MEDICAL CHANNEL

Case Report

CAPILLARY HEMANGIOBLASTOMA

** Tariq N, **Sadiq S, ***Kehar SI*

Department of Clinical Pathology, Jinnah Post Graduate Medical Centre, Karachi.

ABSTRACT

Capillary hemangioblastoma refers to rare benign vascular neoplasm that arises almost exclusively in the central nervous system, seen commonly in females. We present a case of a 20 years old female. The clinical presentation was with raised intra cranial pressure.

Histologically the tumour was composed of large thin walled dilated blood vessels.

KEYWORDS: Capillary Hemangioblastoma Von Hippel-Lindau syndrome.

INTRODUCTION

Capillary hemangioblastoma refers to benign vascular neoplasm that arises almost exclusively in the central nervous system. According to the WHO classification of tumours of the nervous system, hemangioblastomas are classified as grade I tumours of uncertain origin [1&2]. These tumours arises from normal glial cells of the brain. These cells out number the neurons that conduct the impulses and serve to provide metabolic support to the neurons. In 1928 Cushing & Bailey coined the term "Hemangioblastomas" to describe tumours arising from the endothelial cells of the central nervous system [3&4].

SEX AND AGE DISTRIBUTION

Hemangioblastomas are common in the men than in women. In most clinical series, the male: female ratio is approximately 2:1 although hemangioblastomas may develop at any age, rarely affect children, and the usual age at diagnosis in 3rd and 5th decades.

Though histologically benign slow growing tumours that contain both solid & cystic components their development is often unfavorable due to high frequency of recurrence and multicentricity especially when occurring in a familial set up i.e. Von Hippel Lindau syndrome (VHL). VHL- disease is a heritable systemic syndrome that manifest in the CNS with multiple intra cranial and retinal hemangioblastomas. Although the surgical out come are similar for both sporadically occurring and VHL disease associated hamangioblastomas. Patients with VHL disease have a higher morbidity and mortality secondary to multifocal disease. [5,6,7]

These tumour account for 1.5-2.5 % of all intra cranial and 7-12% of posterior

Correspondence

DR. NAILA TARIQ
Incharge
Deptt. of Clinical Pathology
Jinnah Post Graduate Medical Centre, Karachi.

fossa tumours.

Hemangioblastoma are the most common primary adult intra axial posterior fossa tumour comprising 8-12% [8].

The second most common location of hemangioblastoma is the spinal cord [9,10,11,12] where the frequency ranges from 2-3% of primary spinal cord neoplasm's to 7-11% of spinal cord tumours. These tumours also occurs in other locations such as the supratentorial compartment,

[13,14] Optic nerve [15,16,17,18] Peripheral nerves [19], or the soft tissues of extremities is extremely rare site [20]. Surgery is the standard modality of treatment though their hypervascularity and location present a formidable challenge.

DISCUSSION

Hemangioblastomas may occur sporadically or as part of VHL complex.[21,22] An autosomal dominant hereditary syndrome that includes retinal angiomatosis, central nervous system hemangioblastomas, and various visceral tumours most commonly involving the kidneys and adrenal glands.

In our case report the patient was 20 years old female which was not in concordance with the literature where male preponderance is more however the patient was in the younger age bracket. [23,24]

The triad of headache, vomiting and cerebellar symptoms has been most common clinical features. Our case also presented with features of raised intra cranial pressure. This is similar to the previous series in which the incidence of papilloedema has ranged from 39 -70% and cerebellar involvement from 40-80%. [25]

Cranial nerve involvement is also seen in 17-35 % of the cases. There may be increased erythrocytes and is believed to be due to secretion of erythropoietin like substance by the neoplastic tissue. The hemoglobin concentration is our case was 15gms% as compared to 16-18 gms % reported in Western literature. [26]

RBC count was 5.76 x 10 /ul while hematocrit was 48.4 %

Radiologically the lesion can either be cystic or solid or both. However 60-72 % are cystic tumours [27].

