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Case Report

Intra Venricular Adult Pilocytic Astrocytoma of 4th Ventricle – A Rare Variant

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Abstract

Background: Pilocytic astrocytoma are low grade, WHO grade 1 tumors arising predominantly in pediatric population. They account for 25% of pediatric and 1.5% of adult brain tumors. Intra ventricular (IV) tumors are a separate entity per se, but IV variants of PAs have been rarely reported in literature. Because of low volume of such cases reported in literature, a definite plan regarding their pre-op diagnosis , and management plan is lacking.

Case Description: Our patient is a 25 year old male, who presented with obstructive hydrocephalus secondary to 4th ventricular lesion. He underwent posterior fossa craniotomy plus excision of this tumour, that turned out as Pilocytic astrocytoma on histopathology.

Conclusion: Intra ventricular Pilocytic astrocytomas are extremely rare. Patients mostly do well following gross total resection of these tumors. but because of rare nature of these lesions, well established guidelines need to be made for their diagnosis , treatment and follow up.

Key words: intra-ventricular adult pilocytic astrocytoma (ivpas); pilocytic astrocytoma (pa); gross total resection (gtr); hydrocephalus; immunohistochemical markers

Introduction

Pilocytic astrocytoma are low grade, WHO grade 1 tumors arising predominantly in pediatric population. Histologically, they classically manifest in biphasic pattern comprising a composition of elongated to plump bipolar astrocytes that are arranged in a fascicular architecture. These are accompanied by foci of compact piloidal tissue (that demonstrate the highly characteristic feature of fine fibrillary (hair-like) processes and a typical abundance of Rosenthal fibers. [1,11]

They represent 25 % of pediatric and around 1.5% of adult brain tumors [3] By immunohistochemistry, the tumor cells are positive for vimentin and glial fibrillary acid protein (GFAP).

These tumors may exhibit alterations in the mitogen-activated protein kinase (MAPK) pathway, most commonly KIAA1549/ BRAF fusion gene. [1,5,11]. However this mutation was absent in PA reported by Hendrix et al [5]. Hence KIAA/BRAF mutation is thought to be present in many but not all cases of IVPAs.

Pediatric PAs are mainly located infratentorially, the commonest site being the cerebellum.[1] In contrast, adult pilocytic tumors are rare and have aggressive clinical course compared to their pediatric counterpart.[10] As per an institutional series on Pilocytic tumors published in 2014, Adult pilocytic astrocytomas, occur at a median age of around 29 (range 18- 72 yrs). The incidence decreases substantially with age, and they are rarely diagnosed in patients over 50 years of age. [1,10]

They are mainly extra cerebellar, commonly located in supratentorial region, with cerebrum, brainstem, optic pathways, hypothalamus, ventricles, cerebellum and spinal cord in decreasing oreder of their occurrence. IVPA, as reported in our case, account for roughly 13 % of total PAs encountered in this series.[10]

Intraventricular brain tumors are a distinct entity per se accounting for less than 10 % of intracranial tumors.[4], with a broad list of differentials including choroid plexus tumors , epndymoma , medulloblastoma, neurocytoma, sub ependymal giant cell astrocytoma etc. [1] Because of the low incidence of PA within the ventricles , only a few cases of IVPAs have been reported in literature, with the most recent published single center experience of 2022, reporting an incidence of 4-15.6% IVPAs among all Pas.[4]

Case Report:

25 year old male with KPS score of 90, presented via out patient department with complaints of Headache and neck pain for past two years. On examination, patient had GCS of 15/15. Positive neurological examination findings were nystagmus in both eyes and positive cerebellar signs (i.e. Dysdidokokinesia, Past pointing and Rombergs). His MRI brain with contrast was done which shows Altered signal intensity peripherally enhancing lesion in posterior fossa likely arising from fourth ventricle causing compression over brainstem and cervical spinal cord, with proximal obstructive hydrocephalus suggestive of fourth ventricular neoplastic lesions. (FIGURE 1 AND 2)

Patient underwent external ventricular drain via Frazier point followed by sub occipital / Posterior fossa craniotomy, c1 and c2 laminectomy, durotomy in Y shaped fashion and inter tonsillar approach for debulking of intraventricular and intra spinal tumor. Aqueductoplasty was done and layered closure of wound was done to avoid CSF leak.

The tumor was found to be protruding through the foramen of megendie on opening the dura., and it was debulked to near total extent using CUSA.

Per-operative finding was a firm, grayish colored, mildly vascular and moderately suckable tumor.

Post operatively, the patient was neurologically sound. His EVD was retained till 3rd post-operative day and was subsequently removed prior to discharge.

He had persistent nystagmus, mild diplopia, on and off headache , on his follow up visits , but those settled on conservative management .

Histopathologic findings were consistent with the diagnosis of Pilocytic astrocytoma. These included glial neoplasm exhibiting biphasic appearance with few areas of compact fibrillar portion having elongated nuclei and bipolar piloid processes. Focal areas shows microcystic spaces with round to oval nuclei, rosenthal fibres and eosinophillic granular bodies. Majority of the areas showed thick walled hyaliniazed vessels and microcalcification, with no significant atypia , mitosis or necrosis seen. GFAP was positive and ki-67 was 1-2 % (FIGURE 3A TO 3D)

DISCUSSION:

We present a case of a middle aged male who presented with 4th ventricle tumor causing obstructive hydrocephalus for which he underwent External Ventricular Drain placement followed by tumor excision, that showed up as pilocytic astrocytoma on histopathology.

