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Case Report

Papillary meningioma with a history of renal clear cell carcinoma and lung adenocarcinoma: a case report

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Abstract: Papillary meningioma is a rare subtype of malignant meningiomas. The case of papillary meningioma is extremely rarer than other tumors that may pose a diagnostic dilemma to the pathologist. Here we report a rare case of papillary meningioma following renal clear cell carcinoma and lung adenocarcinoma and suggest a useful indicator for accurate diagnosis. A 52-year-old female patient was diagnosed with renal clear cell carcinoma and lung adenocarcinoma two and half years ago, respectively. Four years ago, she presented with nausea, dizziness, and left ear pain when her headache was severe. The symptoms became progressively worse and more frequent, so she was subjected to left cerebellar craniotomy for resection of the tumor. On the basis of its morphologic and immunohistochemical features, the tumor was diagnosed as a papillary meningioma. Papillary meningioma needs to be differentiated from other intracranial tumors. Early diagnosis of papillary meningioma could significantly reduce the progression of subsequent invasion and mortality.

Keywords: Papillary meningioma, lung adenocarcinoma, renal clear cell carcinoma

Introduction

Meningiomas are the most common primary intracranial tumors. Atypical and malignant meningioma comprise a small fraction of the total (≤5%), and show a slight male predominance [1]. Malignant meningioma includes three types: namely papillary meningioma, rhabdoid meningioma, and anaplastic meningioma. According to the 2016 World Health Organization's (WHO) classifications, papillary meningioma (hereafter PM) is an uncommon subtype of meningioma. It is defined as grade III of the central nervous system tumors [2, 3]. PM occurs more commonly in young male patients and is usually accompanied by significant symptoms of intracranial hypertension, such as severe headaches, vomiting, and blurred vision. Here we report an extremely rare case of a female patient with three primary malignant tumors of the brain, kidney and lung in 6 years.

Case presentation

A 52-year-old female had unexplained gross hematuria 6 years ago which gradually worsened. Color Doppler revealed a space-occupying lesion in the right renal intralesional. The patient underwent laparoscopic radical nephrectomy for resection of the right kidney. A postoperative pathological examination showed grade I-II renal clear cell carcinoma (**Figure 1A**, **1B**). Four and a half years ago, the patient felt the pain in her left chest on occasion. In the examination, she was diagnosed with lung adenocarcinoma (**Figure 2A**, **2B**), and the occupied lesions were discovered at the left hilar. The final diagnosis of lung adenocarcinoma was made by percutaneous lung puncture biopsy under CT guidance. Then the EGFR gene detection confirmed the mutations. She was started on gefitinib in irregular intervals. Four years ago, this patient arrived at our hospital with the complaint of intermittent headaches accompanied by nausea, dizziness, and left ear pain when the headache was severe. The above symptoms gradually worsened and became more frequent. She was subjected to left cerebellar craniotomy for resection of the tumor on March 1st, 2014. After 4 years, the follow-up neuroimaging showed no signs of recurrence.

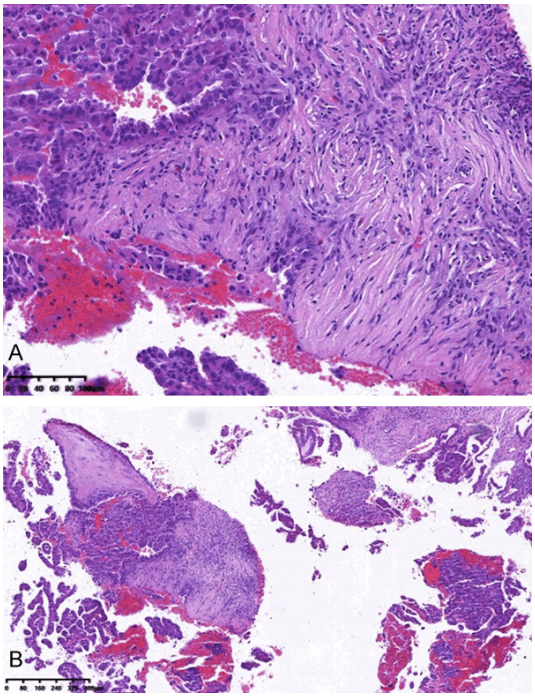

Figure 1. Pathologic diagnosis showed pulmonary adenocarcinoma.

