A dorsal intramedullary astrocytoma presenting as syringobulbia: A case report.

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Citation

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Abstract

A 26 years old male presented with complaints of dysphagia and hoarseness of voice with poor gag reflex. Magnetic resonance imaging (MRI) of the spine and brain showed medium sized, ovoid shaped, intensely enhancing intramedullary lesion at D5-7 associated with extensive syringomyelia from cervicomedullry junction to D11 and syringobulbia. He underwent D5-8 laminectomy with near total excision of the lesion. The histopathology revealed astrocytoma (WHO Grade-1). The patient improved completely after surgery with resolution of syrinx. In the present case, the unusual clinical presentation, diagnostic challenges, and treatment dilemmas have been discussed.

KEY MESSAGE

Intramedullary astrocytoma presenting with features of syringobulbia is extremely rare. Nevertheless, it must be considered when a patient presents with features of bulbar palsy since its total resection can be achieved with remarkable clinical outcome.

INTRODUCTION

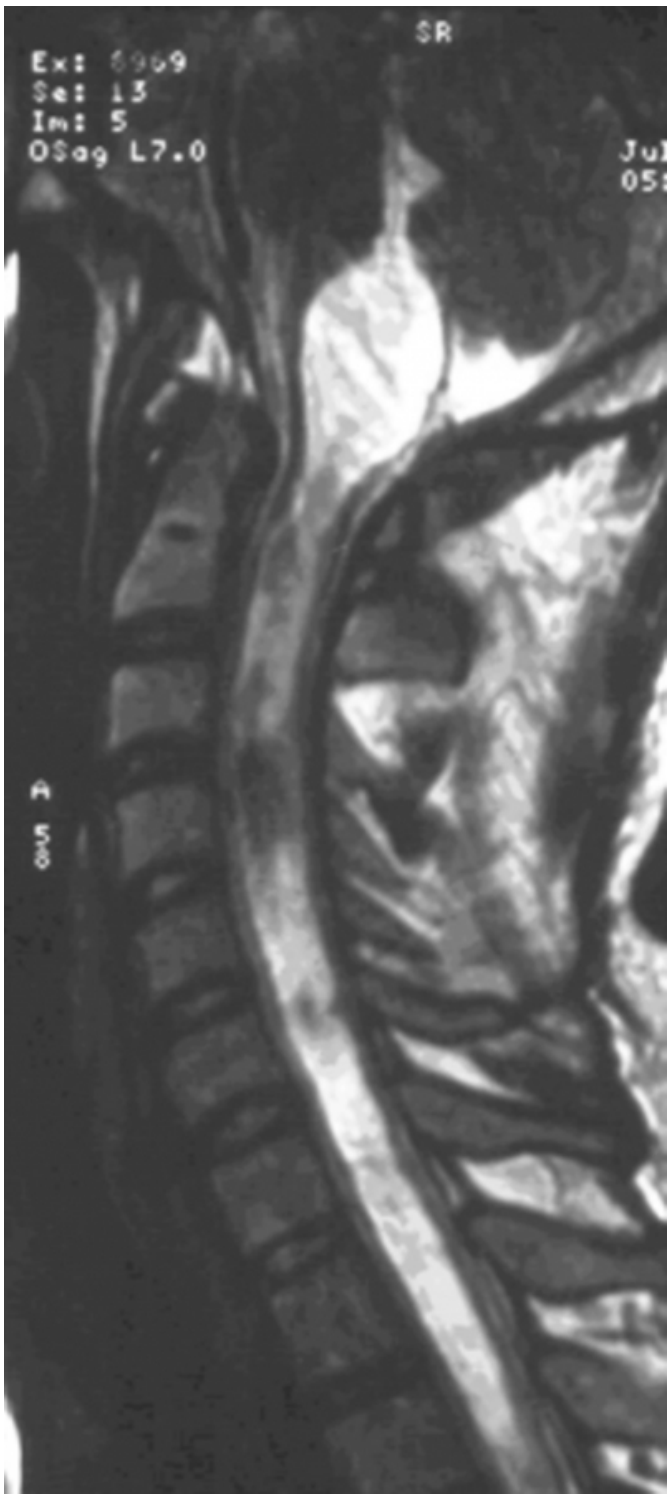
Intramedullary astrocytoma is the most common spinal cord tumor in children & second most common, next only to ependymoma, in adults [1]. Intramedullary astrocytoma is a heterogenous group with respect to natural history, gross characteristics, histological and biological features. We report a case of dorsal intramedullary astrocytoma that presented with bulbar palsy. The diagnostic challenges and the treatment dilemma have been discussed.

