Annals of Medicine and Surgery 65 (2021) 102300

Contents lists available at ScienceDirect

Annals of Medicine and Surgery

journal homepage: www.elsevier.com/locate/amsu

Case Report

Adult Pilocytic Astrocytoma in the insula: Case report and review of the literature

Baha'eddin A. Muhsen, MD, MRCSI, FEBNS [a,*], Ansam Ghzawi [b], Hasan Hashem [e], Maher Elayyan [a], Bayan Maraqa [c], Mahmoud Al Masri [d]

[a] Division of Neurosurgery, Department of Surgery, King Hussein Cancer Center, Amman, Jordan
[b] Faculty of Medicine- Yarmouk University, Irbid, Jordan
[c] Department of Pathology, King Hussein Cancer Center, Amman, Jordan
[d] Department of Surgery, King Hussein Cancer Center, Amman, Jordan
[e] Division of Pediatric Hematology Oncology, King Hussein Cancer Center, Amman, Jordan

ARTICLE INFO

Keywords:
Pilocytic Astrocytoma
Adult
Insular
Microsurgery
Outcomes

ABSTRACT

Introduction: and Importance: Adult Pilocytic Astrocytomas (APA) are infrequent low grade tumors. While supratentorial APA is considered rare, insular APA is extremely rare.

Case presentation: We present a case of pure insular APA along with surgical outcomes. Tractography and functional MRI were obtained pre-operatively. The patient underwent neuro-navigation guided microsurgical resection with sub-cortical white matter mapping, utilizing Intra-operative MRI guidance. The Sylvain fissure was opened to secure the M3 branches, and near total resection was achieved.

Clinical discussion: APA in the insula is a very rare presentation and is considered challenging. Its proximity to the middle cerebral and lenticulostriate arteries, motor areas, and language areas makes accessing and resecting the tumor challenging. A multidisciplinary approach by an experienced team is needed to plan the management of young adult patients and reach the best outcomes.

Conclusion: Implementing microsurgical techniques, modern imaging modalities and intraoperative mapping helps to achieve maximal safe resection without risking functions.

Introduction

Adult Pilocytic astrocytomas (APA) are rare tumors. They are classified as Word Health Organization grade I gliomas [6]. PAs in general tend to be more prominent in children and adolescents. The incidence of PAs decreases significantly from 11.4% in children and adolescents to only 0.8% of CNS tumors in adults [7]. In addition, tumors location distribution seems to be related to the age of the patients. As it is more prominent to observe cerebellar presentations in pediatrics, in contrast to supratentorial location prominence in adults [10]. PAs present usually manifest with a space-occupying lesion affecting the posterior fossa as obstructive hydrocephalus with clinical pictures of headache, vomiting, nausea, and papilledema [9]. The excision of the tumor surgically is the treatment of choice aiming at tumor-free margins, with the least neurological insults. With recurrence, further resection surgically is mainly performed. APA in the cerebral hemispheres is very rare in the literature, insular location is even extremely rare. Surgical resection of insular tumors is challenging due to their proximity to the middle cerebral artery (MCA), lenticulostriate arteries (LSA), motor areas, and language areas. Herein, we present a rare case of adult pilocytic astrocytoma presenting in the insula that have been managed at a cancer centre in Jordan and discuss it with the available literature. This case has been reported according to **SCARE** criteria for case reports [1].

Case report

A 23 year-old right-handed male patient presented to the ER by his parents with severe progressive headache associated with short-term memory loss and refractory seizures (KPS 60). The patient is known to have epilepsy since the age of 3 years and has a history of left-sided

Abbreviations: KPS, Karnofsky scale; MCA, middle cerebral artery; LSA, lenticulostriate arteries.

