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CENTRAL NEUROCYTOMA - A CASE REPORT

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Abstract :
Central neurocytoma is a rare neoplasm accounting for less than 1 percent of all intracranial tumours. It most commonly grows into the lateral or third ventricle from the septum pellucidum or foramen of Monro. It is the most common neoplasm involving septum pellucidum in young adults. Here we present a case of central neurocytoma diagnosed in a 26 year old female because of its rarity.

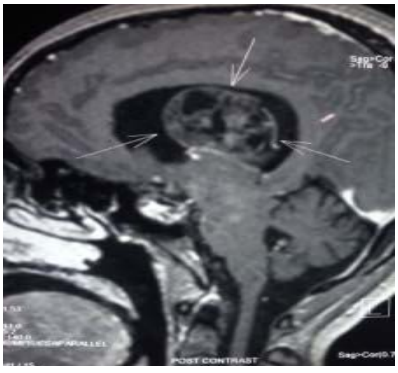
Keyword :
Central Neurocytoma , Septum pellucidum.

INTRODUCTION:

Central neurocytoma was first described by Hassoun et al., in 1982. It is a rare tumor of the central nervous system with neurocytic differentiation and favorable prognosis. In the four-tier World Health Organization classification of tumors of the nervous system, it corresponds to a grade II/IV tumor. Although neurocytoma comprises only 0.25- 0.5% of all intracranial tumors, they are the most frequent intraventricular tumor in adults.

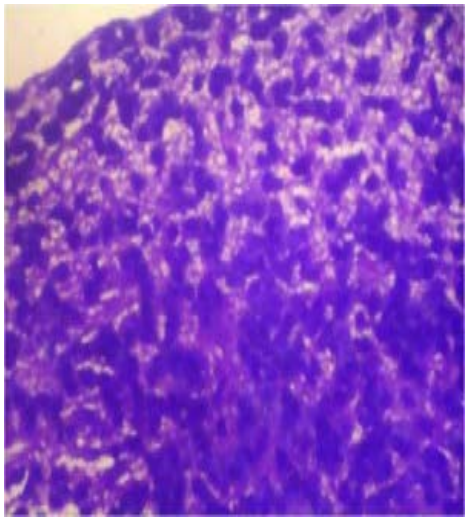
CASE REPORT:

A 26 year old female presented with complaints of headache for four months which was insidious in onset progressive in nature. On examination her general condition and central nervous system functions were normal. MRI revealed a well circumscribed, contrast enhancing intraventricular space occupying lesion with numerous cystic areas giving soap bubble appearance attached to septum pellucidum.Gross total resection of the tumour was done and the specimen was sent for histopathological examination.

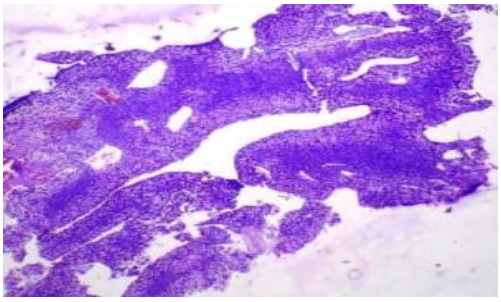


Intraventricular space occupying lesion with soap bubble appearance

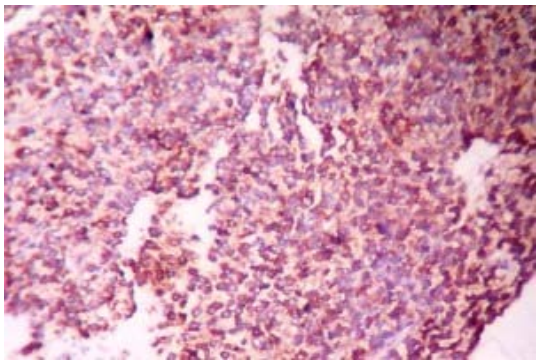
PATHOLOGICAL FINDINGS:
GROSS:
Received multiple gray brown and gray white soft tissue measuring 1 cc in aggregate.
MICROSCOPY:
Histopathological examination revealed a neoplasm composed of monotonous sheets of round to polyhedral cells with moderate amount of eosinophilic cytoplasm and round to oval nuclei with stippled chromatin surrounded by neurofibrillary background arranged in cellular and hypocellular areas. Focal area shows pseudorosette appearance.



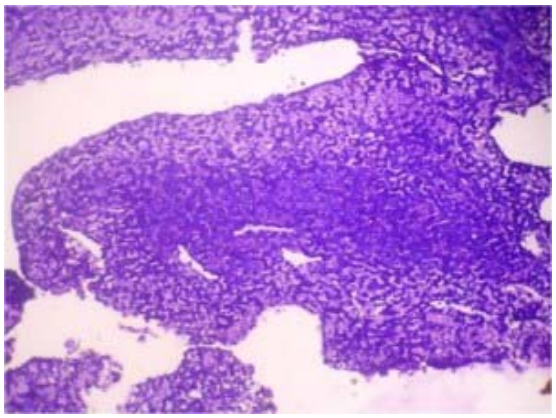
40X View - tumor cells in neurofibrillary background



10X View - tumour cells are arranged in hyper and hypocellular areas 10X View - monotonous sheets of round cells in a neurofibrillary background



Immuno histochemistry showed Synaptophysin positivity and GFAP negativity.



