CASE REPORT

Spindle cell Haemangioendothelioma, A rare Vascular Tumour with Presentation as Intraparenchymal Haemorrhage, A Case Report

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ABSTRACT

Spindle cell hemangioendothelioma is a rare vascular tumor featured by a histologic manifestation intermediate between benign hemangioma and malignant angiosarcoma. A 45 year old female patient presented with loss of consciousness and left sided weakness with CT brain suggestive of hematoma in right gangliocapsular region. A computed tomography angiography brain revealed asuspected aneurysm in right internal carotid artery in clinoid segment. Right pterional craniotomy was done with removal of vascular tumoralongwith hematoma which was adhered to right internal carotid artery and clinoid process. The histopathologic report was suggestive of spindle cell hemangioendothelioma of the brain. It is hoped that our report will contribute further understanding of the neuropathology and natural history of this unusual tumor.

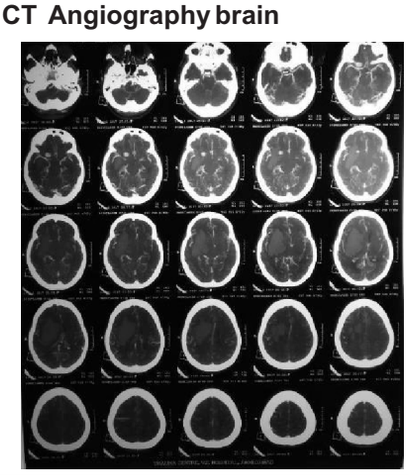
INTRODUCTION

Intracranial spindle cell hemangioendothelioma (HE) is a rare borderline angiomatous tumor of vascular origin and histologically intermediate of hemangioma and angiosarcoma. HE involving intracranial structures occasionally results in serious local compressive symptoms, including cranial nerve palsy or a potentially fatal increase in intracranial pressure. Despite the low proliferation indices, the clinical course of intracranial HE can be complicated.2A total resection is essential where possible, otherwise radiotherapy and/or chemotherapy are required.2 Pre-operative embolization of the feeding-artery is recommended. As intracranial HE is seldom encountered in clinical practice, considerable confusion exists with regard to its correct diagnosis and management.

CASE REPORT

A 45 year old female patient with history of cardiac disease presented with loss of consciousness and left sided weakness. On examination, patient was comatose with GCS: eye opening on painful stimuli and withdrawal response with no verbal response and left sided hemiparesis. Patient was intubated and put on ventilator support. Pupils were equal in size with normal reacting. All laboratory tests were within normal limits. A computed tomography brain revealed hematoma in right gangliocapsular region. Computed tomographic angiography (Figure1-3) was done. Based on the findings of a homogeneously enhanced, well-defined lesion in right internal carotid artery in clinoid segment, a provisional diagnosis of right ICA aneurysm was made. Right pterional craniotomy with removal of vascular tumor and hematoma which was adhered to right internal carotid artery and clinoid process was done. Tumour was approximately 0.9*0.9*0.7 cm sized, reddish brown in colour and soft in consistency . The histopathologic report was suggestive of spindle cell hemangioendothelioma of the brain (Figure 4). Patient expired on third post-operative day due to preexisting cardiac condition. As patient expired on third post operative day, Immunohistochemistry was not done.

Figure I : CT Angiography brain



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Figurell: CT Angiography brain

