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Case Report:

Meningeal Hemangiopericytoma of Brain: Role of Radiation Therapy

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Abstract: Hemangiopericytoma is an uncommon vascular tumour. Complete surgical resection is treatment of choice. However, late local recurrences and distant extraneural metastases ranging from 12% to 57% are reported in literature after complete removal. Post operative radiotherapy is indicated in unresectable or incompletely excised tumour. We present a case of meningeal hemangiopericytoma in a 60 years old female patient treated with surgery and adjuvant radiotherapy. There is no evidence of disease at primary site and no sign or symptoms of metastatic disease in the patient after three years.
Key Words: Brain tumor; Radiotherapy; Hemangiopericytoma

Introduction:
Hemangiopericytomas are an uncommon intracranial tumor of mesenchymal origin (MHPC) and constitute less than 1% of all central nervous system tumors.[1] They have been observed at the falx, occipital, spinal dura, tentorium and cerebellopontine angle.[2]
These tumors are usually present as dura-based masses and clinically indistinguishable from meningiomas. MHPCs are characterized by locally aggressive biological behavior with tendency to locally recur and metastasize extra cranially.[3] MHPC occurs most frequently during the fifth decade.
Management of these tumors includes aggressive total excision. However, late local recurrences and distant extraneural metastases ranging from 12% to 57% are reported in literature after complete removal. Adjuvant radiotherapy is indicated in completely excised tumor with an aim to maximizing local tumor control.[4] The aim of this study was to evaluate the effectiveness of adjuvant radiotherapy and determine the outcome.

Case Report:
A 60 years old female patient present with weakness in limbs, associated with inability to walk, loss of sensation, disorientation and loss of memory of four months duration. There were also complaints of loss of bowel and bladder control of two months duration. There was no history of seizers and trauma. General and physical examination were normal. Computed tomography of brain showed a large hypodense area in left frontal, temporo -parietal and occipital regions. Right lateral ventricle was dilated with periventricular edema and left lateral ventricle and third ventricle were compressed.[Fig 1,2] CECT of chest and abdomen were normal and there was no evidence of bone metastases on radioisotope bone scan examination.
Patient underwent left parieto-occipital craniotomy and excision of tumor.(Fig 3) The histopathological examination of growth revealed Meningeal Hemangiopericytoma. Reticulin stain showed staghorn vessels with increased perivascular reticulin. On immunohistochemistry EMA and PR were negative, CD 34 and VWF were positive in vascular channels.(Fig 4-7) Patient was treated with adjuvant external beam radiotherapy with dose of 56 Gy in 30 fractions over 6 weeks after six weeks of surgery. Patient is disease free three years after treatment.

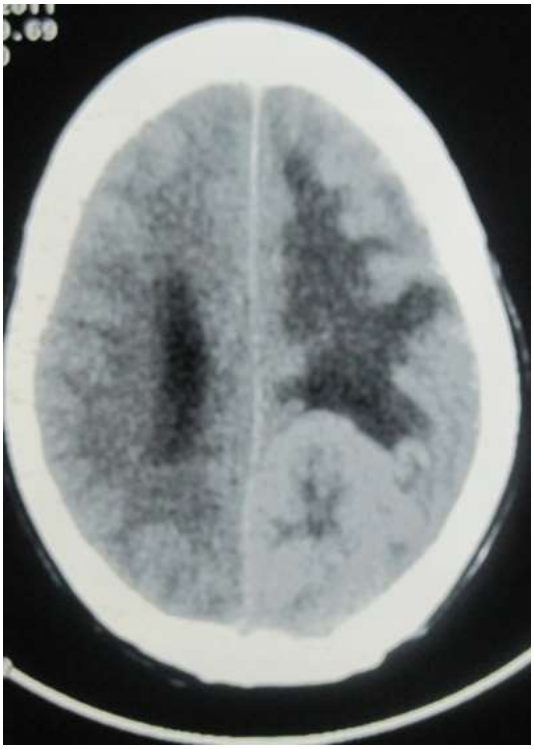
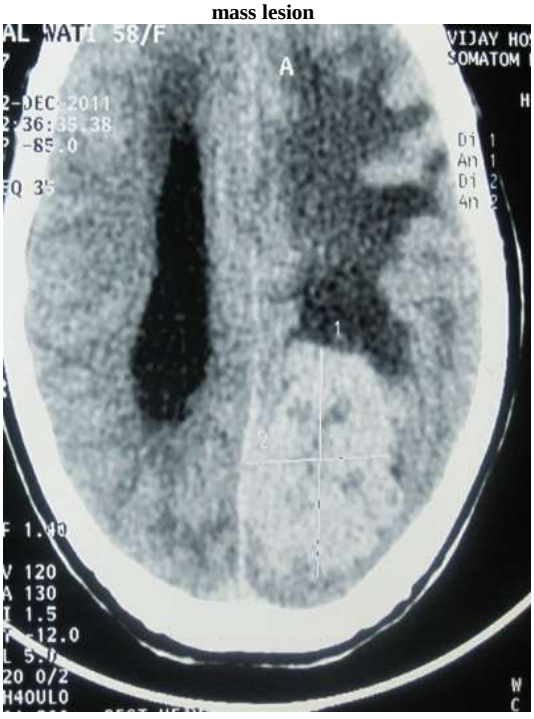


