Pineal parenchymal tumor of intermediate differentiation

Pineal parenchymal tumor of intermediate differentiation (PPTID) is a rare disease, first classified by the World Health Organization in 2000.

In 2007, the World Health Organisation (WHO) reclassified pineal parenchymal tumours (PPT) from two subgroups pineocytomas (PCs) and pineoblastomas (PBs) into four, including pineal parenchymal tumours of intermediate differentiation (PPTID). PPTID have been further divided into low- and high-grade lesions (WHO II and III), but due to their rarity have proven difficult lesions to diagnose and a paucity of literature means their optimal treatment options are a challenge to define [1].

Case series

2014

The clinical data on five patients diagnosed with PPTID since 2000 were retrospectively reviewed. Patients with cerebrospinal dissemination at diagnosis received biopsy-only surgery, craniospinal and whole-ventricular irradiation and chemotherapy. Patients with locally limited disease at diagnosis received local or whole-ventricular irradiation after surgery. The median relapse-free and overall survival were 72.9 and 94.1 months, respectively. Two of the five patients developed a relapse of cerebrospinal dissemination after treatment and succumbed to the disease. All the patients who received both craniospinal and whole-ventricular irradiation exhibited evidence of cerebral white matter abnormalities in magnetic resonance imaging and developed neurocognitive disorders after treatment. Although PPTID may be aggressive and has cerebrospinal fluid seeding potential, PPTID patients may survive long-term, even after recurrence. Considering the long survival time and the late adverse effects due to intensive treatment, the irradiation field and usage of chemotherapy after surgery require optimization [2].

Case reports

Bando et al., from Shinko Hospital, Kobe, Japan, report a rare case of pineocytoma (PC) in a 63-year-old woman who presented with lower-extremity weakness and gait disturbance. A pineal mass lesion was detected on MRI. A diagnosis of (PC) was established after microsurgical gross total resection, and the patient received no adjuvant therapy after surgery. Two years after surgery, a partial recurrence was recognized and Gamma Knife radiosurgery was performed. Fours years later, the patient developed diffuse leptomeningeal dissemination. She was successfully treated with craniospinal irradiation. Leptomeningeal dissemination may develop 6 years after the initial diagnosis of PC. A histopathological study of the recurrent tumor revealed a malignant change from PC to PPTID. The present case shows the importance of long-term follow-up of patients with pineal parenchymal tumor following resection and the efficacy of craniospinal irradiation in the treatment of leptomeningeal dissemination [3].

1)
Amato-Watkins AC, Lammie A, Hayhurst C, Leach P. Pineal parenchymal tumours of intermediate differentiation - An evidence-based review of a new pathological entity. Br J Neurosurg. 2015 Nov 16:1-5. [Epub ahead of print] PubMed PMID: 26571134.

2)

Watanabe T, Mizowaki T, Arakawa Y, Iizuka Y, Ogura K, Sakanaka K, Miyamoto S, Hiraoka M. Pineal parenchymal tumor of intermediate differentiation: Treatment outcomes of five cases. Mol Clin Oncol. 2014 Mar;2(2):197-202. Epub 2013 Dec 23. PubMed PMID: 24649332; PubMed Central PMCID: PMC3917789.

3)

Bando T, Ueno Y, Shinoda N, Imai Y, Ichikawa K, Kuramoto Y, Kuroyama T, Shimo D, Mikami K, Hori S, Matsumoto M, Hirai O. Therapeutic strategy for pineal parenchymal tumor of intermediate differentiation (PPTID): case report of PPTID with malignant transformation to pineocytoma with leptomeningeal dissemination 6 years after surgery. J Neurosurg. 2018 Jul 20:1-7. doi: 10.3171/2018.2.JNS171876. [Epub ahead of print] PubMed PMID: 30028263.