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MRI is the best tool to diagnose haemangioblastoma which shows an enlarging mass clearly delineated from the surrounding brain or spinal cord tissue.

Plain computed tomography (CT) scan may reveal hypodensity of the tumoural cyst and associated hydrocephalus. CT scan with intravenous contrast show uniform enhancement of the tumour nodule that in association with the adjacent cyst, may be extremely characteristic of posterior fossa haemangioblastomas. Our case study also had CT scan with contrast showing a post surgical defect in the occipital bone. Post contrast images showed small rim enhancing lesion in the left side of cerebellum associated with edema measuring 1.5 cms causing pressure on the 4th ventricle resulting in moderate to severe hydrocephalus. The radiological finding in our case study are similar to Western study. [28].

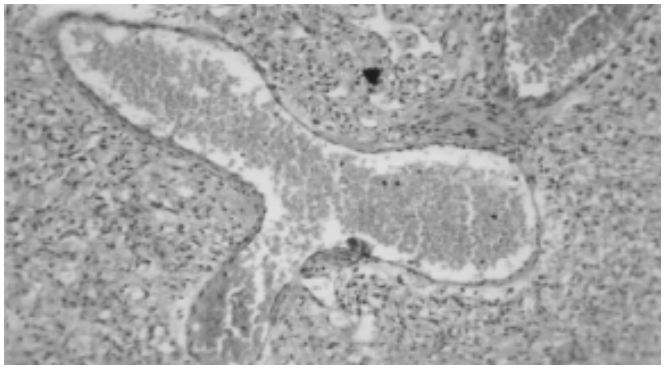
HISTOLOGIC FINDINGS:

Haemangioblastomas are vascular neoplasms. Two hisological subtypes (Cellular and reticular) have been described in primary haemangioblastomas of the central nervous system and have been found to correlate with the probability of tumour recurrence. [29]. Our case histologically showed tumour composed of large thin walled dilated capillaries with interspersed round cells having eosinophilic cytoplasms. No mitosis seen. Photomicrograph [1,11]

Complete excision is the goal of surgery. The cyst wall need not be removed . The solid nodule tumour mass is removed microscopically by dissecting the gliotic margins away from the tiny vascular pedicles that are coagulated to avoid intra-operative swelling. The large draining veins have to be preserved with the arterial feeders to the mural nodule isolated and resected. [30,31]

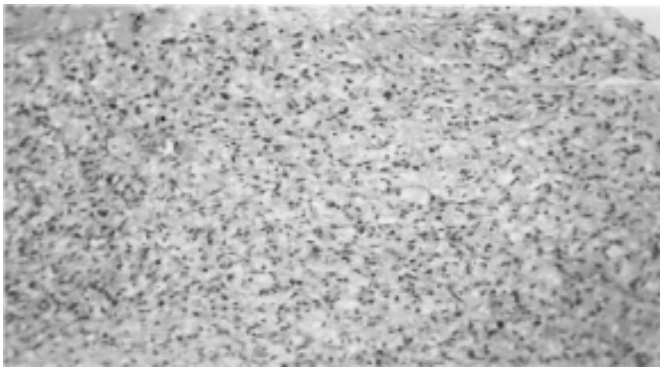
PHOTOMICROGRAPH-I

Thin walled dilated blood capillaries



PHOTOMICROGRAPH-II

Interspered round cells with eosinophilic cytoplasm



Haemangioblastomas treated with gamna knife has show shrinkage of small solitary or medium sized hemangioblastomas. [32,33]

Mortality rate of hemangioblastoma in the literature is 4 – 36 %

CONCLUSION

Haemangioblastomas are benign tumours of uncertain origin that are located predominantly in the posterior cranial fossa and the spinal cord. Although most hemangioblastomas are sporadic, they are associated with autosomally dominant VHL disease in approximately 25% of cases.

The tumours may be solid or cyst and present with raised intra cranial pressure due to obstruction of CSF pathways. Most hemangioblastomas can be cured with surgical resection and long term recurrence rates seem to depend on the presence of VHL disease and multicentric lesions.

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