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Case Report

Papillary tumor of the pineal region

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ABSTRACT

Pineal region tumors make up 0.4-1.0% of intracranial tumors in adults. These tumors may arise from pineal gland itself or structures around pineal gland which are all termed as pineal region tumors. Papillary tumor of the pineal region is a non-parenchymal tumor of the pineal region. It was first documented in 2003. We presented a 21 years old female patient with pineal tumor. Supracerebellar infratentorial approach was performed. The total excision of the lesion was achieved successfully. Histopathological examination revealed papillary tumor of the pineal region. The case is discussed in the light of our preoperative surgical experience, its pre-and post-operative radiological and histopathological evaluation. Supracerebellar infratentorial approach is encountered as the favoured approach generally. As recurrence rates are high, adjuvant treatment is advised after surgery. Further research on this pathology will enlighten the neurosurgeons for effective treatment.

Keywords: Papillary, Pineal, Region, Tumor

INTRODUCTION

Papillary tumor of the pineal region (PTPR) is a relatively newly accounted addition to the World Health Organization (WHO) classification.[1] It was firstly documented by Jouvet et al. in 2003.[2] Pineal region tumors make up 0.4-1.0% of intracranial tumors in adults and 3.0-8.0% of brain tumors in children.[3] Pathologies in or around pineal region are mainly out of neuroaxis in origin but may also derive from pineal gland. Whereas its primary pathologies potentially arise from pineal parenchymal cells; secondary pathologies do not originate from neural structures such as germ cells, ependymal cells, glial cells, the meninges, and metastases.

In the human pineal gland, the rosette-like clusters of cells are considered to be ependymal cells, which invade the pineal parenchymal from the subcommisural organ in the first months of the perinatal period. The tumor described here as PTPR appears to be derived from the specialized ependymal cells of the subcommissural organ.[4]

Parenchymal tumors originating from the pineal gland itself account for 14-27% of pineal region tumors. In addition to grade I pineocytoma and grade IV pineoblastoma, PTPR was recognized in the 2007 (WHO) classification as an intermediate-grade malignancy (II or III).

Rarely, other brain tumors, including meningioma, glioblastoma, ependymoma, plexus papilloma and neuroendocrine tumors (NETs), can also occur in the pineal region.[5,6] Interestingly, nonparenchymal tumors arising from pineal region are more common than parenchymal tumors. In this paper, a patient with PTPR is reported and this rare pathology is discussed and presented in detail with its enriched radiological, pathological and surgical contents.

CASE REPORT

A 21-year old female is admitted to neurosurgery clinic with severe headache and dizziness. Her neurological examination was intact. Fundoscopy revealed bilateral slight papilledema. Initial magnetic resonance imaging (MRI) scans showed a pineal mass with contrast enhancement.

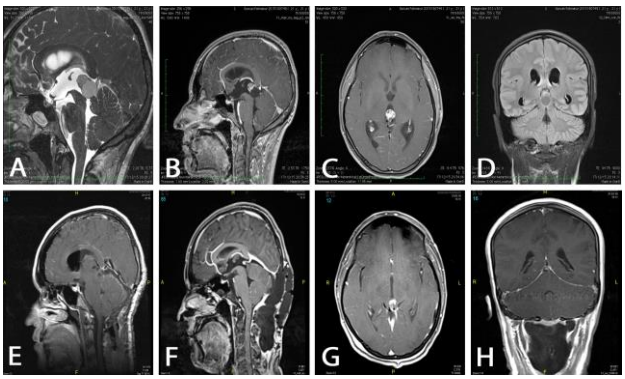

Figure 1: (A) Preoperative CISS MR sequence shows heterogenous mass in pineal region. (B-D) Preoperative MRI scans show an enhanced mass with cystic component (E) Sagittal T1 postcontrast MRI scan one week after surgery showing no contrast enhancement. (F) Postoperative third month follow-up MRI scans showing no residue or recurrence in operative field.

Tumor had a heterogeneous structure containing solid and cystic components. Although sagittal T1 post contrast MRI scans showed cystic components that are iso-intense with cerebrospinal fluid, sagittal CISS MRI revealed that cystic component was much viscous and less intense than cerebrospinal fluid.

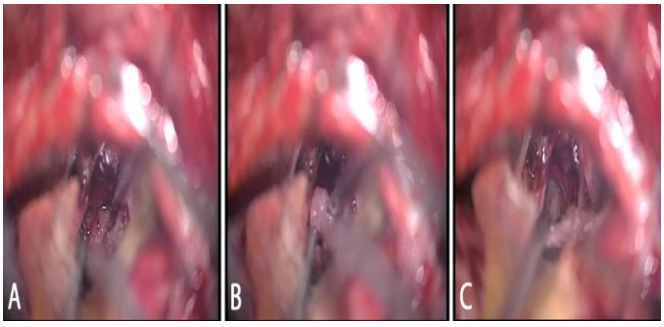

Figure 2: Intraoperative views. (A) Anatomical relationship of tumor with posterior venous complex. (B) Tumor removal (C) Exposure of third ventricle wall forming the posterior border of tumor.

The patient was operated with supracerebellar infratentorial approach. Total tumor resection was achieved. During surgery, pineal region vasculature was preserved. The tumor was observed as soft, easily aspirated and dissected from surrounding tissue. Solid and cystic components were also observed intraoperatively and after the exposure of third ventricle wall, the surgery was ended considering the total tumor removal.

