

Papillary Tumor of the Pineal Region: A Case Report

Hamid Nasrollahi**, Bita Geramizadeh**, Mansour Ansari***,
Seyed Hassan Hamedei*, Mohammad Mohammadianpanah****,
Shapour Omidvari***, Niloofar Ahmadloo**, Ahmad Mosalaei*****

*Department of Radiation Oncology, Nemazee Hospital, Shiraz University of Medical Sciences, Shiraz, Iran

** Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran

***Breast Diseases Research Center, Shiraz University of Medical Sciences, Shiraz, Iran

****Colorectal Research Center, Shiraz University of Medical Sciences, Shiraz, Iran

*****Shiraz Institute for Cancer Research, Shiraz University of Medical Sciences, Shiraz, Iran

Abstract

Pineal region tumors are uncommon lesions in the central nervous system. Papillary tumor of the pineal region is recently recognized as a separate disease. Its incidence, treatment, and outcome are not well-defined. We have reported the case of a 6 year-old-boy with papillary tumor of the pineal region. He presented with headaches, nausea and vomiting and, after a biopsy, was referred for radiotherapy. The patient received 54 Gy irradiation to the pineal region followed by a chemotherapy regimen of cisplatin, vincristine, and lomostine. His tumor decreased slightly. After 4 years, the patient has remained well and attends school. He is under routine follow-up. We suggest radical radiotherapy as the main treatment for papillary tumors of the pineal region.

Keywords: Pineal gland, Neoplasm, Papillary tumor

Introduction

Pineal region tumors comprise only 1% of primary central nervous system (CNS) tumors.¹ The germ cell tumor is the most common primary tumor in this region. In this space, pineal parenchymal tumors, astrocytomas, and meningiomas can be seen.^{1,2} Previously, these tumors have been irradiated empirically without pathological diagnosis and

tissue biopsy.³

The papillary tumor of the pineal region (PTPR) was introduced for the first time in 2003 as a new entity.⁴ Jouvett et al. reported 6 patients who had distinct pathologies from the pineal region.⁵ They have observed which PTPR have a characteristic epithelial-like growth pattern that most likely originates from the subcommissural organ.⁵ Hence,

♦Corresponding Author:

Hamid Nasrollahi, MD
Department of Radiation
Oncology, Nemazee Hospital
Shiraz University of Medical
Sciences, Shiraz, Iran
Tel: +98 713 6125170
Fax: +98 713 6474320
Email: nasrolahiamid@yahoo.com

distinction between the PTPR, parenchymal pineal tumors, and germinoma tumors can be difficult.⁶

This tumor is very rare; therefore, proper treatment and prognosis are not well-documented.^{1,7} Herein, we report the case of an six-year-old child with PTPR to share our experience with other clinicians.

Case Report

A 6-year-old boy presented to our clinic in September, 2012 with severe nausea, vomiting, and headaches for 3-4 weeks. He was admitted to the Pediatric Gastroenterology Ward where his work-ups were all normal. A brain magnetic resonance imaging (MRI) showed a lesion in the pineal region (Figure 1). The neurological examination was normal. The patient underwent biopsy and the pathology showed very tiny fragments of tumor tissue with one or two papilla that had a fibrovascular core lined by eosinophilic cells with indistinct cytoplasmic borders, and a

columnar to cuboidal shape. Blood vessels appeared to be lined by multiple layers of tumor cells with mild pleomorphism (Figure 2). The immunohistochemistry (IHC) study was reactive for vimentin and S-100 protein, with low Ki67. Epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP), and synaptophysin were negative. Pathologic examination favored PTPR. The patient received local radiotherapy with a dose of 54 Gy in 27 fractions. Brain MRI during the follow-up remained unchanged. He received 6 cycles of cisplatin, vincristine, and lomostine. The subsequent 3 MRIs showed that the lesion decreased and eventually disappeared. After 4 years, the patient has remained well with no physical or mental problems.

Discussion

Papillary tumor of the pineal region is a disease of the 21st century. Morphological similarities

