaging- Case report. *Journal of Neurosurgery* **94**(2) 318–21.

Mariani C, Cislaghi MG, Barbieri S, Filizzolo F, Di Palma F and Farina E (1991). The natural history and results of surgery in 50 cases of syringomyelia. *Journal of Neurology* **238**(8) 433–8.

Milhorat TH (2000). Classification of syringomyelia : Neurosurgical Focus. *Journal of Neurosurgery Publishing Group* **8**(3) 1–6.

Morgan D and Williams B (1992). Syringobulbia: a surgical appraisal. *Journal of Neurology, Neurosurgery and Psychiatry* **55**(12) 1132–41.

Muthukumar N (2012). Syringomyelia as a presenting feature of shunt dysfunction: Implications for the pathogenesis of syringomyelia. *Journal of Craniovertebral Junction and Spine* **3**(1) 26–31.

Nacir B, Arslan Cebeci S, Cetinkaya E, Karagoz A and Erdem HR (2010). Neuropathic arthropathy progressing with multiple joint involvement in the upper extremity due to syringomyelia and type I Arnold-Chiari malformation. *Rheumatology International* **30**(7) 979–83.

Romero-Pinel L, Fernández S, Torres A, Majós C, Ferran E and Martínez-Yélamos S (2006). Syringomyelia extending to the corona radiata. *Journal of Neurology* **253**(6) 817–8.

Seki T, Hida K, Lee J and Iwasaki Y (2004). Hiccups attributable to syringobulbia and/or syringomyelia associated with a Chiari I malformation: case report. *Neurosurgery* **54**(1) 224–6.

Tubbs RS, Bailey M, Barrow WC, Loukas M, Shoja MM and Oakes WJ (2009). Morphometric analysis of the craniocervical juncture in children with Chiari I malformation and concomitant syringobulbia. *Child's Nervous System : ChNS : official journal of the International Society for Pediatric Neurosurgery* **25**(6) 689–92.

Viswanatha B (**2009)**. Syringomyelia with syringobulbia presenting as vocal fold paralysis. *Ear, Nose & Throat Journal* **88**(7) E20.