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Case Report

Subpial Cervical Subependymoma: Report of an Unusual Tumor with Review of Literature

Abstract

Subependymoma is rare benign neoplasm (World Health Organization Grade I) usually found in the 4th ventricle and lateral ventricles. They were first described by Boykin as a separate entity in 1954. Subependymoma constitutes only 1%–2% of spinal ependymal tumors. Majority of the spinal subependymoma is intramedullary, with a rare few reported in the extramedullary plane. Clinicoradiologically, subependymoma often mimic more frequent, aggressive tumors of the spine (astrocytoma and ependymoma) which makes them difficult to differentiate. In fact, the diagnosis of subependymoma comes as a histopathological surprise. Maximal safe resection holds the key to good postoperative outcome with a very limited role of adjuvant therapy. Complete excision of the tumor, though desirable, is not feasible in all cases. Owing to their rarity and lack of characteristic clinicoradiological features, there is limited information currently available regarding their preoperative diagnosis and "optimal" management strategy. In this case report, we are discussing a case of eccentric subpial cervical subependymoma discussing important differentiating radiological features, and surgical nuances with an attempt to define "optimal" management strategy.

Keywords: *Bamboo leaf sign, cervical, subependymoma, subpial*

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Introduction

Subependymoma constitutes about 1%–2% of spinal ependymal tumors.[1,2] Even though they may occur anywhere along the spinal cord, C1–C2 is the most frequent location (24%).[2-5]

Subependymoma often mimic aggressive intramedullary tumors on radiology and frequently present with pain, sensory-motor deficits, bowel, and bladder dysfunction like them. Due to their benign biological behavior, complete surgical excision is usually considered curative. Owing to a highly controversial role of adjuvant therapy, no consensus has been reached on "optimal" management of these cases.

We report a case of eccentric subpial subependymoma discussing important differentiating radiological features and surgical nuances with an attempt to define "optimal" management strategy.

Case Report

A 36-year-old male presented with pain and progressive paresthesia in right-sided limbs for 3 years without sphincter dysfunction. Examination revealed spastic weakness (Medical Research Council Grade 4/5) in both upper limbs and right lower limb with modified McCormick Grade 2 disability. He also had 30%–40% sensory loss to touch and pain below C5 with impaired posterior column sensations. Magnetic resonance imaging (MRI) showed a well-defined mass lesion extending from cervicomedullary junction to C5, causing expansion of the cord without syrinx formation or tumor cysts. Tumor was eccentrically placed, anterolaterally on right side pushing the spinal cord toward left. It was T1 hypo to isointense, T2 hyperintense without significant contrast enhancement [Figure 1]. Common intramedullary lesions such as astrocytoma and ependymoma were considered among the differential diagnoses.

C1–C5 laminectomy for tumor excision was done. Intraoperatively, tumor was greyish, soft, moderately vascular lesion extending along subpial plane on the right side, without distinct planes at either end. Upper half of the tumor was found anterolateral to the cord on right side, and lower part was seen extending ventral to the cord. No myelotomy was required for tumor excision.

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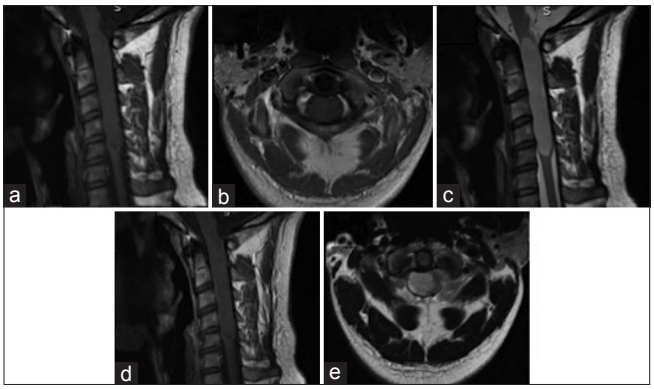

Figure 1: The typical radiological findings in spinal subependymoma. A well-defined eccentrically placed, intramedullary lesion can be seen extending from cervicomedullary junction to C5 causing distinctive steep dilation of spinal cord: Bamboo leaf sign. Lesion is iso to hypointense on T1-weighted images (a and b) and hyperintense on T2-weighted image. There is no evidence of perilesional edema or syrinx formation (c). No significant enhancement is seen on T1-weighted postcontrast image (d and e)

Histopathologically, tumor cells with mildly enlarged anisomorphic nuclei were seen clustered in the acellular fibrillary matrix. Mitotic activity and necrosis were absent. These features were consistent with subependymoma. Immunohistochemistry showed weak tumor cell positivity for glial fibrillary acidic protein (GFAP) and S-100, and negative for neuron-specific enolase and epithelial membrane antigen (EMA) with low Ki-67 index (1%) [Figure 2].

Postoperatively, motor power in all four limbs worsened by one grade compared to the baseline. We planned for radiological follow-up without any adjuvant therapy. At 2-month follow-up, motor power and spasticity improved with functional recovery.