CASE HISTORY

A 26 yrs old businessman presented with complaints of difficulty in swallowing for 10 day and recurrent vomiting for 5 days. For these symptoms he underwent extensive investigations including upper gastrointestinal endoscopy, laryngoscopy, barium swallow and Computed tomography (CTScan) of the abdomen. All these studies were reported to be normal. By the time he presented to us, his dysphagia had further worsened and he had also developed hoarseness of voice. On examination his speech had nasal twang with depressed pharyngeal movement. However the palatal movement was normal. Pinprick and temperature sensation over C3-7 dermatome (cape like distribution) and proprioception in both lower limbs were diminished. There were no pyramidal or cerebellar signs. Magnetic resonance imaging (MRI) of the brain showed syringobulbia (Figure 1).

Figure 1

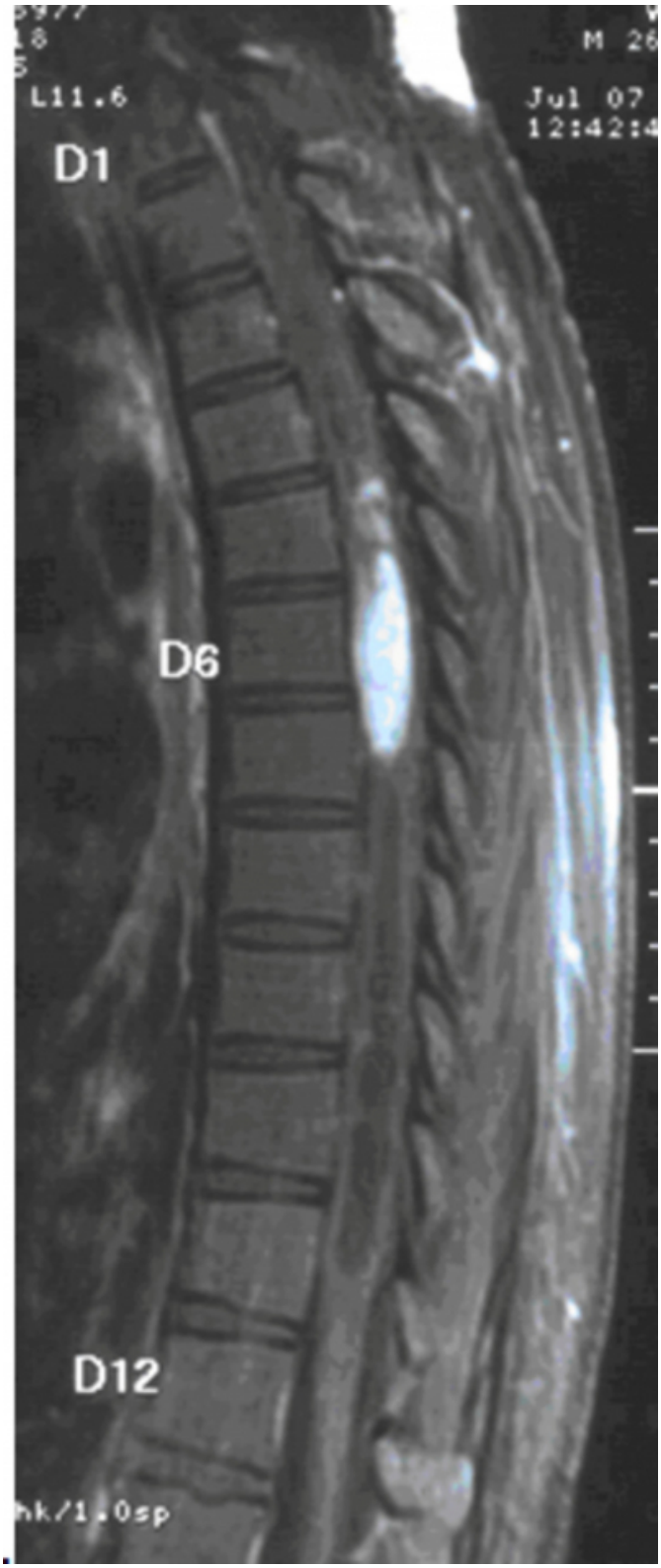
Figure 1: T2 WI MRI showing syringobulbia with cervicodorsal syrinx



