# Intramedullary Pilocytic Astrocytoma Presenting with Holocord Syringomyelia: A Pathophysiological Enigma

Ashutosh Kumar[1] Kavindra Singh[2] Lily Pal[3]

[1] Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India
[2] Department of Neurosurgery, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India
[3] Department of Pathology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

**Address for correspondence** Kavindra Singh, MCh, Department of Neurosurgery, All India Institute of Medical Sciences, Rishikesh 249203, Uttarakhand, India (e-mail: singh.kavindra8@gmail.com).

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Syringomyelia is a condition characterized by cystic cavitation of the spinal cord, having a fluid composition similar to that of cerebrospinal fluid. Holocord syrinx is a terminology used when it occupies the entire spinal cord. Historically, it was seen in patients with Chiari malformation or with spinal hemangioblastomas. Its association with other intramedullary tumors like ependymomas has been reported only in very few cases and has never been reported with pilocytic astrocytomas. Various hypotheses have been proposed regarding the underlying basis of syrinx formation in patients with intramedullary spinal cord tumors. Here we present the case of a patient with a lower dorsal intramedullary tumor in association with a holocord syrinx. This case also highlights the importance of thorough spinal cord imaging in patients harboring a holocord syrinx to look for neoplastic lesions, which may be missed if the lesion is small or noncontrast enhancing.

A 46-year-old man presented with complaints of backache and spastic paraparesis for 1.5 years. He also complained of difficulty in passing urine along with a history of constipation for 6 months. He had undergone a lumboperitoneal shunt at an outside institution 9 months back, without any improvement in his symptomatology. Imaging revealed syrinx spanning the entire spinal cord, with a heterogeneously enhancing intramedullary tumor extending from D9 to D11 (►**Fig. 1**). Intraoperatively the tumor was pale-yellow, firm, mildly vascular, and solid-cystic with areas of calcification, and the entire solid component of the tumor was removed. Histopathological examination showed tumor cells with pleomorphic, oval to spindle nuclei along with eosinophilic cytoplasm (►**Fig. 2**). A diagnosis of pilocytic astrocytoma was made. The patient's postoperative course was uneventful with a slight improvement in power at 6 months of follow-up.

The association of syringomyelia in patients with spinal cord tumors was first reported in 1875 by Simon et al. A holocord syrinx is a rare condition seen in patients with either Chiari malformation[1] or intramedullary spinal cord tumors.[1] Classically hemangioblastomas[2] (HGBs) are known to have an extensive syrinx, even with small tumors and rarely ependymomas[3] have also been associated with a holocord syrinx. Two mechanisms have been postulated for this phenomenon: first, the transudation of fluid from pathological tumor vessels, especially in vascular tumors like HGB, and the second is due to obstruction to cerebrospinal fluid flow and extracellular fluid flow by the tumor.[4] It is hypothesized that subarachnoid and extracellular spaces are single fluid compartments with interrelated fluid flow. A block in one of these spaces will be reflected as increased flow in another compartment. As per the second hypothesis, the higher the tumor level, the greater the chances of the syrinx. Why some patients harbor a holocord syrinx even with a small tumor at a lower level, as in this case, remains an enigma.

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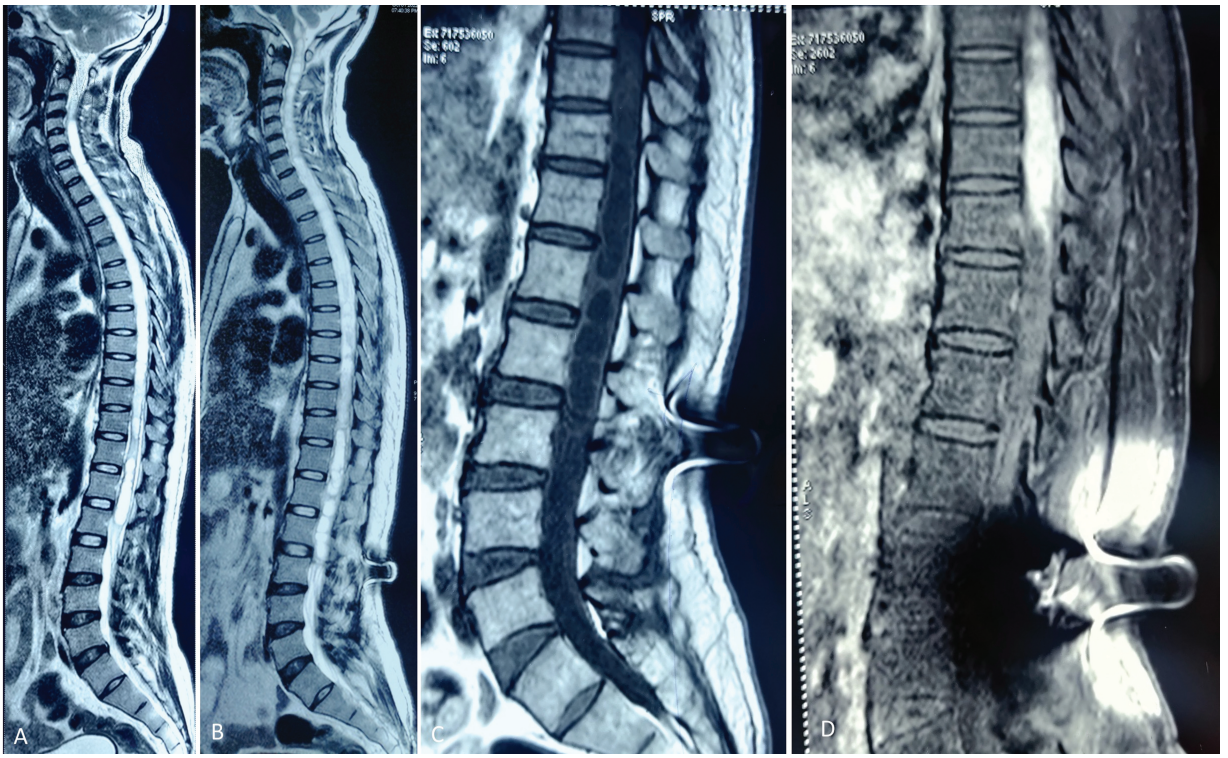


