Case Report

The Diagnostic Odyssey: Papillary Meningioma - A Rare and Aggressive Entity

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Abstract

Background: Papillary meningioma (PM), a rare subtype, accounts for less than 1% of all meningiomas, has a high potential for metastasis, a higher rate of recurrence, and a worse prognosis than other types of meningiomas, categorized as WHO Grade 3 malignant tumors.

Case details: A 39-year-old male came with complaints of swelling and proptosis of the left eye, left-sided hemicranial headache for the last two months, and blurring of vision for the last month. On MRI scan, a likely diagnosis of hemangiopericytoma with a differential diagnosis of metastatic deposits was given. Excision of the eye swelling was done, and histopathological examination revealed the diagnosis of papillary meningioma (histological grade 3), which was further confirmed on immunohistochemistry.

Conclusion: Papillary meningioma is an exceptional variant of meningioma with an increased incidence of local recurrences and potential for distant metastasis. Its timely recognition may have important implications for management as well as prognostication in order to reduce morbidity and mortality.

Keywords:

Papillary meningioma, meningioma, aggressive malignant variant, rare variant, CNS tumors

Introduction

Meningiomas, the most common intracranial tumors, constitute 20% to 36% of all primary cerebral tumors and are derived from arachnoid cap cells [1,2]. The 2021 WHO Classification of CNS Tumors categorizes meningiomas into three grades based on histopathological and molecular features. WHO Grade 1 (benign) includes meningothelial, fibrous (fibroblastic), transitional (mixed), psammomatous, angiomatous, microcystic, secretory, lymphoplasmacyte-rich, and metaplastic meningiomas. WHO Grade 2 (atypical) comprises chordoid, clear cell, and atypical meningiomas. WHO Grade 3 (anaplastic/malignant) includes rhabdoid, papillary, and anaplastic (malignant) meningiomas. The classification considers specific criteria such as mitotic activity, brain invasion, and histological patterns, which correlate with tumor aggressiveness and prognosis [7].

According to the World Health Organization (WHO) classification of CNS tumors (2021), papillary meningiomas (PM) are a specific form of meningioma that have a very low overall incidence and are classified as grade 3 malignant tumors [1]. Histologically, the majority of the tumor exhibits a pseudo-papillary pattern, characterized by the presence of tumor cells around thin-walled blood vessels in a perivascular pseudo-rosette pattern, along with areas of necrosis. Rhabdoid cytology and papillary architecture are sometimes found together in meningiomas, and this combination has been linked to much more aggressive histopathological and clinical behavior [1].

Papillary meningiomas have been reported in both children and adults. These tumors are commonly associated with peritumoral edema, bone hyperostosis or destruction, and sometimes cyst formation. Papillary meningiomas typically exhibit aggressive behavior, including brain invasion, recurrence, and metastasis.

It is crucial to note that other meningioma variants can also show papillary architecture on histopathology. However, the histopathological diagnosis of PM must only be made when the entire mass exhibits this specific histological architecture. It can also be quite challenging to distinguish this type of meningioma from high-grade glial tumors [1,2].

In this article, we report the histopathological findings of a rare case of papillary meningioma in a 39-year-old male.

Case Report

A male patient in his 30s presented with complaints of swelling and proptosis of the left eye, left-sided hemicranial headache for the last two months, and blurring of vision in the left eye for the last month. The patient had been apparently well two months prior. No history of loss of smell, vision, seizures, or consciousness was noted. The patient was a chronic smoker and alcoholic. There was no past history of diabetes or hypertension. On examination of the left eye, a swelling was present over the lateral aspect, measuring 4 x 5 cm. The right eye was normal.

An MRI scan revealed a likely diagnosis of hemangiopericytoma, with a close differential diagnosis of metastatic deposits. Total removal of the tumor mass was accomplished after a left pterional craniotomy, and the specimen was sent for histopathological examination.