* Corresponding author. Neurosurgical-Oncology and Skull Base Surgery, Department of Surgery, Division of Neurosurgery, King Hussein Cancer Center, 202 Queen Rania Al Abdullah St., P.O Box 1269, Amman, 11941, Jordan.
E-mail addresses: BM.14911@khcc.jo, Bmuhsen08@gmail.com (B.A. Muhsen).

https://doi.org/10.1016/j.amsu.2021.102300
Received 1 March 2021; Received in revised form 7 April 2021; Accepted 7 April 2021
2049-0801/© 2021 Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY license (http://creativecommons.org/licenses/by/4.0/).

insular brain tumor for four years. He was observed conservatively for four years through serial Magnetic Resonance Imaging (MRIs). On examination, the patient showed loss of tandem gait, pronator drift and weakness in the right side. CT scan was performed to exclude any active brain hemorrhage. Subsequent MRI revealed significant increase in the size of both cystic and solid parts of the tumor with midline shift and obstructive hydrocephalus (Fig. (1A, B)). Awake craniotomy option was discussed with the patient but was uncooperative. Preoperative imaging assessment with Tractography MRI showed displacement of the corticospinal tract (Fig. 2) by the tumor. Functional MRI was performed to identify speech area (Fig. 2). Neuronavigation guided Surgical resection with cortical mapping and subcortical white matter mapping was performed by *BM MD, MRCSI, FEBNS* - a senior neurosurgical oncology and skull base surgery specialist practicing in a tertiary cancer centre-after opening the sylvain fissure to secure the M3 branches and cortictomy over the preplanned trajectory (Fig. 1, A). Motor cortex and corticospinal tract mapping using a bipolar probe as a stimulator was utilized (Fig. 3). Near total resection was safely achieved Figure (1C, D). Histopathological study revealed pilocytic astrocytoma with varying proportions of compacted bipolar cells and loose microcystic areas. Numerous Eosinophilic granular bodies are seen, as well as many entrapped ganglion cells. Rare mitotic figures are identified. The tumor cells are positive for GFAP, negative for CD34, P53 and IDH-1. ATRX is Lost in tumor cell nuclei. Ki67 index is around 4%. The postoperative KPS was 80 until the last follow up of 12 weeks.

Discussion

Pilocytic astrocytoma is very rare in adults, with only scarce data available on the tumor characteristics, clinical course and prognostic factors. To our knowledge, this is the first report describing pilocytic astrocytoma in the insula for an adult patient. The presentation of the tumor usually mimics the manifestation caused by a space-occupying lesion affecting the posterior fossa such as obstructive hydrocephalus with a clinical picture of headache, seizures, vomiting, nausea, and papilledema.

The tumors location in adults goes in another direction when compared with the pediatric population, as its more prominent in the posterior fossa. Ye et al. [13], Ishkanian et al. [5] and Bond et al. [2] all have published studies that analyzed the patients data and found that the tumor distribution was infratentorial in more than 50% of the cases, which is similar to pediatric PA.

Theeler et al. [11] have analyzed the data of 127 adult patients with PA and found a predominance of supratentorial tumors in 53% of the cases. However, 13% of their cohort were reported as ventricular tumors, without further specifications. Boschetti et al. [3] showed the same with 73.9% supratentorial distribution in their adult APA cohort.

For all PA's, the excision of the tumor surgically is the treatment of choice aiming at tumor-free margins, with the least neurological insults. With recurrence, further resection surgically is usually performed.