Fig 1, 2: Pre operative CECT scan showing intracranial mass lesion



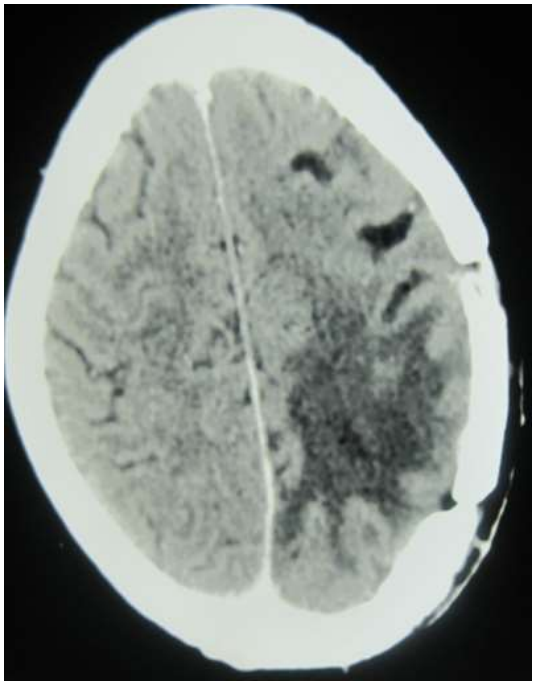


Fig 3: Post operative CT scan shwoing tumor clearance

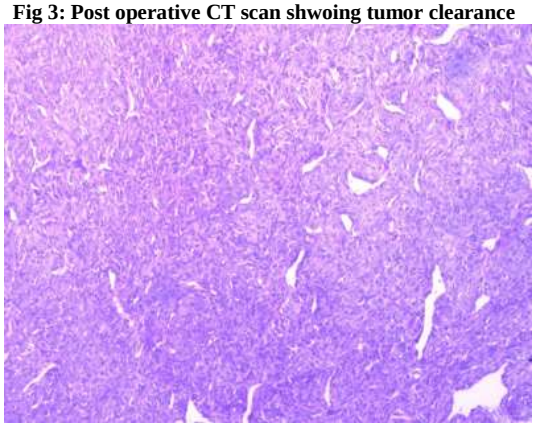


Fig 4: Photomicrograph showing oval to spindle shaped tumor cells along with staghorn vessels.

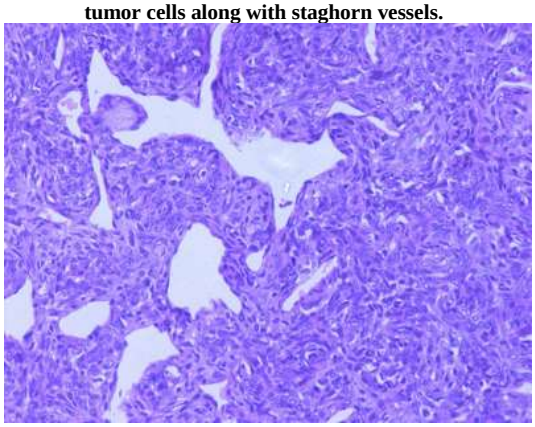


Fig 5: Photomicrograph showing staghorn vessels with increased perivascular reticulin