The caudal extent of the syringobulbia was further investigated with MRI of the spine which showed medium sized, ovoid shaped, intensely enhancing intramedullary lesion at D5-7 level associated with the rostral syringobulbia and caudally extending syringohydromyelia from cervicomedullary junction to D11 (Figure 2).

Figure 2

Figure 2: Post contrast MRI showing enhancing lesion at D6 level, with caudal extension of the syrinx to D11.

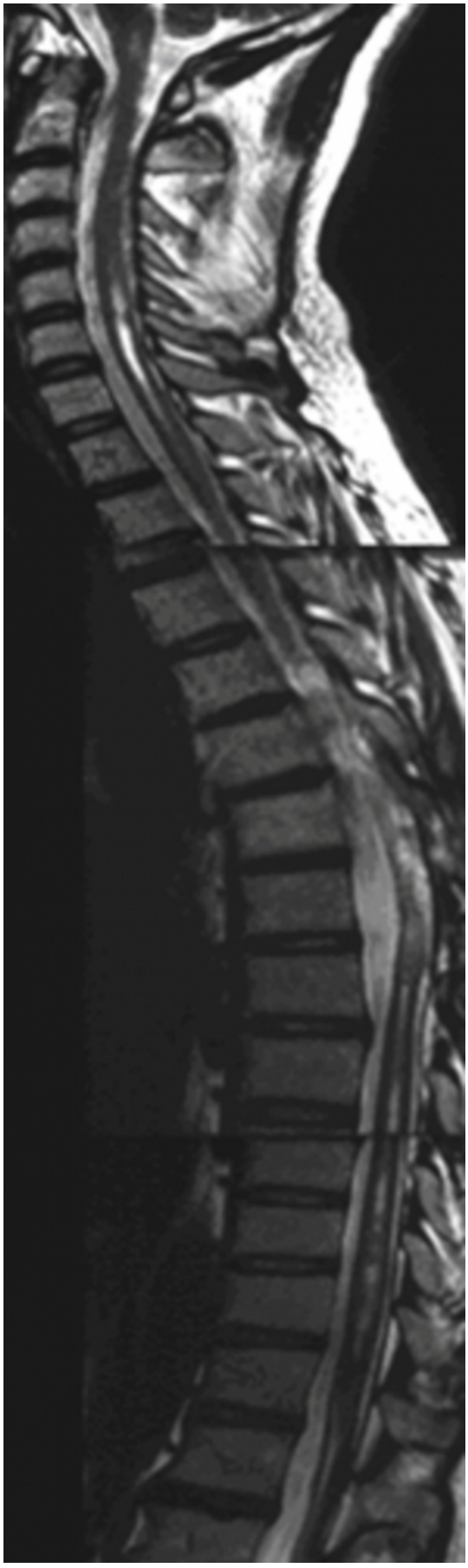


He underwent D5-8 laminectomy and gross total excision of

the intramedullary tumor. Post-operatively his swallowing improved within a week. The histopathology revealed an intramedullary astrocytoma (WHO grade – 1). At 2months follow up, he had complete recovery and MRI showed complete tumor excision with resolution of syrinx (Figure 3). Patient is now free of symptoms 2 years after surgery.

Figure 3

Figure 3: Follow up MRI 2 months after surgery showing complete resolution of the syrinx and complete excision of the tumor.