Cyrinea et al. [4] investigated lesions in the eloquent and

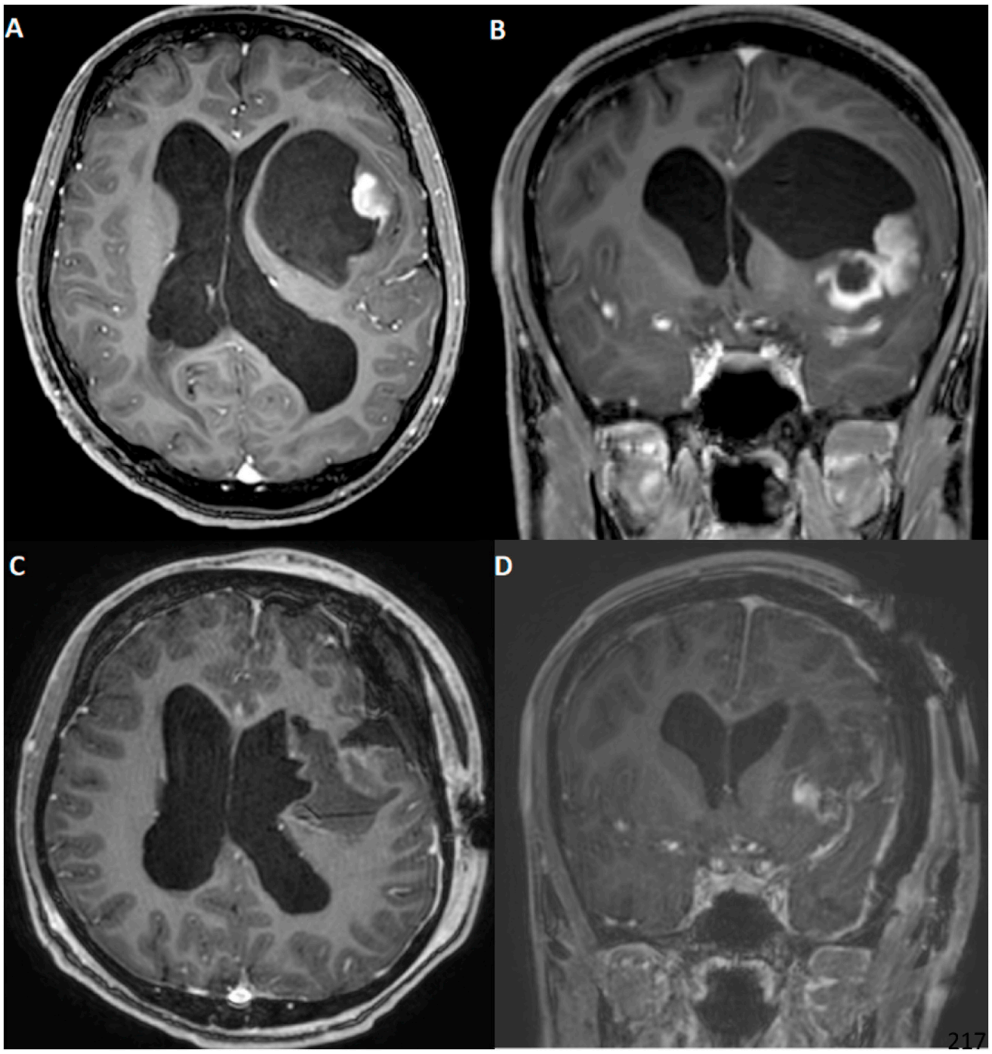

Fig. 1. Brain MRI with contrast; A) Axial preopertaive, B) Coronal preopertaive, C) Axial post pertaive, D) Coronal postopertaive views.

