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Metastatic rhabdoid meningioma of the parotid – Mimicking primary salivary gland neoplasm

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ABSTRACT

INTRODUCTION: Tumors involving the parotid are predominantly primary with metastatic lesions forming a miniscule population. Meningioma metastasizing to the parotid is extremely rare and hence can often be mistaken for the more common primary salivary gland neoplasms.

PRESENTATION OF CASE: A 59-year-old male presented with a swelling in the left parotid region. Fine needle aspiration cytology was suggestive of myoepithelial predominant pleomorphic adenoma. A superficial parotidectomy performed revealed a tumor composed of rhabdoid cells with abundant finely granular eosinophilic cytoplasm raising a possibility of myoepithelioma. Immunohistochemistry for myoepithelial markers was negative. A critical review elicited a history of surgical excision of a recurrent rhabdoid meningioma twice. A possibility of metastasis was considered and a second panel of immunomarkers demonstrated vimentin and epithelial membrane antigen positivity. Neuroimaging studies demonstrated a space occupying lesion in the frontal lobe suggestive of a recurrent/residual tumor. In view of the history, neuroradiology, histopathology and immunohistochemistry, a final diagnosis of metastatic rhabdoid meningioma to the parotid was rendered.

DISCUSSION: Morphologically, metastatic rhabdoid meningioma may mimic a primary or metastatic carcinoma, melanoma and sarcoma. Accurate diagnosis can be made by careful clinical evaluation and histopathological examination of the tumor. These tumors are composed of rhabdomyoblast like cells with abundant eosinophilic cytoplasm. The present case demonstrated characteristic histopathological features confirmed by immunohistochemistry.

CONCLUSION: Rhabdoid meningioma is an aggressive tumor with a high propensity to recur and metastasize. The present case highlights the importance of clinical, radiological and histopathological correlation to accurately diagnose these rare entities.

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1. Introduction

Meningiomas are rare tumors arising from the dura mater and are the most common neoplasm of the central nervous system. They are categorized as by the World Health Organization (2007) as WHO grade 1, 2 (atypical) and 3 (anaplastic). Grade 2 and 3 tumors have an aggressive behavior. Rhabdoid meningioma is an extremely rare subtype of WHO grade 3 meningiomas characterized by the presence of cells resembling rhabdomyoblasts and exhibiting Vimentin immunoreactivity. These tumors are associated with a poor prognosis and a high rate of recurrence [1,2]. Meningiomas rarely metastasize via the haematogenous route and

the lung and pleura are the preferred sites [8]. We describe a rare case of recurrent and metastatic rhabdoid meningioma affecting the parotid.

2. Case

A 59-year old male presented with a gradually progressive, painless swelling in the left parotid region of 1 year duration. Clinically, the lump was firm with a distinct boundary producing structural distortion of left side of the face. The patient had previously been operated for rhabdoid meningioma 7 years back followed by two cycles of radiotherapy. He required repeat surgery after 2 years for recurrence. Fine needle aspiration cytology of the parotid lesion was reported as benign pleomorphic adenoma. A T2 weighted coronal cut (magnetic resonance image) of the brain (MRI) showed a residual lesion in the left frontal lobe with a penumbra of edema (Fig. 1). A conservative superficial parotidectomy was performed.

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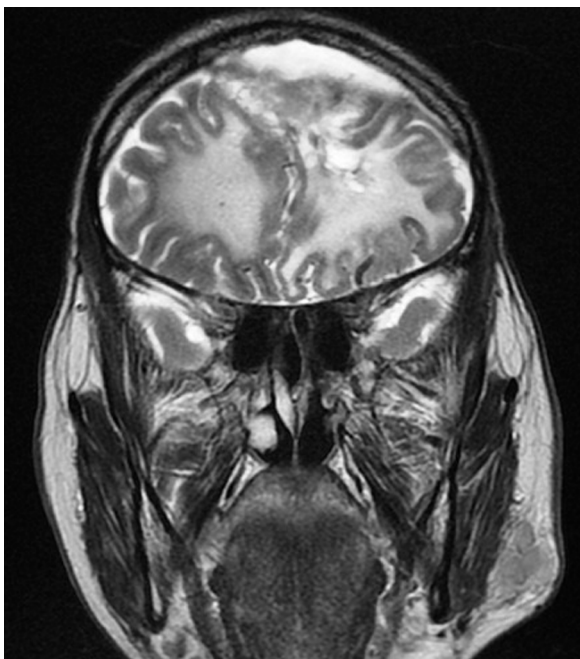


Fig. 1. MRI Brain (T2w image) scan shows residual tumor.

Microscopy showed a tumor composed of syncytial sheets with globules, pseudo-acinar and capillary patterns of oncocytoid polygonal and spindle cells with abundant finely granular eosinophilic cytoplasm and vesicular nuclei with pseudo-inclusions. Mitoses were rare and necrosis was absent (Fig. 2). Immunohistochemistry revealed a strong and diffuse positivity for vimentin (Fig. 3a) and EMA (Fig. 3b) with a focal reactivity for cytokeratin. A final diagnosis of metastatic rhabdoid meningioma was given. He is asymptomatic on follow up and has denied any further management of the residual tumor in the central nervous system.

3. Discussion

Meningiomas account for 15–20% of all primary tumors of the central nervous system. These tumors display varied morphological features and are described as typical (grade 1) lesions, atypical including clear cell and chordoid variants (grade 2) and anaplastic which encompasses papillary and rhabdoid meningiomas. Rhabdoid meningiomas are uncommon and are known to have a very poor prognosis with a median survival of less than three years and recurrence rates as high as 50–78% [4]. Extraneuroaxial metastasis and brain invasion have also been reported [3,4,6].