IVPAs are rare tumors with a rough incidence of around 5-15 % in literature. They are commonly found in lateral ventricles and are less common in the 4th ventricle.

The ventricular system comprises of the ependymal cells and choroid plexus, and lacks any astrocytic conformation, Hence, all intraventricular astrocytomas are thought to be resulting from brain tissue around, and grow secondarily into the ventricles.[11] Another hypothesis may be differentiation of choroid plexus cells within the ventricles into astrocytes which then turn neoplastic.[4]

Headache and visual deterioration was the most common presenting symptom reported in patients with IVPAs.[4] 4th ventricular lesions may cause early signs and symptoms of obstructive hydrocephalus. A rare presentation of hemi facial spasm has also been reported because of 4th ventricle PA. [8]

Radiologically, the tumor tends to be rounded or irregular in shape, with majority being mixed cystic and solid in composition. Cystic portions are strongly T2 hyperintense/T1 hypointense, while Solid portions are moderately T2 hyperintense/T1hypointense with heterogeneous contrast enhancement. They may show element of hemorrhage or calcification; calcifications usually denote the benign and slow growing nature of these tumors.[2,9,11]. These characteristics on imaging should assist in making a pre-op diffential of Pas.[7]

Though literature has reported clinical presentation and radiologic appearance of IVPAs, but because of the rarity of these tumors in adult population, any consensus regarding the pre-operative diagnosis, post-operative management and clinical or radiological follow up of adult PA and their intra ventricular variants is utterly lacking.[11]

These tumors usually have a good prognosis. However, recurrence may still occur, with variable rate of 13% to 42% reported in literature.[3,10]

The most recent paper on literature search was published in 2021 that reports a case of 26 year old female diagnosed as a case of 4th ventricular pilocytic astrocytoma, who underwent near total excision and had improvement in her symptoms post-operatively. However, on her 10th month clinco-radiological follow up, she was found to have recurrence in the entire ventricular system and sub-arachnoid spaces. To our knowledge, this has been the first reported case of recurrence in IVPAs. Again, because of the un-resectable nature of this recurrence and patient's decision of not continuing further medical care, a clinical strategy of managing these sort of cases is deficient.[6]

Mostly, the management strategies of adult pilocytic astrocytomas can be applicable to IVPAs. However, variability in presenting features and occasional aggressive behaviour of these tumors warrant separate guidelines to be made regarding the management of IVPAs.

Gross total resection for APA, whenever feasible should be the goal of surgery as it provides better chances of disease free survival as compared to subtotal resection. [3] Adjuvant radiotherapy is also an option to increase progression free survival in these patients.[10] Recurrences may be managed via sub-total resection or radiotherapy alone.[3]. Immunohistochemical markers on the biopsied Specimen adds to further information regarding the behaviour of tumor.

Conclusion:

Generally pilocytic tumors are thought to be a benign process with good overall prognosis. IVPAs, however, are rare, and little is known about their natural course. These should therefore be dealt as a separate entity with close clinical and radiological follow up, particularly in cases with sub-total resections, since they may present with aggressive recurrences.

Consent:

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Refrences:

1. Ahn JS, Harrison W, Hughes E, McLendon RE (2019). Intraventricular pilocytic astrocytoma with KIAA1549/BRAF fusion arising in a 44-year old. Journal of Neuropathology & Experimental Neurology 78(2):187-190.
2. Al Saleem MA, Alsafwani NS, ElSharkawy TM, Al-Suhibani SS, Essawi W(2020). A 37-Year-Old Saudi Man with a Calcified Intraventricular Astrocytoma. The American Journal of Case Reports 21 :e928224-928221.
3. Bond KM, Hughes JD, Porter AL, Orina J, Fang S, Parney IF (2018). Adult pilocytic astrocytoma: an institutional series and systematic literature review for extent of resection and recurrence. World neurosurgery 110:276-283.
4. Dash C, Moorthy S, Garg K, Sawarkar D, Singh PK, Kaur K, et al (2022). Intraventricular Pilocytic Astrocytoma: A Single Centre Experience. Neurology India 70(4):1468.
5. Hendrix J, Xiong Z (2020). Intraventricular pilocytic astrocytoma in an adult patient. Intractable & Rare Diseases Research 9(4):260-262.
6. Khan K, Luther E, Morrell AA, Tan SK, Eichberg DG, Shah AH, et al (2021). Recurrent adult pilocytic astrocytoma presenting with intraventricular and leptomeningeal spread. Surgical neurology international 12.

7. Lee Y-Y, Van Tassel P, Bruner JM, Moser RP, Share JC (1989). Juvenile pilocytic astrocytomas: CT and MR characteristics. American journal of neuroradiology 10(2):363-370.
8. Sandberg DI, Souweidane MM (1999). Hemifacial spasm caused by a pilocytic astrocytoma of the fourth ventricle. Pediatric neurology 21(4):754-756.
9. Sattar S, Akhunzada NZ, Javed G, Uddin Z, Khan YA (2017). Pilocytic astrocytoma: A rare presentation as intraventricular tumor. Surgical Neurology International 8.
10. Theeler BJ, Ellezam B, Sadighi ZS, Mehta V, Tran MD, Adesina AM, et al (2014). Adult pilocytic astrocytomas: clinical features and molecular analysis. Neuro-oncology 16(6):841-847.
11. Xia J, Yin B, Liu L, Lu Y, Geng D, Tian W (2016). Imaging features of pilocytic astrocytoma in cerebral ventricles. Clinical neuroradiology 26(3):341-346.