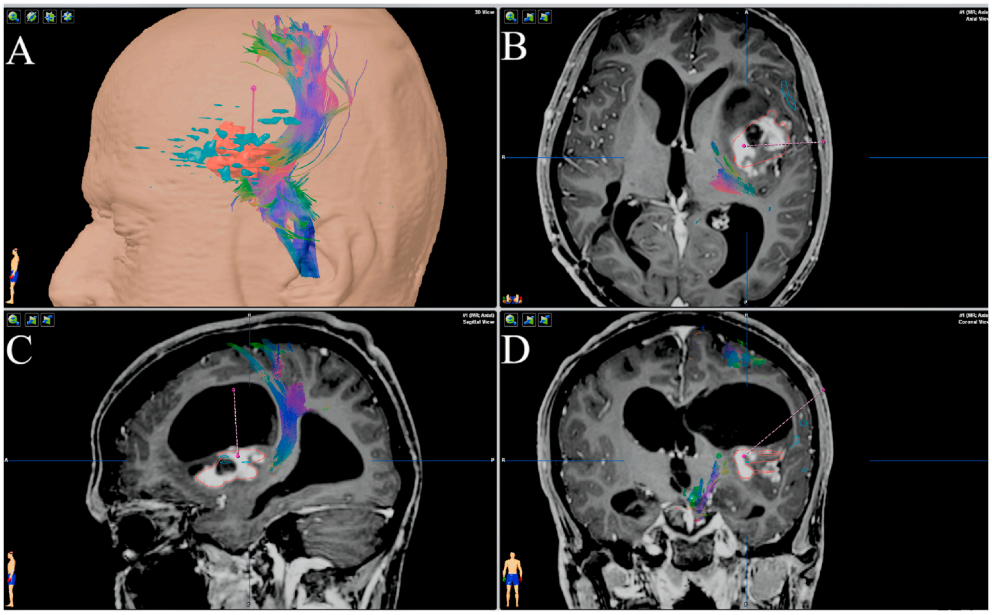

Fig. 2. 3D reonsrtucation of the coticoapinal tract, tumor (in red), speech area (blue), planned trajectory (in pink) **A**, Axial **B**, Sgaiital **C**, Coronal **D** Brain MRI with contrast sequences showing tumor corelation to the coticpsinal (Dark blue fibers) tract and speech area (dotted blue ares). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

deep-seated areas like the optochiasmatic area, diencephalon, and brainstem. These locations are significant challenges in the face of surgical interference while biopsy and subtotal resection may be the only and best treatment choice. In all, it raises the suspicion that a more limited resection extent, due to constrained surgical accessibility, could be related to the elevated progress rates and worse prognosis attributed to specific sites. Though the resection extent usually has a connection to the survival rate in most of the studies assessing these factors, a resection extent appraisal can be complicated and cause a biased investigation.

The extent of resection (EOR) is a main prognostic factor in Overall Survival OS, malignant transformation, and progression-free survival (PFS). Yahanda et al. [12] investigated 284 subjects undergoing grade I gliomas treatment. They had 248 tumor resections; the higher is the EOR, the more improved is the five-year OS with $p = 0.013$ and the five-year PFS with $p < 0.0001$.

Resection of the tumor enhanced the life quality for patients manifesting with intractable epilepsy. Regarding the evidence presented, trying to maximize the tumor resection and minimize the neurological effect should be aimed at insular glioma intervention. The challenge that every neurosurgeon encounters is to control both seizures and lesions while preserving most of the functions. Collected experience in resecting these tumors established significant flexibility in the region. Recognition of social cognition, cognitive functions, and language intraoperatively of the right non-dominant hemisphere (RndH) utilizing intraoperative brain mapping (ioBM) could be of utmost importance in enhancing the resection extent of low-grade gliomas and positively affect the neuropsychological and clinical result at six months [8] which might permit the majority of patients to reimburse overtime after aggressive resections of insular gliomas given that motor locations recognized by stimulation and essential language are maintained in surgery. Implementing microsurgical techniques, modern imaging modalities and intraoperative mapping helps to achieve maximal safe resection.

Conclusion

To the best of our knowledge, this case is completely unique owing to the complete observational follow-up data available for insular APA. Implementing microsurgical techniques, modern imaging modalities and intraoperative mapping helps to achieve maximal safe resection without risking functions.

Ethical approval

N/A.

Source of funding

No funding was received for this research.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Author contribution

Baha'eddin A. Muhsen MD1: Performed the intervention, supervised and reviewed the manuscript.

Ansam Ghzawi MS2: Manuscript writing, final revision.

Hasan Hashem MD5: Editing and reviewing the manuscript. Maher Elayyan1, Bayan Maraqa MD3 and Mahmoud Al Masri4: finalizing the review process.