Discussion

Nearly 50% of subependymoma are identified incidentally. On the other hand, symptomatic cases occur frequently in 5th–6th decade and rarely affect children.[6-8] Symptoms may exist months to several years before diagnosis are made, reflecting their indolent behavior. However, they may mimic more aggressive intramedullary tumors clinically. Pain and sensory deficits are the most common initial presentation with the risk of compressive myelopathy and loss of sphincter control later. Therefore, early surgery may preclude a significant morbidity.

Histopathogenesis of subependymoma still remains elusive, with few authors even reporting them as a variant of ependymoma based on electron microscopic study.[9] Whereas, others believe them to be a separate entity arising from various cells such as subependymal glial cells, subependymal cell plate or as a result of some developmental defect.[10-12] Krishnan *et al*. proposed their origin from subpial white matter progenitor cells, which later descend to eccentric, subpial location.[3]

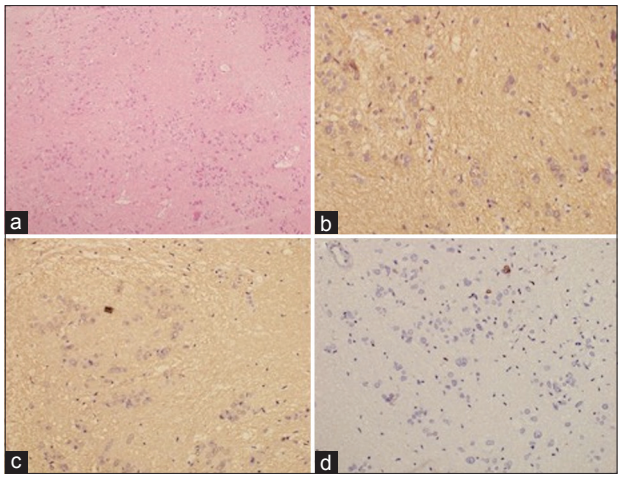

Figure 2: (a) Microphotograph showing tumor composed of loose aggregates of cells with intervening hypocellular fibrillary matrix (H and E, ×10), (b) Immunohistochemistry showing expression of glial fibrillary acidic protein (IHC, ×20), (c) Immunohistochemistry shows expression of S-100 (IHC, ×20), and (d) Ki-67 proliferation index showing nuclear positivity in <1% cells (IHC, ×20)

In contrast with ependymoma, features such as mitotic activity, ependymal rosettes, or perivascular pseudorosettes are rarely found. They exhibit the GFAP and S-100 positivity similar to astrocytic tumors but may show dot-like pattern for EMA due to poor formation of ependymal-type rosettes.[6] Mitotic index is frequently low (<1%).

Radiologically, it is difficult to establish a definitive diagnosis due to lack of characteristic findings and a limited number of reported cases. On MRI, our case showed an eccentrically placed tumor causing distinctive steep dilation of the cervical cord. It was T1 isointense, T2 hyperintense, nonenhancing lesion without peritumoral edema. Such dilation occurs as a result of tumor growth in the subpial plane and has been termed "Bamboo leaf sign." It may help to differentiate them from ependymoma or astrocytoma which cause gradual fusiform enlargement of the cord.[13] Therefore, a high index of suspicion for "subependymoma" should be considered in "ependymoma" which have little or no edema with minimal or no contrast enhancement. However, tumor cysts and syringomyelia are very rarely associated with them.

A gross total resection is considered curative without requiring adjuvant therapy. Intraoperative features such as lobulated shape, minimal vascularity, eccentric subpial location, and distinct anatomical planes from normal cord facilitate the dissection. Sometimes, total excision may still be difficult to achieve due to local infiltration leading to neurological deficits. Therefore, it becomes highly pertinent to define an "optimal" treatment in managing them.

In our case, we were unable to achieve total excision for the following two reasons. First, there was loss of plane between tumor and cord parenchyma at cervicomedullary junction. Second, tumor was extending ventral to the

cord at its lower end with the high risk of cord traction on attempting its removal. A minimal part of tumor was left behind at these locations to prevent worsening or development of new deficits. Therefore, such a safe surgical approach may be considered "optimal."

Transient weakness in immediate postoperative period similar to our case has been reported earlier in nearly 60% of patients despite a "safe" surgical course; nonetheless, most (76%) of them improve with time.[14] A study also suggests high incidence of poor outcome owing to the cervicothoracic location of tumor, poor intramedullary microcirculation, and postoperative kyphotic deformity.[15]

Surgical excision forms the cornerstone of management in cervical subependymoma and also preferred by most surgeons over irradiation in case of recurrence/regrowth of tumor. Currently, the role of adjuvant radiotherapy remains controversial. Although more experience and studies with longer follow-up would be required to gain further evidence.

Conclusion

Cervical subependymoma is an uncommon, benign tumor which lacks characteristic clinicoradiological findings, and often mimic, frequently occurring aggressive tumors. Complete tumor excision, though desirable, is not feasible always. Maximal safe resection holds the key to good postoperative outcome with the limited role of adjuvant therapy.

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.

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Conflicts of interest

There are no conflicts of interest.

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