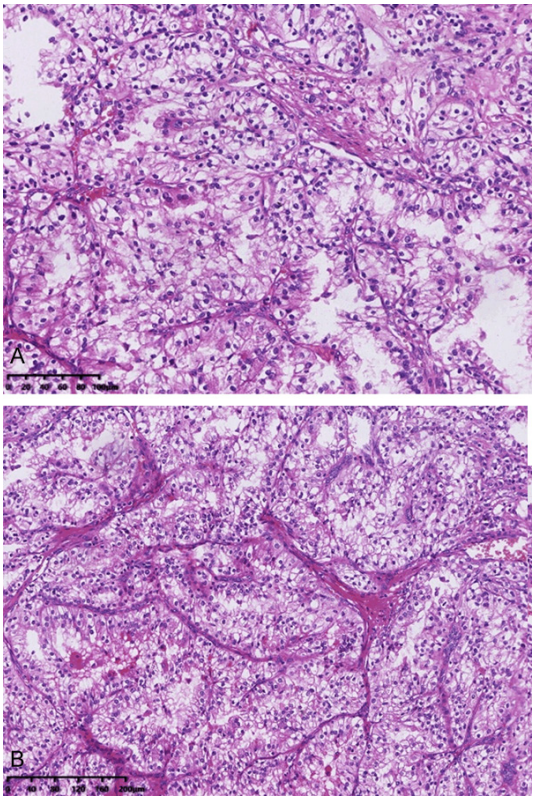

Figure 2. Pathologic diagnosis showed renal clear cell carcinoma.

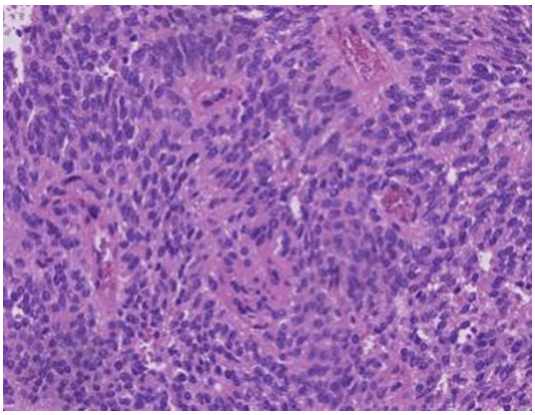

Figure 3. The tumor cells are columnar or cuboids that closely surround the blood vessels and are arranged in pseudo chrysanthemum-shaped.

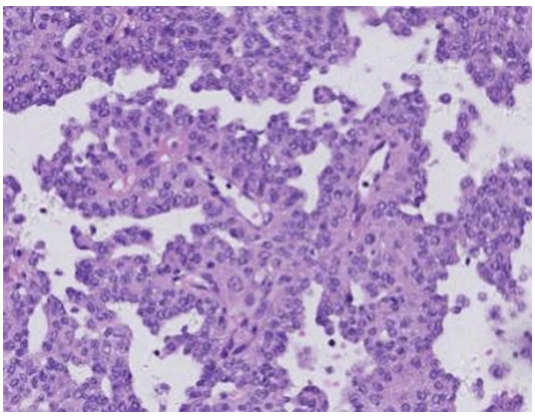

Figure 4. Photomicrograph of the tumor showing sheets of tumor cells arranged in papillary pattern and a papilla with fibrovascular core.

On the basis of its morphologic and immunohistochemical features, the tumor was diagnosed as papillary meningioma. The columnar or cuboid tumor cells closely surrounded the blood vessels and arranged in pseudo chrysanthemum-shaped (**Figure 3**) or papillary (**Figure 4**). The elongated cytoplasmic and nuclear protuberance extended to the vascular wall. Foamy nucleolus was present and the mitotic figures were evident. The tumor cells were relatively dense and characterized by abundant cytoplasm, coarse granular chromatin and round or oval nucleus. There were eosinophil and focal necrosis between tumor cells and blood vessels. The tumor invaded the peripheral brain parenchyma and some of the tumor cells showed significant meningothelial differentiation (**Figure 5**). Large pieces of necrosis and

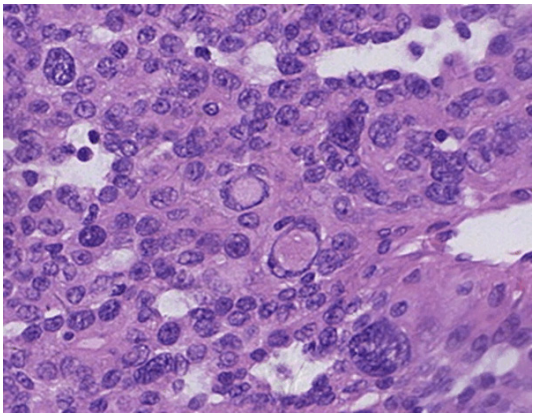

Figure 5. Tumor cell was large, showed foamy nucleolus and light cytoplasmic staining.

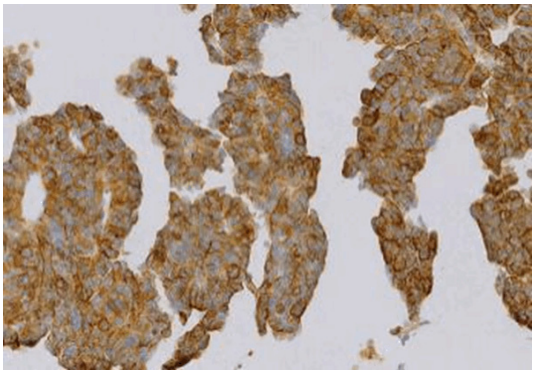

Figure 6. On immunohistochemistry the tumor cells stained positive with EMA.

extensive hemorrhage were observed in the tumor tissues and some tumor cells were nested or scattered in distribution. Immunohistochemically, the tumor cells were positive for Vimentin (**Figure 6**), EMA (**Figure 7**), CD-10, CK and 30% tumor cells positive for Ki-67. The tumor cells were negative for GFAR, S-100, PR, RCC, TTF-1.