Research registration (for case reports detailing a new surgical technique or new equipment/technology)

N/A.

Guarantor

Baha'eddin A. Muhsen MD.

*Division of Neurosurgery, Department of Surgery, King Hussein Cancer Center, Amman, Jordan.,Email: Bmuhsen08@gmail.com,Tel: +962777990888.

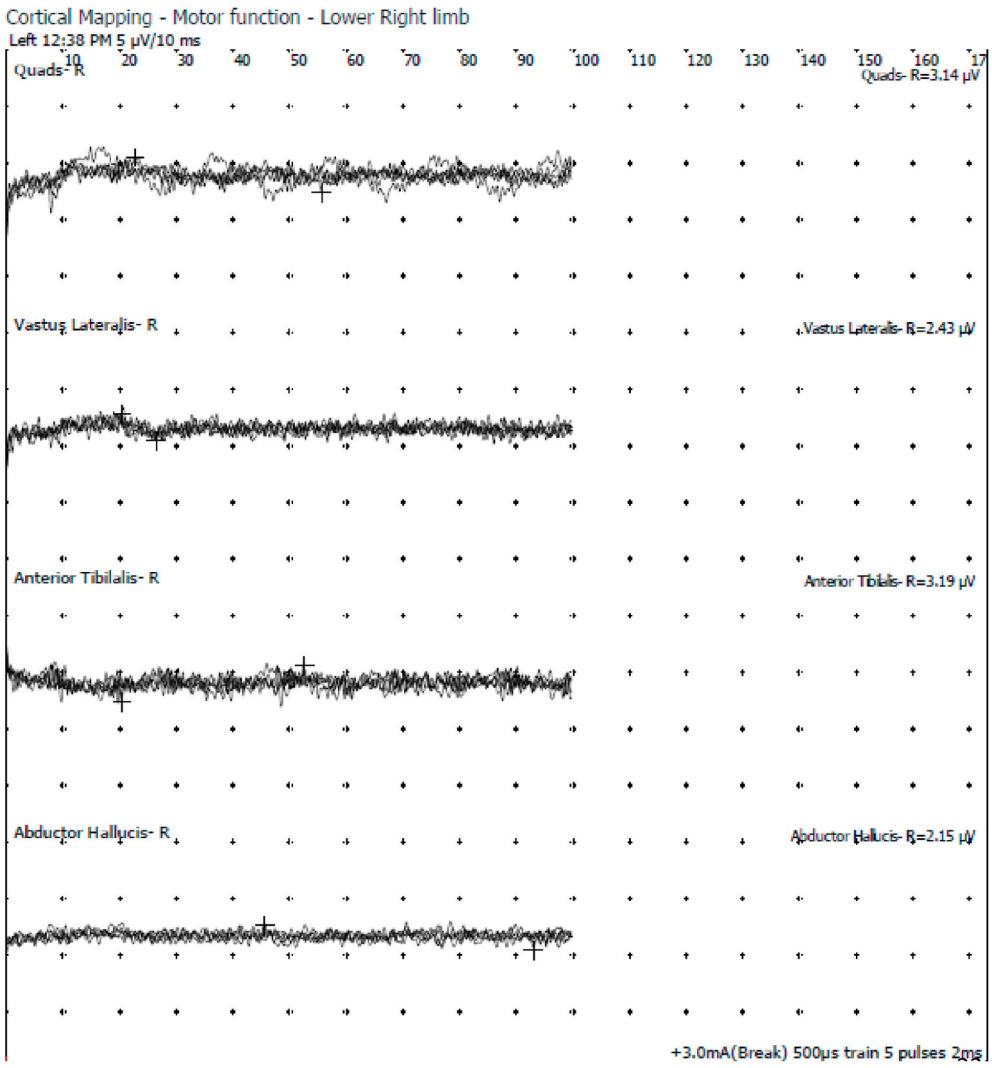

Fig. 3. Motor mapping in the lower limbs.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

No conflicts of interest to disclose

Acknowledgements

We would like to thank our patient for allowing permission to publish the case.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102300.