**Fig. 1** (**A**) Magnetic resonance imaging (MRI) T2-weighted sagittal sequence (before lumboperitoneal shunt) showing hyperintense signal throughout the cord with loculations at some places suggestive of syrinx. (**B**) T2 weighted sagittal sequence after LP shunt showing persistent holocord syrinx with the shunt seen at the level of the third lumbar vertebra. (**C**) T1-weighted sagittal sequence with hypointense signal with loculations extending up to L3, D- T1 + contrast sagittal sequence showing a heterogeneously contrast-enhancing intramedullary tumor extending from the 9th to the 11th dorsal vertebral level.

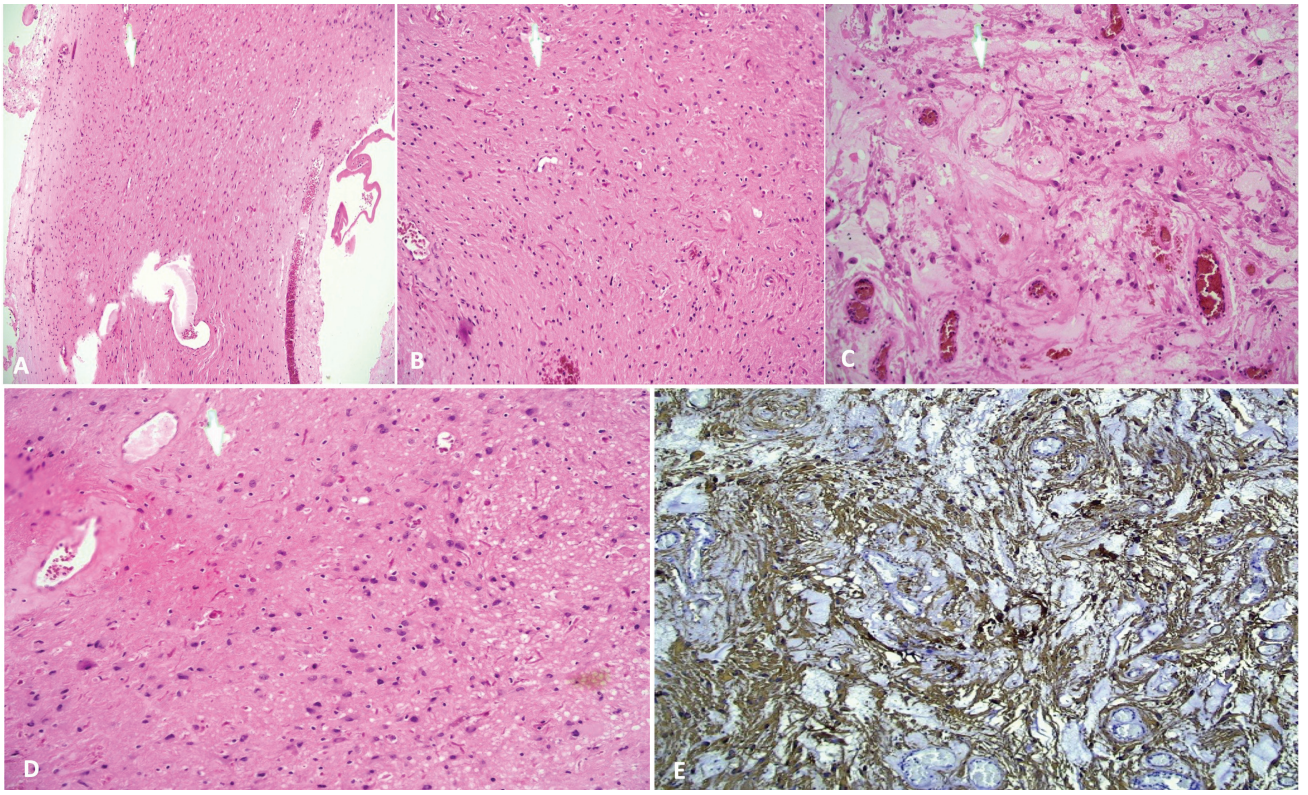


**Fig. 2** (**A**) Low-power view of a sparsely cellular tumor (10x). (**B, D**) Lower magnification of pilocytic astrocytoma showing piloid cells with mildly pleomorphic astrocytes, occasional ganglionlike cells and many Rosenthal fibers (20x). (**C**) Many hyalinized vessels and hemorrhage (10x). (**E**) Astrocytes are glial fibrillary acidic protein (GFAP) positive (10x).

 

**Conflict of Interest**
None declared.

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