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Case report

Intracranial Hemangiopericytoma: A Case Report

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Intracranial hemangiopericytomas are rare vascular tumors, accounting for less than 1% of all primary tumors of the central nervous system. This tumor is usually found outside the CNS, specifically in the lower extremities and retroperitoneum. The standard treatment for this condition involves surgery followed by radiotherapy to reduce the risk of recurrence. This case report presents a case report of a young female diagnosed with intracranial hemangiopericytoma in 2021 at Acharya Tulsi Regional Cancer Institute, Bikaner.

Introduction

Hemangiopericytomas were described by stout as malignant tumours which arises from blood vessels and he described these as sarcoma [1]. The cell of origin of these HPCs are pericytes which interacts with endothelial cells and critical for blood vessel formation and maintenance [2]. HPCs are rare tumours occurs mostly outside CNS only (in lower extremities and retroperitoneum) but sometimes also seen in CNS [3]. A case series done by Guthrie et al. [4] showed that most patients presenting with meningeal HPCs are adults, usually 30-45 year of age. Hemangiopericytoma is more commonly located in supratentorial region. Slight male predominance is seen in this tumour (1.4:1). Presenting Symptoms depends on site of mass and when it arises from CNS then common symptoms are headache, seizure, visual dysfunction, and motor weakness. HPCs are significantly less common than meningioma and their clinical behaviour is more aggressive than that of benign meningioma. Moreover, they have a strong tendency of local recurrence and extracranial metastasis

Case Report

•A 35-year-old female was admitted to our hospital (ATRCTRI Bikaner) for a sudden seizure episode for the first time. She had no previous history of seizure or family history of the disease. She had history of weakness and numbness in her left lower limb for last 3 years. The patient had no history of trauma or fever. The neurological examination was done and we found that power in right limb was decreased compare to left and atrophy was seen in limbs.

• CECT brain was done and it showed approx. 5x3 cm Dural based sphenoid wing mass lesion seen. surgery was done to remove the mass and the specimen was sent to our pathology laboratory for histopathological examination.

• Histological examination showed proliferation of a staghorn vascular pattern of spindle cells with moderate amount of eosinophilic cytoplasm but necrosis was not seen in prepared specimen (Figure 1 and 2).

Figure 1. Contouring at TPS on Right Side of Skull (post op volume) with Staghorn Pattern Seen in Pathology Slide.

Figure 2. Contouring is Shown on TPS with Outer Blue Line Shows PTV and Inner Yellow Line Denotes CTV.

Workup

•Patient underwent routine investigations CBC, RFT, USG and CT which showed a lesion in right side causing contralateral midline deviation. After diagnosis, patient underwent tumor embolization and complete surgical excision in February, 2021.

• An Immunohistochemistry of the specimen came positive for STAT6, CD99, and CD34.

• He was started on Adjuvant Radiotherapy 60 Gy/30# in March 2021.

• He remained in clinical follow up with our department, and is clinically stable and showed good response.

Discussion

HPC is a rare neoplasm, arises from pericytes, a supportive cell of the vasculature. Usually, HPCs is arises in lower extremities or retroperitoneum, but rarely, it can occur within the CNS also. HPCs were previously known as angioblastic meningiomas, as the tumors are very vascular and arose from the meninges [5]. Usual presentation of patient in adulthood, with median occurrence of HPC is in the fifth decade of life. In our case the patient age is 35 year. Initially patient present with focal neurological signs and symptoms and that helps in diagnosis of disease. Recurrence rate of tumor is about 60 to 76 %. Very rarely it can also metastasize to other organ like bone, liver lung and peritoneum with 5-year metastasis rate at 33% [4]. Treatment of meningeal HPCs is surgery like meningiomas but because of extensive vasculature most of the time only surgery is not possible and pre-embolization of the tumor before resection can prevent blood loss [6]. Post op radiotherapy can be given to prevent recurrence, but a study done by Dufour et al. [7] showed that radiotherapy did not change the rate of metastases. In our case patient underwent craniotomy and then after 25 days of surgery patient came for post operative radiotherapy. Patient was treated with 60 Gy in 30 fractions and after 6 months of follow up patient was asymptomatic. Use of chemotherapy like Doxorubin- based regimens have been used, but have yet to establish a clear role in the treatment of HPC [8].

Therefore, we conclude that primary treatment for HPCs are surgery followed by radiotherapy. Chemotherapy has limited role here. For recurrent HPCs Dasatinib can be used because Dasatinib can inhibit PDGFR and it has been shown that overexpression of PDGFR is seen in HPCs [9].

Patient consent declaration

We obtained patient consent for images and other clinical information to reporting in the article. The patient understands that his names and initials will not be published.

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