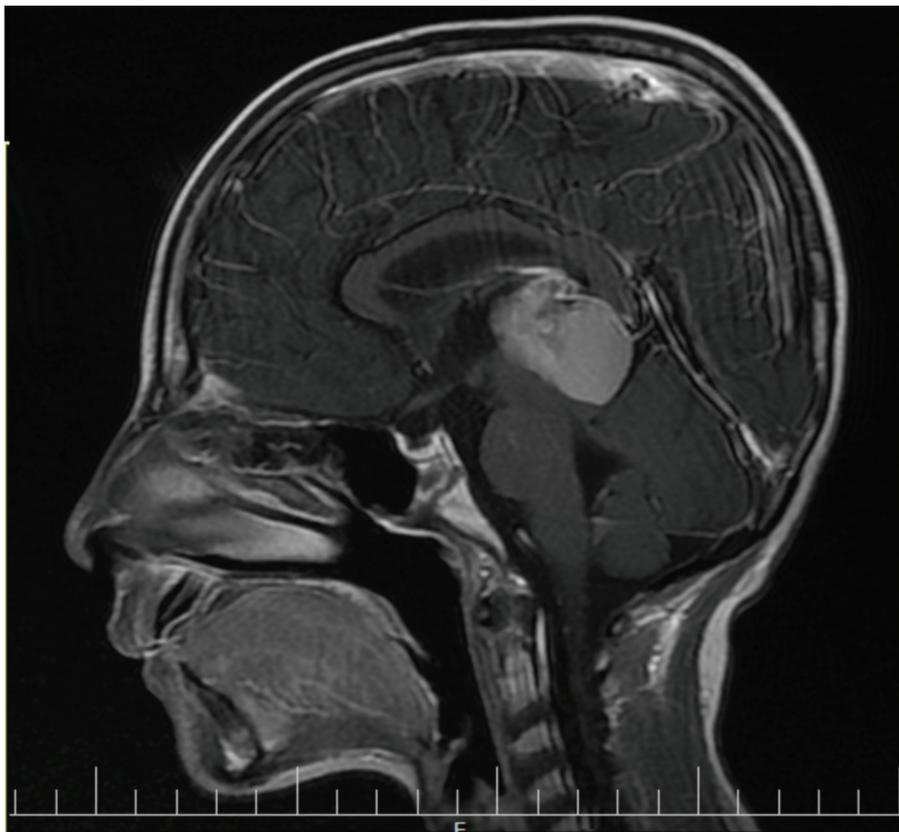


Figure 1. Contrast-enhanced sagittal and axial view of T1-weighted magnetic resonance image (MRI) of the brain that shows a 16×8 mm mass in the pineal region.

may arise from the ependymocytes at the subcommissural organ.⁸ In 2007, the World Health Organization (WHO) Classification of Brain Tumors recognized this entity as a distinct disease. The PTPR is considered a grade II/III brain tumor.^{5,8} Surgery in this region is challenging and associated with serious side effects. Complications of surgery, such as meningitis, Parinaud syndrome, CSF leakage, cranial nerve palsy, and hemiparesis are reported may occur in upto 18% of patients.⁹

Our patient had an indolent clinical course. We found a similar clinical course reported by Nakamura et al. in an 11-year-old boy who initially had surgery and irradiated as Primitive Neuro-Endocrine Tumor (PNET). The tumor reduced by 30% after biopsy, radiotherapy up to 50.4 Gy, and chemotherapy. Their chemotherapy regimen consisted of nimustine hydrochloride and

vincristine for one course. After 15 years, the patient had remained well and disease-free. This was the longest disease-free survival with PTPR that we have found.¹⁰

Primary radiotherapy can produce a complete tumor response and may be helpful, whereas surgery may bring about serious unwanted results.⁹ Primary radiotherapy in the current study patient and as reported by Shibahara et al. did not produce complete tumor eradication. Shibahara et al. reported the case of a 29-year-old female patient with a primary diagnosis of pineoblastoma. After radiotherapy at a dose of 50 Gy, she had no observed response. The patient underwent an additional operation and was tumor-free for 9 months at the time of their report.⁴ Our patient has remained well after 4 years. Although he did not have a complete response, thus far he is well and symptom-free.