Gross examination of the resected tumor revealed multiple irregular tissue fragments, the largest measuring 5 x 3 cm and the smallest 0.5 x 0.2 cm. Histopathological examination revealed tumor tissue arranged in papillary and pseudo-papillary patterns (Figure 1). Individual tumor cells were round to oval, with a high nuclear-to-cytoplasmic ratio, pleomorphic nuclei, coarse nuclear chromatin, and prominent nucleoli. The cytoplasm was scant and eosinophilic (Figure 2). The pseudo-papillary pattern, comprising tumor cells clinging to blood vessels in a perivascular pseudo-rosette pattern, and a few meningothelial whorls were noted (Figure 3). A few rhabdoid cells were present, showing large pleomorphic nuclei with prominent nucleoli and abundant eosinophilic cytoplasm. The tumor tissue also invaded adjacent skeletal muscle bundles. A diagnosis of papillary meningioma (histological grade 3) was made based on histopathology.

Immunohistochemical staining for epithelial membrane antigen (EMA), vimentin, GFAP, and Ki-67 was performed. The tumor cells were immunohistochemically positive for EMA (Figure 4) and vimentin, while negative for GFAP. The Ki-67 index was 72–75%. Immunohistochemistry further confirmed the diagnosis of papillary meningioma, WHO grade 3.

The histological identification of papillary meningioma represents the primary diagnostic challenge. Papillary ependymoma, metastatic papillary adenocarcinoma, and choroid plexus papilloma are among the possible differential diagnoses on

histopathology.

The present case was diagnosed as papillary meningioma (histological grade 3) due to the presence of focal meningothelial whorls. Further, immunohistochemistry showed tumor cell expression of EMA, Ki-67, and vimentin. Unfortunately, the patient expired three months later.

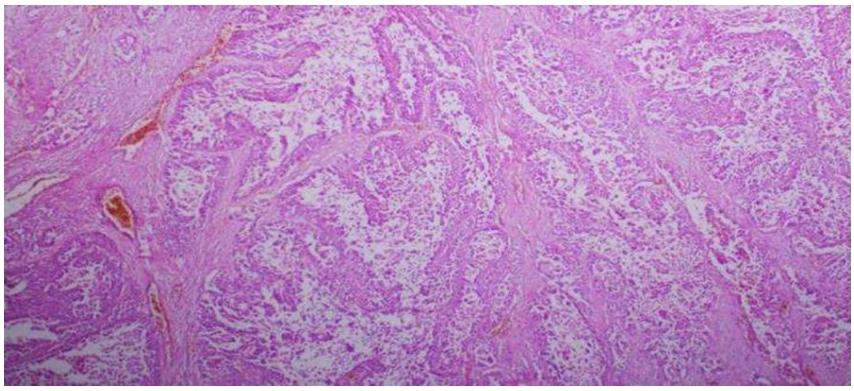

Figure 1: Photomicrograph showing tumor tissue arranged in a papillary and pseudopapillary pattern (H&E, 4x).

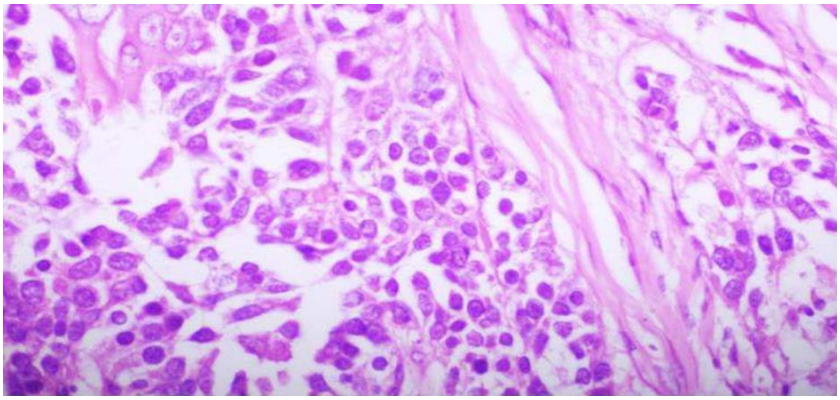

Figure 2: Photomicrograph showing round to oval tumor cells with pleomorphic nuclei, coarse nuclear chromatin, prominent nucleoli, and scant eosinophilic cytoplasm (H&E, 40x).