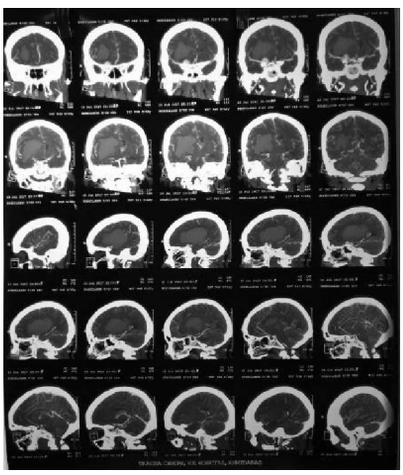


Figure III : CT Angiography brain

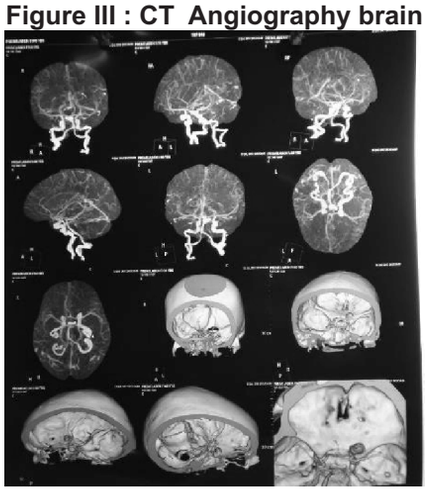
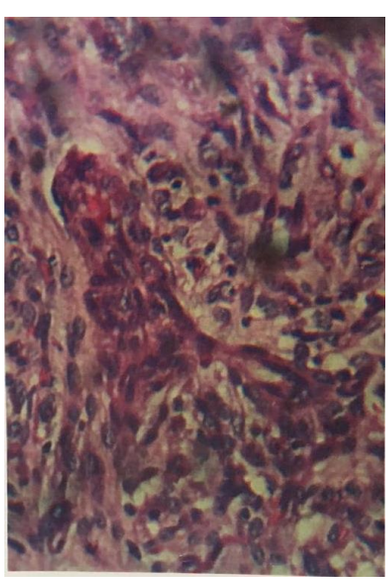


Figure IV : Histopathologic examination

(Spindle cell hemangioendothelioma)



DISCUSSION

Spindle cell hemangioendothelioma (SCH) is a rare vascular neoplasm that was described by Weiss and Enzinger.1Intracranial hemangioendothelioma accounts for <0.02% of all primary intracranial tumors, and it usually arise from brain, dura matter, spine and skull. It involve the other organs as lungs, liver, head and neck , bones and vessels, so always look for site other than cranium.2 Various clinical, radiological and pathological features have been sporadically reported.2,3Spindle cell hemangioendothelioma was first described as a low-grade sarcoma with histologic features intermediate between hemangioma and Kaposi's sarcoma.4 0verlapping of histological feature of tumour makes it difficult to classify into any single category.5 Distinct subtypes described are epithelioid hemangioendo-thelioma, spindle cell hemangioendothelioma, and malignant endovascular papillary angioendothelioma.

Intracranial HE are usually of epitheliod type. 7 Epithelioid hemangioendothelioma has been described in extradural intracranial locations but spindle cell hemangioendothelioma of the brain has been rarely reported.6 Spindle cell hemangioendothelioma is typically described as being composed of two cellular components.6,7 Vascular spaces lined by endothelial cells, had a tendency to digitate into the vascular lumens and separate cellular areas of spindle cells which are distinguished by the focal presence of rounded endothelial cells which form nests or line vascular channels. Both tumor cell types feature vacuolar changes in the cytoplasm of some cells. SCH is benign vascular neoplasm or malformation with local recurrences represents contiguous spread along vessel, rate is as high as 60%.8

HE can affect adults and children, with no clear gender preponderancepresents in the fourth and fifth decades. Clinical symptoms are usually depends on location of tumour and related to raised intracranial pressure, but rarely they can present as hematoma.9,10 The tumors often grew slowly with indolent clinical course, thus the period from the onset of the first symptom to the time of diagnosis is relatively long.

On imaging, tumour appear hyperintense (and/or heterogenous) on T2 and iso to hyperintense (and/or heterogenous) on T1. It shows uniform contrast enhancement on both CT and MRI. 3

Differential diagnosis for HE usually includes meningioma, hemangioma and angiosarcoma.12,13 Hemangioma are benign and shows honeycomb configuration and classic sunburst pattern, with a rapid enhancement at the early stage and homogeneous

enhancement after a delay.14 Primary CNS angiosarcoma is an extremely rare malignancy, with rapid growth and rapid onset on neurological symptomps. Imaging studies show a well-demarcated lesion of the cerebral hemisphere with avid post contrast enhancement. However, by contrast to HE, angiosarcomas usually present as a heterogeneous mass with significant vasogenic edema and intratumoral cyst formation.15,16

The prognosis and treatment of hemangioendo thelioma have not been well established. Essential of the treatment of SCH is total surgical resection whenever possible. Adjuvant therapy is required in a case with subtotal resection and higher grade on histology in form of chemotherapy, radiotherapy and vascular embolization. Hemangioendothelioma are moderately radiosensitive tumors. Local radiotherapy is also useful in treating inaccessible hemangioendothelioma with good long-term local control. Recurrence is usually rare after total resection.3

CONCLUSION

Spindle cell hemangioendothelioma is a low-grade tumor. HE is rare, but should be considered as a possible diagnosis when a tumor presents as a lobulated mass, with hemorrhage, signal voids of vessels, a heterogeneous appearance and delayed enhancement; these factors could potentially distinguish HE from other primary brain neoplasms. CT and MRI may be useful in providing an early and accurate diagnosis; each method is important due to the propensity of the tumor for abundant vascularization and low-grade malignant biological behavior.. Prognosis and treatment of hemangioendothelioma have not been well established. Involved-field radiotherapy is the only modality commonly used following incomplete tumor resection or tumor recurrence.

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