Tumor cell shows Synaptophysin positivity**IMPRESSION**- CENTRAL NEUROCYTOMA

DISCUSSION:
Central neurocytoma is a rare tumour of the central nervous system corresponds to WHO grade II with neurocytic differentiation and has a favourable prognoris.
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Epidemiology: Central neurocytomas are seen in young patients 20-40 years of age, and accounts for less than 1% of all intracranial tumours.[1] It has no reported gender predilection.

Localization- Central neurocytomas are commonly located supratentorially and the anterior portion of one lateral ventricle constitutes 50%, followed by the combined involvement of the lateral and third ventricles (15%) and the involvement of both lateral ventricles (13%) and in the third ventricle in 3% of cases.[2,3] Small neurocytomas have been documented with precise sites of origin at the foramen of Monro, the septum pellucidum, the corpus callosum, the hypothalamus and the thalamus. Tumours having histopathologic, immunohistochemical and biological features similar to central neurocytoma may be found outside the ventricles are termed as "extraventricular neurocytomas". [4,5]

CLINICAL MANIFESTATIONS:
The clinical history is usually short with mean period of 3 months. Most common signs and symptoms are headache, nausea and vomiting due to increased intracranial pressure.Other manifestations are mental disturbances, visual changes, paresthesias, lethargy, loss of balance or tinnitus.

IMAGING FEATURES:
CT- central neurocytomas are isodence or slightly hyperdense, well demarcated, lobulated intraventricular masses. Calcification can be seen in about 50% of the cases. T1- and T2- weighted MRI show a solid mass with occasional cystic components that is heterogeneously isointense or slightly hyperintense relative to the cortex. GROSS: Well demarcated, gray masses often adhere to the ventricular lining or septum pellucidum. It may be partly cystic with varying consistencies from soft to gritty, depending on the amount of calcification. HISTOPATHOLOGY: The tumour is composed of small mature neurocytic cells that are arranged predominantly in sheets. The cells are monotonous, round, regular and densely packed close to one another and separated by a delicate neuropil background. The neoplastic cells with sparse eosinophilic to clear cytoplasm, round or oval nuclei, with finely granular chromatin and small nucleoli. Tumour cells often cluster around small blood vessels stimulating the perivascular pseudorosettes of ependymoma and surrounding central islands of acellular neuropil stimulating Homer Wright- like rosettes.[6] Clear cells and perinuclear halo can occur, mimicking the oligodendroglioma. Calcification and "chicken wire"-like capillaries can also be seen.

ATYPICAL NEUROCYTOMA:
The tumour that exhibits aggressive behaviour as increased proliferation index with MIB-1 labelling index more than 2%, microvascular proliferation, mitosis and necrosis are termed as atypical neurocytoma. However, this diagnostic variant has not been assigned a higher WHO grade. But in the case of spinal neurocytoma, even with low mitotic activity, has aggressive growth and worse prognosis. [1,7,8] Ultrastructurally, central neurocytomas have features of neuronal differentiation that include delicate cytoplasmic processes, microtubules and dense core granules.

MMUNOHISTOCHEMISTRY:
The tumor cells are Synaptophysin positive and GFAP negative.

DIFFERENTIAL DIAGNOSIS: OLIGODENDROGLIOMA:
Presenting as intraventricular mass is

rare and immunohistochemistry shows synaptophysin negativity.

CLEAR CELL EPENDYMOMA:
It shows reactivity for GFAP and Epithelial membrane antigen where as central neurocytomas are GFAP negative.

DYSEMBRYOBLASTIC NEUROEPITHELIAL TUMOR : The tumors that occurs in the third ventricles may suggest neurocytomas. Their mucoid changes, however, are not characteristic of neurocytomas. The small oligodendrocyte- like cells lack reactivity for synaptophysin.

Treatment and Prognosis: The clinical course is usually favourable following complete surgical resection. [1,3] Local control following gross total resection ranges from 85 to 100% at five years and overall five year survival rate ranges from 80 to 95%.

SUMMARY: Central neurocytomas are WHO grade II intraventricular tumor seen in young patients of 20 to 40 years of age, and accounts for less than 1% of intracranial tumors. Although rare central neurocytoma are distinctive and important to differentiate from other small, round, blue-cell tumors, oligodendrogliomas and ependymomas, since central neurocytoma has low proliferative activity and the clinical course of patients are usually favorable.

CONCLUSION:
This case is reported because of its rarity and to highlight the importance of its recognition as it closely mimics many glial neoplasm, since it has a favourable clinical course and has long patient survival rate following gross total resection.

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