

Basilar invagination associated with brainstem tumor opposite the odontoid process

ABSTRACT

An 8-year-old girl presented with the symptom of nasal regurgitation and hoarseness of voice. Investigations revealed severe basilar invagination and a large intra-axial pons-medulla brainstem tumor adjoining the tip of the odontoid process. The child underwent lateral mass plate and screw atlantoaxial fixation and attempted craniovertebral junction realignment. Biopsy of the brainstem tumor was done during the same surgical procedure. Histology of the brainstem tumor revealed it to be a pilocytic astrocytoma. Simultaneous presence of brainstem tumor and basilar invagination is a rare clinical association and a complex therapeutic challenge. The possible cause of genesis of brainstem tumor in an unusual location is speculated.

Keywords: Atlantoaxial fixation, basilar invagination, brainstem glioma

INTRODUCTION

Intraaxial brainstem tumors and basilar invagination are discrete clinical entities. A case is reported wherein both these clinical entities were present together and in close proximity. The clinical therapeutic issues posed in the case are elaborated. To the best of our knowledge, such a combination of entities has never been reported earlier.

CASE REPORT

An 8-year-old female presented with the gradual progressive symptoms of weakness and stiffness of all four limbs, difficulty in swallowing, nasal regurgitation, and hoarseness of voice for a period of approximately 1 year. She complained of disabling pain in the nape of the neck during the same period. On sleeping supine, she had severe giddiness and preferred to lie in the prone position or on her sides. Neurological examination revealed an impaired gag reflex and spasticity in all four limbs and hyperreflexia. She moved all her limbs but was bed-bound. Computed tomography (CT) scan showed Group A basilar invagination, platybasia, assimilation of Atlas and atlantoaxial dislocation, and severe compression of the brainstem. Apart from basilar

invagination, magnetic resonance imaging (MRI) of the brain showed a large intraaxial lesion in the medulla abutting the tip of the odontoid process [Figure 1]. The tumor was hypointense on T1-weighted images, hyperintense on T2-weighted images, and marginally enhanced on contrast administration. As she had difficulty in sleeping supine, the MRI had to be performed with the patient in the prone position. The patient was operated in the prone position


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Submitted: 14-May-23
Published: 13-Jun-23

Accepted: 15-May-23

Access this article online	
Website: www.jcvjs.com	Quick Response Code 
DOI: 10.4103/jcvjs.jcvjs_54_23	

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How to cite this article: Shah A, Vutha R, Goel A. Basilar invagination associated with brainstem tumor opposite the odontoid process. J Craniovert Jun Spine 2023;14:205-7.

