Dear Editor,

Hemangiopericytoma is a rare mesenchymal tumor derived from pericytes of Zimmerman.[1] It accounts for <1% of all central nervous system (CNS) tumors. Recurrences in meningeal hemangiopericytoma may occur either at the primary site or at distant sites. This may be a late event occurring 5-10 years after initial detection of the disease. The present case report highlights the rarity of this tumor and the incidental detection of liver metastases 8 years after treatment for the primary tumor.

A 54-year-old man presented in 2004 with headache and abnormal movements of the right upper limb of 3 weeks duration. Magnetic resonance imaging (MRI) brain was suggestive of parasagittal meningioma [Figure 1a and b]. Left parietal craniotomy was done [Figure 1c]. Biopsy revealed grade II hemangiopericytoma. Postoperative radiotherapy to the brain to a dose of 54 Gy was given. Eight years later he was detected to have liver lesions on ultrasound abdomen on a routine health check-up. Contrast-enhanced computed tomography abdomen was suggestive of liver metastases [Figure 2]. Fine-needle aspiration cytology of liver lesions was positive for CD34 in vessels of neoplastic fragments confirming metastases from meningeal hemangiopericytoma [Figure 3a-e]. He then received palliative chemotherapy with ifosfamide and doxorubicin. Interim CT scan indicated partial response. He has received six cycles of chemotherapy and is asymptomatic at 10 months follow up.

Hemangiopericytomas were described in 1942 by Stout and Murray.[2] Begg and Garret first described meningeal hemangiopericytoma and proposed the term hemangiopericytoma.[3] The most common sites of involvement are the lower extremity, pelvis, and retroperitoneum.[4] Meninges are uncommon sites of presentation. Intracranial hemangiopericytomas were earlier classified as angioblastic meningioma, but now they are considered a separate entity with different biologic characteristics and immunohistochemistry profile as compared to meningioma.

Headache is the most common symptom. A few patients may present with features of raised intracranial tension due to mass effect or hydrocephalus. Extensive disease or local recurrences may present with cranial nerve palsies. Hemangiopericytomas are dural based and show white matter buckling. Guthrie et al.,[5] reported the common sites of occurrence of primary meningeal hemangiopericytoma as parasagittal, posterior fossa, and tentorium.

Hemangiopericytomas are dural-based, vascular, extra-axial masses. These tumors lack hyperostosis and calcification, which is a feature observed in meningioma.[6] Hemangiopericytomas are characterized by a narrow based dural attachment unlike meningiomas that have a broad based attachment.[7] They typically appear as hyperdense heterogeneous tumors on unenhanced CT scans and isointense on T1-and T2-weighted images and show heterogeneous enhancement on contrast.

Histopathology reveals spindle-shaped cells with multiple vascular spaces. There is a high degree of cellularity. High power view commonly shows vesicular nuclei with nucleoli and six to eight mitotic figures. Immunohistochemistry is positive for vimentin, S-100, smooth muscle actin, muscle-specific actin, CD34, and factor XIIIa. Lesional cells do not stain for factor VIII. Presence of mitoses, necrosis, and vascular invasion predicts aggressive behavior and poor prognosis. Certain chromosomal abnormalities such as t (12; 19) and t (13; 22) have been reported.[8]

Local recurrences are common and vary from 45 to 90% in most case series. Most of the recurrences occur at the primary tumor site. In a study by Galanis et al.,[9] 59% recurrences occurred at the primary tumor site, 34% at
both primary and distant CNS sites, and 7% had diffuse leptomeningeal spread.[8] Kim et al.,[9] in their review of 31 cases of primary meningeal hemangiopericytomas reported 91.7% of recurrences at the primary tumor site. Recurrences in meningeal hemangiopericytoma are a late event occurring 5-10 years after initial detection of the disease. In a study by Brunori et al.,[10] the time to first recurrence was 7 years. Due to the late recurrences close and prolonged follow-up is warranted.

Meningeal hemangiopericytoma has a propensity to metastasize to distant organs. Some of the common sites of metastases are liver, lungs, bone, kidney, and pancreas. Galanis et al.,[8] noted that 82% metastases occur to the bone, whereas in 41% metastases spread to the liver. Longer duration from time to first treatment increases the chances of metastases; hence, close and regular follow-up is necessary. There have been reports of metastases occurring 16 years after primary treatment. Guthrie et al., demonstrated that the 5, 10, 15-year probability of developing metastases was 13%, 33%, and 64%, respectively.[5] In our case, the metastases to liver was detected 8 years after the primary presentation. In a report by Brunori et al.,[10] lung, liver, and kidney metastases were detected at autopsy in a patient who survived 15 years after the first operation.

Surgery is the mainstay of treatment and the tumor must be excised whenever possible. Complete excision has a favorable effect on recurrence and survival. It improves survival and reduces the chance of first local recurrence. Kim et al., documented that complete excision at the first operation extended the duration before first recurrence from 43 months to 111 months.[8]

Preoperative radiotherapy is indicated in high-risk surgical cases and surgically inaccessible tumors. Uemura et al.,[11] reported that in patients who received preoperative radiotherapy, removal of tumor was easier with less hemorrhage. Histological examination postradiation showed reduction in tumor cells with hyaline degeneration.[11]

Postoperative radiotherapy is associated with better survival. This was done in our case too in which postoperative radiotherapy was given. Guthrie et al., have recommended prophylactic postoperative radiotherapy after complete tumor excision to reduce local recurrence rates.[5] Radiotherapy extended the time before first recurrence from 34 months to 75 months and extended survival from 62 to 92 months. A dose of 50 Gy is recommended after first tumor excision. The role of chemotherapy is more in the palliative setting than as primary treatment. Treatment regimens commonly include actinomycin D, adriamycin, vincristine, and cyclophosphamide and methotrexate. Response rates to chemotherapy range between 30-40% and less than 50% are disease free at 5 years after diagnosis of metastasis.[12]
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