Patient was taken to intensive care unit for a day and discharged one week after the operation. She had no neurological deficits. Postoperative MRI scan was performed one week after surgery revealing no contrast enhancement. Second MRI scan was performed 6 weeks after surgery for follow up and evaluation of any metastatic seeding. Cranial and spinal MRI scan was negative for seeding metastasis.

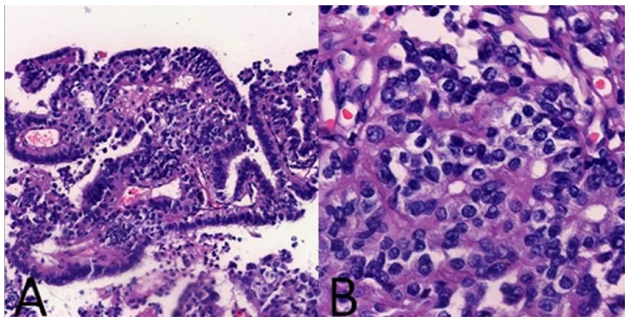

Figure 3: (A) Papillary structures with vascular cores and surrounding columnar cells; H&Ex169. (B) Solid areas comprising uniform round cells with round regular nuclei and eosinophilic cytoplasm. Some of them have clear cytoplasm with perinuclear halo. Lumen forming rosettes can also be noted; H&Ex504.

Pathological examination showed a tumor consisting of both papillary structures and solid areas. Papillary structures were actually pseudopapillae with epithelioid columnar cells surrounding fibrovascular cores. The solid areas showed sheets of round cells. Some of these cells demonstrated slightly clear cytoplasm with perinuclear halo. Lumen forming rosettes were also noted at the solid areas. There was no mitosis. Microvascular proliferation and necrosis did not detect. Immunohistochemical studies showed positivity of neuron specific enolase (NSE), vimentin, Pan-cytokeratin (Pan-CK: AE1+AE3) and synaptophysin. NSE and vimentin expression of tumor cells were diffuse.

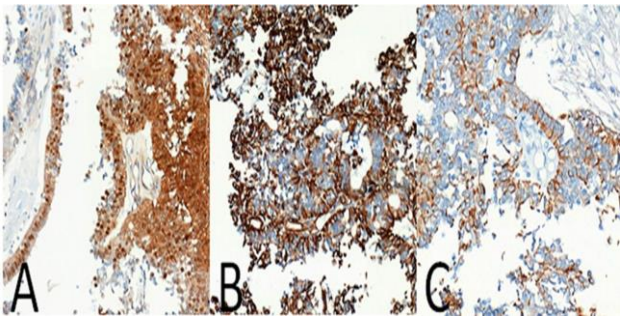

Figure 4: (A) Diffuse NSE expression of tumor cells; x169, (B) Diffuse vimentin expression of tumor cells; x175, (C) Pan-CK expression of tumor cells forming papillary structures; x250.

Pan-cytokeratin expression was mostly seen in the cells surrounding papillary structures. A few cells were positive with synaptophysin. Ki-67 (MIB-1) labeling index were 2%. CD117 showed diffuse but mild staining. Tumor cells were did not stained with glial fibrillary acidic protein (GFAP), S-100, p53, Chromogranin-A, epithelial membrane antigen (EMA), thyroid transcription factor-1 (TTF-1), CD10, Sall-4, placental alkaline phosphatase (PLAP), β-human chorionic gonadotropin (β-hCG). Tumor was diagnosed as PTPR with a comment explaining the uncertainty of these tumors' WHO grading criteria and tendency of local recurrence.

DISCUSSION

This case of PTPR showed similarities with the literature. There have been case reports presenting patients with similar clinical characteristics. Ninety-seven cases of PTPR have been reported in the literature so far including this study.[7] As PTPR is a new addition to the literature, more research is needed for the purpose of understanding its natural history for ideal treatment scheme and modality. As in germinomas, the definite diagnosis of PTPR may be possible with specific marks from blood or cerebrospinal fluid samples and cured without the necessity of surgery. In a literature review, only 1 spinal seeding metastasis is shown.[8]

Papillary tumor must always be included in the differential diagnosis of the pineal region tumors. The optimal treatment for this lesion appears to be surgery combined with adjuvant treatment. Because of the anatomical location of the pineal gland, gross total resection is hard to achieve. Radiotherapy appears to provide a decent local control of the disease. There are reports of definitive radiotherapy treatment. This treatment should be considered when surgical removal of the lesion poses high risk. There is a case of biopsy confirmed papillary tumor case with Gamma Knife SRS treatment. The case was followed disease free for 7 years.[9]

This study is also the first to stress the importance of CISS MRI sequence on radiological examination of PTPR. As surgery and radiotherapy targets pineal region and neighbouring anatomical structures, post-operative cognition defects must be considered. As many patients presented in the literature are younger than 30 years old, long-term effects of treatment and quality of life must be carefully evaluated. Multidisciplinary treatment for pineal region tumors is advised.

When dealing with pineal region tumors, endoscopic approach should always be considered. Depending on the neurosurgeon's choice a surgical removal should be the first line of treatment. Whether micro neuro surgical or neuroendoscopic approach is used, adjuvant treatment is advised for local control and disease-free survival.

CONCLUSION

PTPR is a new addition to the literature and should always be taken into account when dealing with pineal region tumors. Supracerebellar infratentorial approach is encountered as the favoured approach generally. As recurrence rates are high, adjuvant treatment is advised after surgery. Further research on this pathology will enlighten the neurosurgeons for effective treatment.

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