References

[1] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, SCARE Group, The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. S1743–9191 (20) (2020) 30771–30778.

[2] K.M. Bond, J.D. Hughes, A.L. Porter, J. Orina, S. Fang, I.F. Parney, Adult pilocytic astrocytoma: an institutional series and systematic literature review for extent of resection and recurrence, World Neurosurg. 110 (2018) 276–283.

[3] G. Boschetti, A.J. Santos, K.P. Fermon, G.L. de Freitas Honório, G. Batistella, S. Z. Kusma, J.N. Stávale, M.A. de Paiva Neto, S.M. Fleury Malheiros, Adult pilocytic astrocytomas: a Brazilian series, World Neurosurg. 133 (2020) e115–e120.

[4] S. Cyrine, Z. Sonia, T. Mounir, S. Badderedine, T. Kalthoum, K. Hedi, M. Moncef, Pilocytic astrocytoma: a retrospective study of 32 cases, Clin. Neurol. Neurosurg. 115 (8) (2013) 1220–1225.

[5] A. Ishkanian, N.J. Laperriere, W. Xu, B.-A. Millar, D. Payne, W. Mason, A. Sahgal, Upfront observation versus radiation for adult pilocytic astrocytoma, Cancer 117 (17) (2011) 4070–4079.

[6] D.N. Louis, A. Perry, G. Reifenberger, A. von Deimling, D. Figarella-Branger, W. K. Cavenee, H. Ohgaki, O.D. Wiestler, P. Kleihues, D.W. Ellison, The 2016 world Health organization classification of tumors of the central nervous system: a summary, Acta Neuropathol. 131 (6) (2016) 803–820.

[7] Q.T. Ostrom, H. Gittleman, G. Truitt, A. Boscia, C. Kruchko, J.S. Barnholtz-Sloan, CBTRUS statistical report: primary brain and other central nervous system tumors diagnosed in the United States in 2011-2015, Neuro Oncol. vol. 20 (2018) suppl_4): iv1–iv86.

[8] R. Prat-Acín, I. Galeano-Senabre, P. López-Ruiz, A. Ayuso-Sacido, R. Espert-Tortajada, Intraoperative brain mapping of language, cognitive functions, and social cognition in awake surgery of low-grade gliomas located in the right non-dominant hemisphere, Clin. Neurol. Neurosurg. 106363 (2020), 106363.

[9] B.M. Stein, M.S. Tenner, R.A. Fraser, Hydrocephalus following removal of cerebellar astrocytomas in children, J. Neurosurg. 36 (6) (1972) 763–768.

[10] C. Stüer, B. Vilz, M. Majores, A. Becker, J. Schramm, M. Simon, Frequent recurrence and progression in pilocytic astrocytoma in adults: adult Pilocytic Astrocytoma, Cancer 110 (12) (2007) 2799–2808.

[11] B.J. Theeler, B. Ellezam, Z.S. Sadighi, V. Mehta, M.D. Tran, A.M. Adesina, J. M. Bruner, V.K. Puduvalli, Adult pilocytic astrocytomas: clinical features and molecular analysis, Neuro Oncol. 16 (6) (2014) 841–847.
[12] A.T. Yahanda, B. Patel, G. Sutherland, et al., A multi-institutional analysis of factors influencing surgical outcomes for patients with newly diagnosed grade I gliomas, World Neurosurg. 135 (2020) e754–e764.
[13] J.M. Ye, M.J. Ye, S. Kranz, P. Lo, A 10 year retrospective study of surgical outcomes of adult intracranial pilocytic astrocytoma, J. Clin. Neurosci. 21 (12) (2014) 2160–2164.