Trigone Neurenteric cyst

Neurenteric cyst (NC) shows benign histopathology and rarely demonstrate malignant transformation. We herein describe a case of NC that exhibited malignant transformation. A 65-year-old female presented with gait disturbance due to compression by a cystic mass on the dorsal surface of the medulla oblongata. Partial resection was performed twice, leading to improvement of her symptoms. Two years after the second surgery, gadolinium-perfused T1-weighted magnetic resonance imaging revealed an invasive lesion with contrast enhancement at the trigone of the left lateral ventricle for which partial resection followed by radiotherapy was performed. However, mass regrowth was observed, with the patient eventually succumbing to her disease 11 months after her third surgery. Histopathological analyses of the first and second surgical specimens identified pseudostratified cuboidal epithelial cells, with no nuclear or cellular atypia resembling gastrointestinal mucosa, lining the inner surface of the cystic wall. Based on these findings the lesion was diagnosed as NC. The third surgical specimen exhibited apparent malignant features of the epithelial cells with elongated and hyperchromatic nuclei, several mitotic figures, small necrotic foci, and a patternless or sheet-like arrangement. Based on these findings, the lesion was diagnosed as NC with malignant transformation. Next-generation sequencing revealed KRAS p.G12D mutation in all specimens. Additionally, the third surgical specimen harbored the following 12 de novo gene alterations: ARID1A loss, BAP1 p.F170L, CDKN1B loss, CDKN2A loss, CDKN2B loss, FLCN loss, PTCH1 loss, PTEN loss, PTPRD loss, SUFU loss, TP53 loss, and TSC1 loss. The aforementioned results suggest that KRAS mutation is associated with the development of the NC, and that the additional gene alterations contribute to malignant transformation of the NC [1)].

1)

Saito S, Natsumeda M, Sainouchi M, Takino T, Shibuya K, On J, Kanemaru Y, Ogura R, Okada M, Oishi M, Shimada Y, Wakai T, Okuda S, Ajioka Y, Kakita A, Fujii Y. Elucidating the multiple genetic alterations involved in the malignant transformation of a KRAS mutant neurenteric cyst. A case report. Neuropathology. 2022 Dec;42(6):519-525. doi: 10.1111/neup.12822. Epub 2022 Sep 22. PMID: 36146951.