Intracranial germ cell tumor case reports

A mixed germ cell tumor including immature teratoma exhibiting growing teratoma syndrome is presented. The pathogenesis of growing teratoma syndrome remains unclear, and its treatment strategy has not been established. GCTs are often located within the ventricles, causing hydrocephalus, which sometimes improves after the removal of the tumor due to the restoration of cerebrospinal fluid (CSF) flow. On the other hand, even if the flow route of CSF from the third ventricle to arachnoid granulations on the brain surface quadrigeminal cistern is restored after removal of the tumor, hydrocephalus may not improve.

A case whose intractable hydrocephalus improved after penetrating the aqueductal membrane via endoscopy is described. An 11-year-old boy was treated for pineal intracranial growing teratoma syndrome (IGTS). The tumor grew rapidly in a short period, and hydrocephalus progressed despite an endoscopic third ventriculostomy (ETV). Although the obstruction was removed by radiation, chemotherapy, and total tumor resection, the hydrocephalus did not improve. Endoscopic membrane perforation was performed because a membrane-like structure was seen at the entrance of the cerebral aqueduct on magnetic resonance imaging. The hydrocephalus improved immediately after the operation, and the patient's consciousness disturbance also improved significantly.

The purpose of this report is to update the current knowledge and standards of management for patients with growing teratoma syndrome, as well as to drive future translational and clinical studies by recognizing the unmet needs concerning hydrocephalus [1)].

A ten-year-old girl initially presented with a large suprasellar mixed malignant germ cell tumor GCT with a near-complete response after initial induction chemotherapy and irradiation. Three and half years after initial therapy, she presented with progressively worsening vision in her left eye. Magnetic resonance imaging showed infiltrative changes within the left optic nerve but no discrete mass. Serum and cerebrospinal fluid (CSF) tumor markers were not elevated and CSF cytology was negative. Left optic nerve biopsy confirmed the presence of mature teratoma and pure germinoma components. She was treated with gross total resection of the left eye and optic nerve and chemotherapy. Histopathologic evaluation of the optic nerve showed only mature teratoma elements but with pure germinoma cells infiltrating the inner layers of the retina.

Loco-regional extension of suprasellar GCT to the optic nerve is not uncommon; however, infiltration of the tumor into the retina is not reported in the literature. Early detection of optic pathway involvement and proper delineation of the irradiation field may prevent GCT infiltration of the retina with subsequent vision loss [2)].

1)
Kajiwara S, Nakamura H, Sakata K, Komaki S, Negoto T, Morioka M. Endoscopic aqueductal membrane fenestration was effective for intractable hydrocephalus after removal of a nongerminomatous germ cell tumor exhibiting growing teratoma syndrome: a case report. BMC Pediatr. 2022 Nov 28;22(1):683. doi: 10.1186/s12887-022-03743-y. PMID: 36443673.
2)
Abu Arja MH, Stalling M, Governale LS, Pierson CR, Rusin JA, Palmer JD, Finlay JL, Olshefski R, Boué DR. Germinoma Involving the Retina: An Unusual Presentation of Recurrent Intracranial Mixed Germ Cell Tumor. World Neurosurg. 2019 Jan 7. pii: S1878-8750(19)30002-6. doi: 10.1016/j.wneu.2018.12.143. [Epub ahead of print] PubMed PMID: 30630044.