DISCUSSION

About 3% of central nervous system astrocytoma arises within the spinal cord [2], of which, 60% occur in the cervical and cervicothoracic region [3]. Thoracic, lumbosacral, or conus medullaris are less common locations. Intramedullary astrocytoma presents with variable clinical feature. Early symptoms are usually nonspecific and slowly progressive. Symptoms are often present 3 to 4 years before the diagnosis [45]. Pain and weakness are the most frequent presenting symptoms [356]. Pain typically localizes to the level of the tumor and is rarely radicular [1]. Cervical lesion produces upper extremity symptoms while thoracic tumors produce spasticity and sensory disturbances [1]. Other symptoms include clumsiness, gait difficulty, loss of pain, temperature, and proprioception, while bowel, bladder, or sexual dysfunction is seen with more advanced disease. In this case, patient presented with symptoms of rapid onset dysphagia, and hoarseness of voice with associated signs of depressed gag reflex, and nasal twang voice suggestive of bulbar palsy. The causes for bulbar palsy are medullary infarction, medullary tumor, syringobulbia, demyelination, motor neuron disease [7]. Gadolinium enhanced Magnetic Resonance Imaging (MRI) of the spine is the investigation of choice for intramedullary astrocytoma [8]. Spinal cord is enlarged and contrast study reveals ill-defined mass due to their irregular margins. Heterogenous uptake and patchy irregular margins are common because of intratumoral cysts or necrosis [8]. The association of intramedullary tumors with syringomyelia is well recognized [3914]. In the retrospective analysis of 100 intramedullary tumors by Madjid Samii, 45% presented with associated syringes. Ependymoma and hemangioblastomas were the most common tumor types to be associated with syringes. Astrocytoma tended to demonstrate syringes less often (20%). A syrinx was more likely to be found above (49%) than below (11%) the tumor level [10]. A syrinx forms predominantly above the intramedullary tumor and, the higher the spinal level, the more likely a syrinx tends to occur [9]. In his series, in 4 patients syringes extended up into the medulla oblongata. Syringobulbia coexisting with syringomyelia is reported in the literature in 16 cases in which the tumor was primarily intramedullary [9]. Of these, three had Von Hippel-Lindau's disease and one had vascular tumor. In the present case, patient had intramedullary astrocytoma at D5-7 level with associated syringomyelia and syringobulbia. Interesting feature about this patient is that he presented with the symptoms of syringobulbia. In literature there are few cases described where cervicomedullary astrocytoma is associated with syringobulbia [910]. To the best of our knowledge, there has not been any case report in the literature of intramedullary astrocytoma of dorsal spine presenting with clinical symptoms and signs of syringobulbia.

Although some benign astrocytoma are well circumscribed and allows gross total resection, most of them exhibit variable infiltration into the surrounding spinal cord. A definitive correlation between the extent of resection and tumor control has not been established [3610]. Therefore, preservation of neurological function, rather than complete tumor resection should be the treatment priority [11]. Tumor removal should be limited to tissue that is clearly distinguishable from the surrounding spinal cord [1]. The decision for surgical intervention in patients with a slowly progressive minor motor or sensory deficit is difficult, particularly if imaging studies suggests an infiltrating astrocytoma, which may not be removed without significant risk of neurological deficit [12]. As in our patient, we were in a dilemma whether to excise the tumor in neurologically preserved patient or to drain the syrinx cranially to alleviate the symptoms of syringobulbia. If a syrinx accompanies the tumor, this should be interpreted as a favorable prognostic sign, because it indicates a displacing rather than a infiltrating tumor and thus suggests resectibility of mass. It is sufficient to operate on the solid portion only. The accompanying syrinx will decrease automatically if the tumor has been removed [10]. Outcome of surgical treatment is directly related to the patient's preoperative status, the location of the tumor, and the presence of spinal cord atrophy and arachnoid's scarring [351013]. In general, intramedullary astrocytomas are low grade, slowly growing neoplasm. Long recurrence free survivals are common, particularly in younger patients [6].

CONCLUSION

Intramedullary astrocytoma presenting with features of syringobulbia is extremely rare. Nevertheless, it must be considered when a patient presents with features of bulbar palsy since its total resection can be achieved with remarkable clinical outcome.

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