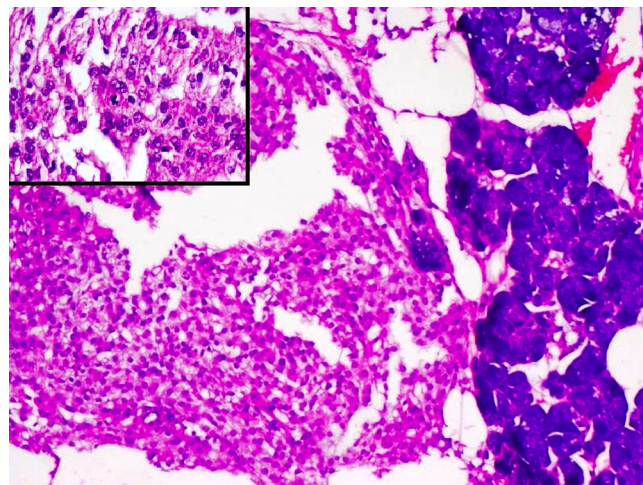


Fig. 2. Microphotograph showing sheets of oncocytoid cells with occasional mitosis and adjacent normal salivary gland parenchyma (H&E, 20 \times), Inset (H&E, 40 \times).

It has been estimated that only 0.1% of meningiomas metastasize. Metastasis is more commonly seen in patients who have undergone craniotomy. The common sites of metastasis are lung, pleura, skeletal system and liver. Metastasis to the parotid is rare and only few cases have been described [5,7].

Morphologically, metastatic rhabdoid meningioma may mimic a primary or metastatic carcinoma, melanoma and sarcoma. Accurate diagnosis can be made by careful clinical evaluation and histopathological examination of the tumor. These tumors are composed of rhabdomyoblast like cells with abundant eosinophilic cytoplasm expressing markers of intermediate filaments such as vimentin and cytokeratin [2]. The present case demonstrated characteristic histopathological features confirmed by immunohistochemistry.

Meningiomas are usually benign and it is usually the location and approach to the tumor that plays the pivotal role in management. Surgical excision is the treatment for this disease entity, however, anaplastic meningiomas require adjunct radiotherapy as well. With this approach, locoregional recurrence is still between 50% and 70%, depending on resection status. Few smaller studies have shown that postoperative radiotherapy can increase progression-free survival, which translates into increased 5-year survival. However, meningiomas are known to be radioresistant, and radiation doses of 60 Gy or higher have been shown to be necessary for tumor control [8].

To conclude, rhabdoid meningioma is an aggressive variant of meningioma with a high propensity to recur and metastasize. These tumors can metastasize to any organ system, lung being the most

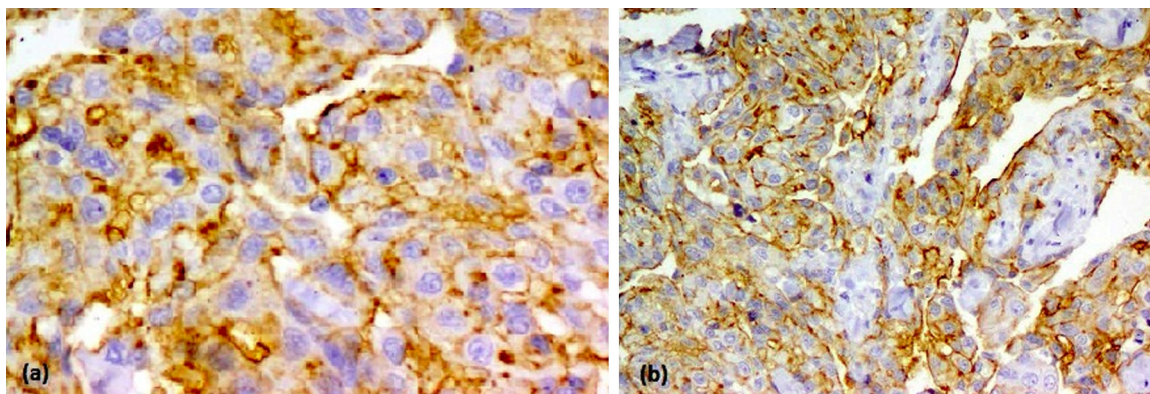


Fig. 3. Microphotograph showing immunoreactivity to Vimentin (a) and EMA (b).

common site. Metastasis to the parotid is a rare occurrence. The present case emphasizes the need for a high degree of clinical suspicion for early detection and treatment of metastasis in such unusual locations.

Conflict of interests

No conflict of interests.

Funding

None.

Ethical approval

Written informed consent was taken from the patient.

Author contributions

Dr. Yashdeep Sarma and Dr. Rajesh Nair - Study design, data and writing Dr. Pradeep Kumar Tripathi - Data collection Dr. Bhavna Nayal, Dr. Summet Kaur Dil - Pathology Dr. Vinod Kumar - Final editing and revisions.

Key learning points

- Neuroimaging studies demonstrated a space-occupying lesion in the frontal lobe suggestive of a recurrent/residual tumour. In view of the history, neuroradiology, histopathology and immunohistochemistry, a final diagnosis of metastatic rhabdoid meningioma to the parotid was rendered.
- Morphologically, metastatic rhabdoid meningioma may mimic a primary or metastatic carcinoma, melanoma and sarcoma. Accurate diagnosis can be made by careful clinical evaluation and histopathological examination of the tumour.

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