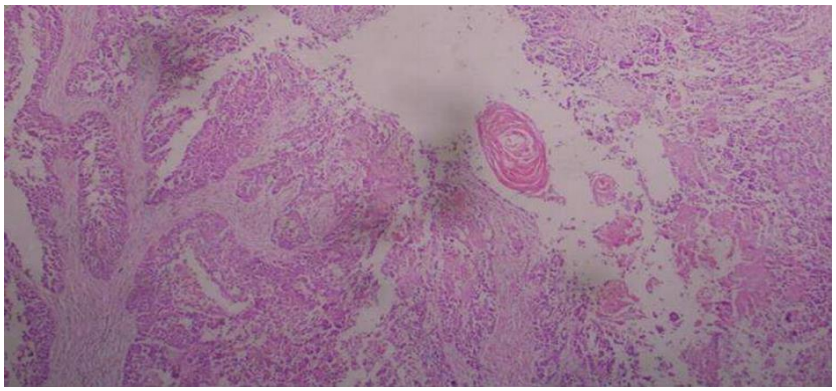

Figure 3: Photomicrograph showing a few meningothelial whorls (H&E, 4x)

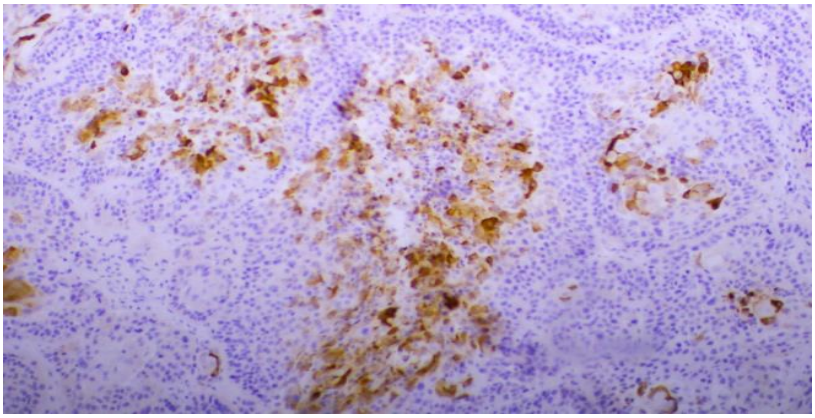

Figure 4: Photomicrograph showing immunohistochemical expression of EMA in tumor cells (10x).

Discussion

Papillary meningioma (PM), a malignant variant of meningioma, accounting for 1.0–2.5% of all meningiomas, was identified for the first time by Cushing and Eisenhardt in 1938 [3]. They discovered a meningioma with a papillary appearance that had lung metastases and intracerebral recurrence.

When Ludwin et al. [4] analyzed the histopathological features of 17 patients with papillary meningioma, they found that, in 8/17 (47%) cases, PM was more prevalent in younger age groups than other meningioma variants. Mitoses, local recurrences, brain invasion, and extracranial metastases were reported in 7/17 (41%) cases, 10/17 (59%) cases, and 4/17 (23.5%) cases, respectively.

The above-mentioned analysis was supported by Brignolio and Favario [5], who evaluated eight patients with papillary meningioma and found that the formation of a papillary architecture in a meningioma is related to both histopathological and clinical aggressiveness.

Similarly, Radhakrishnan et al. [6] analyzed six adult cases of papillary meningioma and found that the majority of the cases had histopathological findings suggestive of infiltration of the bone and brain.

There is no established consensus regarding how papillary meningiomas should be managed due to their rarity. However, extensive surgical excision combined with postoperative radiotherapy has become the accepted standard of care.

In the above-reported patient, a craniotomy was performed, total resection of the tumor was achieved, and the diagnosis of papillary meningioma (histological grade 3) was confirmed through histological and immunohistochemical examination.

Conclusion

Papillary meningioma (PM) is a rare and unique type of meningioma. It must be distinguished from other intracranial tumors showing a papillary appearance on the basis of histopathology. PM is aggressive, usually exhibits invasion of the bone and parenchyma, and is capable of metastasizing outside the cranium. Local or distant metastases can be prevented if they are identified in a timely manner, thereby decreasing the mortality and morbidity associated with PM.

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