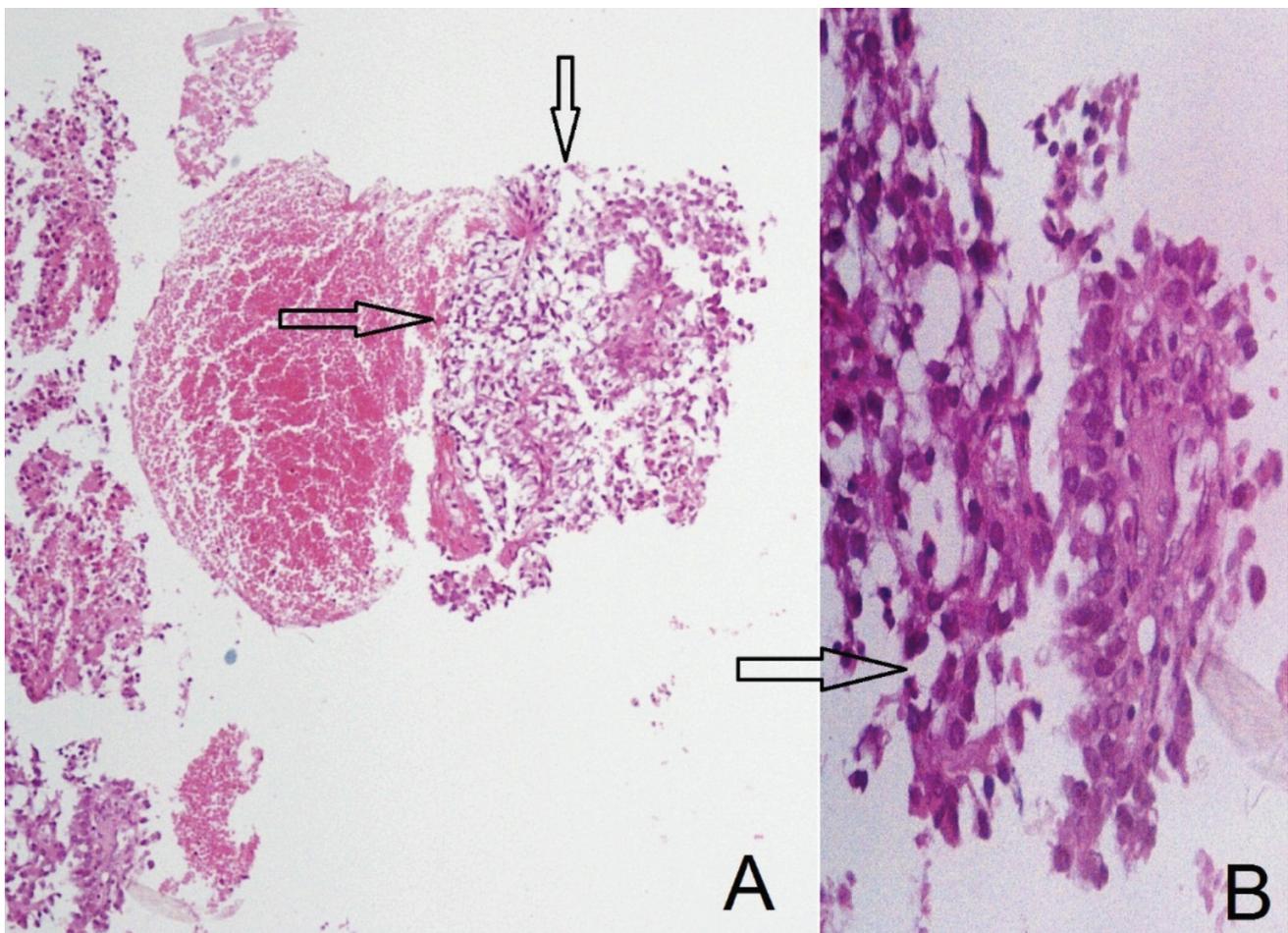


Figure 2. Sections from the brain tumor show papillary structures lined by cuboidal to columnar cells. Inset shows a high power view of one papilla (Figure A: $\times 40$, Figure B: $\times 200$, H & E).

We found no report of distant metastasis, but CSF seeding has been reported after frequent local recurrences.¹¹ Hong et al. reported the case of a 39-year-old woman that had five pineal region tumor recurrences. She was treated as a pineoblastoma, and subsequently as an anaplastic ependymoma. Re-evaluation showed that the primary tumor was PTPR. The patient received stereotactic radiotherapy several times during the recurrences. She was relatively well, however she eventually developed CSF seeding in addition to local recurrence and expired 15 years after primary tumor presentation.¹¹ Hence, PTPR appears to have an indolent course despite CSF seeding.

Papillary tumor of the pineal region may have an aggressive behavior with early recurrence and poor response to treatment. Kim et al. reported the case of a woman with leptomeningeal spread over a two-week period after primary tumor treatment. She had multi-focal disease with poor response to treatment (radiotherapy with Gamma Knife). Chemotherapy was not recommended and she succumbed to her disease 11 months after presentation.¹² Others have reported poor responses and outcomes. Sato et al. reported that an 18-year-old male patient had early CSF dissemination and poor response to both chemotherapy and radiotherapy. He received ifosfamide, cisplatin and etoposide, along with craniospinal radiotherapy and a tumor boost to 55.8 Gy.¹³ Their patient developed early leptomeningeal spread and had a short (12 month) survival.¹³

The role of chemotherapy is not well-documented. Lorenzetti et al. reported the case of a woman who underwent surgery and developed recurrence after 2 years. She received irradiation, but had no satisfactory response. The patient was prescribed temozolomide and showed a partial response. The mass enlarged after 8 cycles of treatment; however, they continued temozolomide for 18 cycles and the tumor showed a good response.¹⁴ Sato et al. also reported that an 18-year-old male had no response to ifosfamide, cisplatin, and etoposide chemotherapy.¹³ The current study

patient had relatively no response to etoposide, lomostine, ifosfamide, and cisplatin.

Local recurrence is a major problem. Yano et al. have reported the case of a 17-year-old male previously treated as a papillary ependymoma. After 9 disease-free years, he had multiple recurrences and showed a good response to additional surgery, re-irradiation, and chemotherapy. At first, he received radiotherapy, then, 9 years later he developed recurrence for which he received re-irradiation and chemotherapy. After 17 and 68 months, he underwent additional radiotherapy. His tumor disappeared 21 months after the last irradiation.¹⁵

We continue to closely follow our case to detect and treat any inadvertent recurrences.

Conclusion

Papillary tumor of the pineal region usually has an indolent clinical course, but recurrence is frequent. It seems that chemotherapy is not very helpful.

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Conflict of Interest

No conflict of interest is declared.

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