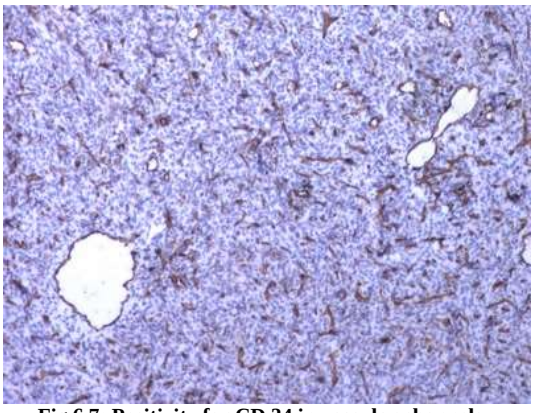
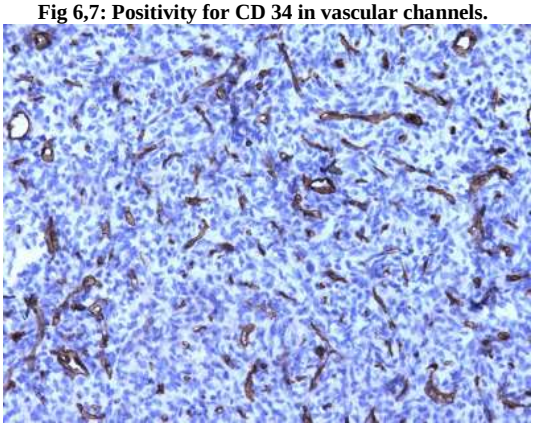


Fig 6,7: Positivity for CD 34 in vascular channels.



Discussion:

Hemangiopericytoma is a rare soft tissue tumor. Extracranial hemangiopericytoma was first reported by Stout and Murray in 1942.[5] Clinically these lesions are indistinguishable from meningiomas. The World Health Organization (WHO) classification of brain tumors has considered them as 'mesenchymal, non-meningothelial tumors.[6] Aggressive surgical resection remains cornerstone of treatment. However, complete excision is sometimes difficult to achieve due to high vascularity and tendency of these tumours to grow along sinus.[2]

Traditionally radiotherapy had been used for unresectable tumor or recurrent tumors. Now, Post operative radiotherapy has a well established role in management of these tumours and associated with increased survival. Administration of post operative radiotherapy is most strongly related to the overall prognosis.[7] Radiation responses are dose dependent, with > 50 Gray (50 Gy–60Gy) providing superior long-term disease-free survival.[8]

In addition, it has been reported that adjuvant post operative radiotherapy can be beneficial in reducing local recurrence rate that also be resulted in prolongation of disease free and overall survival. Other modalities such as helical tomography and stereotactic radiosurgery are indicated in recurrent and metastatic disease.

However, aggressive biological behaviour and a tendency of local recurrence after complete excision of tumor are characteristic features of hemangiopericytoma. Moreover these tumors also have the unique characteristic of metastases extracranially, mainly to bone, liver and lung. Guthrie et al. demonstrated increasing metastatic frequency, reporting 5-, 10- and 15-year metastasis rates of 13, 33 and 64%, respectively.[9] There is limited role of chemotherapy in metastatic setting as reported in clinical series.

To evaluate the outcome the long term extensive follow up is mandatory that included routine investigation such as liver function test, kidney function test, and Chest X ray, Ultrasound examination of abdomen and pelvis and radionuclide bone scan.

Conclusion:

The patient was treated with adjuvant external beam radiotherapy with dose 56 Gy in 30 fractions. Postoperative radiotherapy has resulted in a significantly better local control which has a major impact on the survival of primary meningeal hemangiopericytoma. Radiation therapy is a recommended modality as adjuvant treatment after complete surgical removal of this tumor so as to achieve high probability of an increased recurrence – free interval and overall survival. Extensive long term follow up also recommended ruling out local recurrences and late extracranial metastases.

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