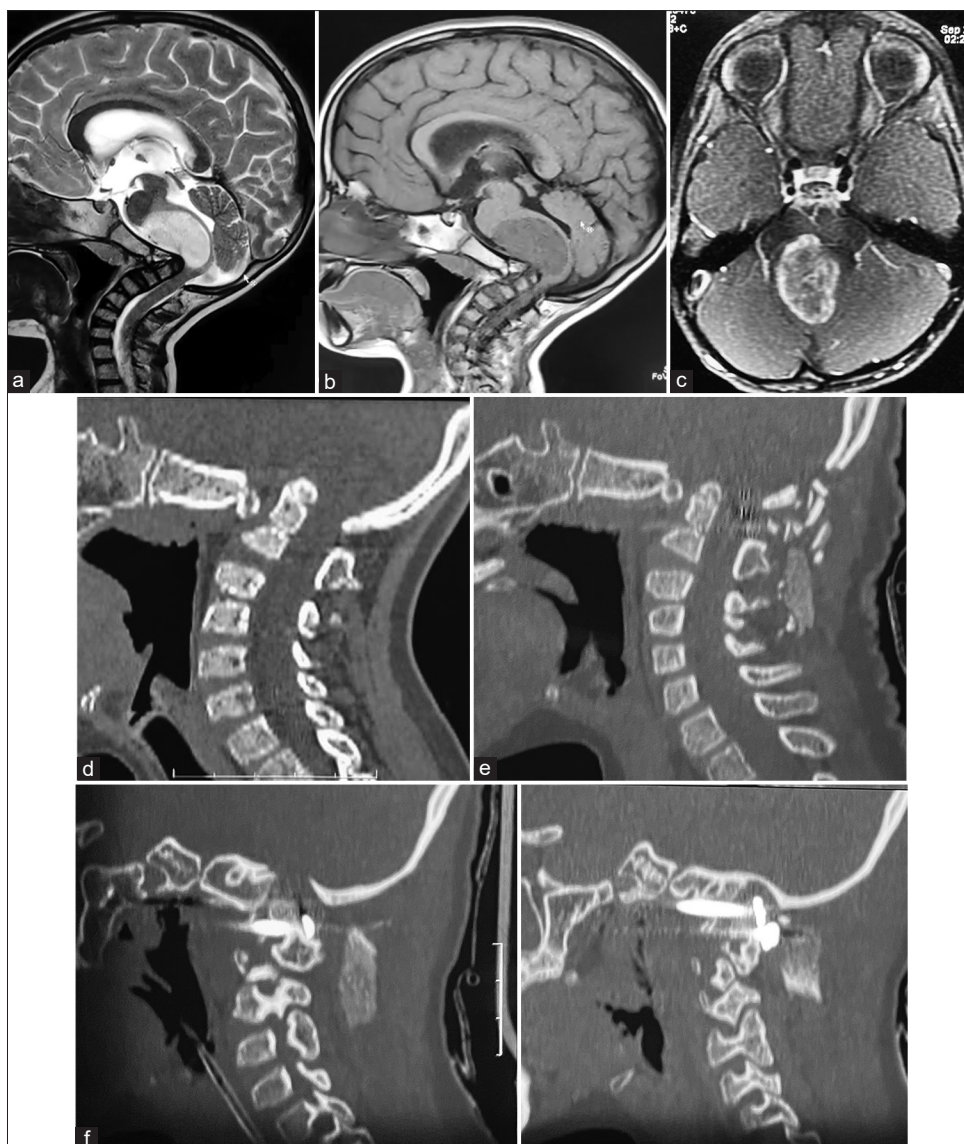


Figure 1: (a) T2-weighted sagittal MRI showing basilar invagination and a large brainstem hyperintense tumor. (b) T1-MRI showing the lesion to be relatively hypointense. (c) Contrast-enhanced axial image of MRI showing contrast enhancement. (d) Sagittal image of CT scan showing basilar invagination. Partial assimilation of atlas is seen. (e) Postoperative CT scan showing reduction of basilar invagination. (f) Postoperative CT scan showing the implant and bone placed for fusion. MRI – Magnetic resonance imaging, CT – Computed tomography

under cervical traction. Lateral mass plate and screw fixation was done as described by the senior author.^[1,2] The essential steps of the surgery have been described. The craniovertebral junction was exposed and the C1-C2 joints were opened with considerable difficulty. Bone graft harvested from the iliac crest was inserted into the C1-C2 joints and a C1 lateral mass and C2 pedicle screw fixation were performed on both sides. A midline suboccipital craniectomy was then performed. On retracting the cerebellum, the bulge of the medulla was exposed. The tumor was visualized and an incision was taken over the site of maximum medullary bulge. The tumor was soft in consistency. Considering the location of the tumor, its large size and infiltrative nature, only a biopsy of the tumor was performed. The patient improved in her

symptoms of nasal regurgitation and hoarseness of voice. She was able to sleep supine. Postoperative CT scan showed reduction of the basilar invagination and good re-alignment of the craniovertebral junction [Figure 1]. Histology of the brainstem tumor revealed it to be a pilocytic astrocytoma. Six days after the surgery the child started developing spells of apnea and this was followed by loss of consciousness. She was electively intubated and observed. On the ventilator she regained consciousness and had good movement of all her limbs with good respiratory effort. She was weaned off and extubated after 5 days and remained well for 2 days after which she became unconscious after an apneic spell and had to be re-intubated. The cycle was repeated one more time. But ultimately the patient became ventilator

dependent. The parents did not wish for tracheostomy or any further treatment of the patient and ultimately the patient succumbed after 2 months of surgery.

DISCUSSION

Basilar invagination Type 1 as identified in the presented case has been traditionally identified as herniation of spine into the brain.^[3,4] The odontoid process impinges or compresses the brainstem and the process results in a variety of neural and musculoskeletal alterations. For the first time in the literature, we identified that basilar invagination is a consequence of chronic atlantoaxial instability and atlantoaxial stabilization is the treatment.^[4] For basilar invagination type 1, we introduced the concept of craniovertebral junction realignment.^[4] The surgical technique involves opening up of the atlantoaxial joint, denuding of the articular cartilage, and distraction of the facets with bone graft or by intraarticular spacers.^[1,4,5] Over the years, we have realized that in basilar invagination, atlantoaxial instability is the issue and atlantoaxial stabilization is the treatment. Instead of using metal implants, we currently prefer the use of bone graft for the purpose of distraction and for fusion.^[6-8]

Basilar invagination in the presented case was associated with brainstem tumor that was in surprisingly close proximity to the tip of the odontoid process. The presence of both basilar invagination and of brainstem tumor presented a therapeutic challenge. The pathogenesis of brainstem glioma in such a location raises some curious issues. It can be speculated that chronically unstable atlantoaxial joint and relentless pressure and/or injury over the brainstem by the odontoid process could be the nodal pathogenetic point of the development of brainstem tumor. Such chronic injury has been implicated as a cause of genesis of cancers in other body location, particularly oral cancers.

Despite satisfactory craniovertebral junction stabilization, our patient could not survive in the postoperative period and an opportunity to evaluate and understand the “special” nature of brainstem tumor was lost.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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