Discussion

The clinical symptoms of PM are various, including headaches, seizures, dizziness, hemiplegia, vomiting, tinnitus, and visual impairment. These presenting symptoms depend on the location of the mass, the tissues it impinges on, and the vessels it obstructs. Unlike other subtypes of meningioma, the time for PM's early treatment is relatively short, which may be related to its aggressive biological behavior. According to current reports, PM have been discovered at the sagittal sinus and cerebral convexity frequently [2]. Recent studies report that PM occurred in the posterior cranial fossa, jugular foramen, and oculomotor nerve [4-6]. Jiang and colleagues [7] reported that PM also occurred in the brainstem, which exhibited high mortality. PM is prone to metastasis. Lung or bones are the most favored site of PM extracranial metastases [8].

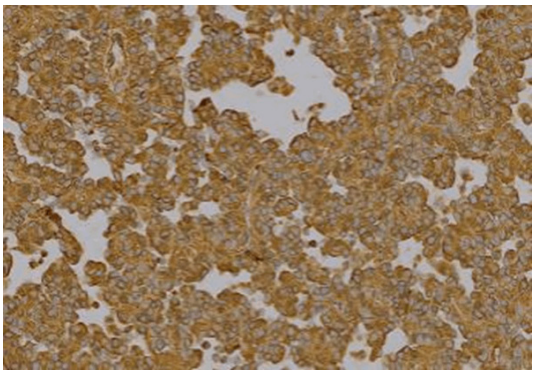

Figure 7. On immunohistochemistry the tumor cells stained positive with Vimentin.

Wang el at. [9] reported that gross total tumor restriction seems to reduce the risk of recurrence and mortality. Postoperative radiotherapy was also recommended to be used as an essential therapeutic schedule for the patients with papillary meningioma [10, 11]. PM is an aggressive type of meningioma with rapid progression and poor prognosis. Therefore, long-term postoperative follow up is critical to understanding the meningioma's aggressive behaviors. The available research shows that the tumors were mostly located in the cerebral convexity and showed irregular margins, heterogeneous enhancement, and severe peritumoral brain edema on preoperative images [9]. MIB-1 labeling index might predict the recurrence. High MIB-1 labeling indices indicate aggressive behavior of the papillary meningioma [12]. But the relationship between MIB-1 labeling index and progression free survival is not clear. Published reports from this research [9] indicate that the papillary meningioma recurrence and mortality was only correlated with the existence of intertumoral necrosis and incomplete resection. Papillary meningioma is an aggressive variant of meningioma and usually display aggressive clinical behavior. Early diagnosis of PM could significantly reduce the progression

of subsequent disease invasion and mortality [13]. Due to the rarity of PM, there is no standard treatment plan for the primary PM in the worldwide context. Some investigators recommend complete tumor resection and postoperative radiotherapy [7, 14]. Stereotactic radiotherapy could be utilized as a substitution choice for patients with asymptomatic recurrence or high-risk surgery. Mattozo [15] performed a statistical analysis of the efficacy of stereotactic radiotherapy in patients with asymptomatic recurrent meningioma. The results showed that stereotactic radiotherapy can effectively control the growth of grade I and II meningiomas. However, the efficacy of stereotactic radiotherapy in grade III meningioma is poor. The relationship among the three tumors in this case is unknown. It is speculated that genetic abnormality may be one of the reasons. If possible, we will study it further.

Differential diagnosis

(1) Papillary adenocarcinoma: It is commonly from lung cancer metastasis, with a more obvious papillary structure, greater cell atypicality, more mitotic figures. Immunohistochemically, EMA and CK were positive, but negative for Vimentin. (2) Papillary ependymoma: The tumor cells are arranged in pseudo chrysanthemum-shaped, and have single cell morphology, small atypia. The mitotic figures are rare. Immunohistochemically, D2-40 nuclear and GFAP were positive. (3) Astroblastoma: The tumor boundary is clear. The tumor cells of the single-stage cell are arranged radially, papillary or band-shaped around the blood vessel. The nucleus is round or oval, and the atypia is small. The common blood vessel wall is transparent and denatured. Immunohistochemically, S-100, Vimentin, GFAP were positive. (4) Choroid plexus papilloma: The papillary structure is more pronounced and diffuse, the tumor cells are cubic or columnar, the nucleus is round or oval, the atypia is small, and nuclear division is rare. Immunohistochemically, CK, Vimentin were positive, GFAP can be positive, the most important feature is EMA negative.

Disclosure of conflict of interest

None.

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