

Central neurocytoma: A case report

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Central neurocytoma of the right lateral ventri was described in a 40-year-old man who presented with left hemiparesis. Histopathologically, the tumor cells were monotonously round and possessed perinuclear halo (fried egg appearance) resembling oligodendroglioma. Correct diagnosis requires accurate clinical data, high degree of suspicion and synaptophysin immunostaining.

Key words: Central neurocytoma, Synaptophysin.

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ได้รายงานผู้ป่วยชายอายุ 40 ปี มาพบแพทย์ด้วยอาการอ่อนแรงของแขนและขาซีกซ้าย ตรวจพบก้อนเนื้ออกในช่องข้างของสมองด้านขวา ผลการตรวจทางพยาธิวิทยาพบเซลล์เนื้ออกรูปร่างกลม ขนาดใกล้เคียงกัน มีช่องว่างรอบนิวเคลียส ดูคล้ายไขดาว การย้อมพิเศษ immunohistochemical พบว่า เซลล์เนื้ออกให้ผลบวกต่อ *synaptophysin* ยืนยันการวินิจฉัยว่าเป็น *Central neurocytoma*

Neoplasms of the central nervous system (CNS) are classified according to their histogeneses, if known.⁽¹⁾ During the past two decades, several entities such as pleomorphic xanthoastrocytoma, desmoplastic cerebral astrocytoma of infancy, and dysembryoplastic neuroepithelial tumor have existed.⁽²⁻⁴⁾ In our opinion, some of them are, in fact, not real "new" diseases but, on the other hand, "newly-recognized" ones since clinical study, immunohistochemistry, electronmicroscopy, tissue culture, and other advanced techniques have provided us more information about the origins and behavior of the CNS tumors.

The purpose of this communication is to present a case of central neurocytoma, a rather recent

entity of neural tumor, which, to our knowledge, has not previously been described in Thailand.

Case report

A 40-year-old man experienced numbness of the left arm for two months. Weakness of left extremities was observed a week before admission. Physical examination confirmed that the patient had left hemiparesis and also upper motor neuron type facial palsy of the same side.

A CT scan exhibited an enhancing mass in the right lateral ventricle, extending from the frontal horn to the body (Fig.1). The patient underwent transcallosal craniotomy with partial removal of the tumor.

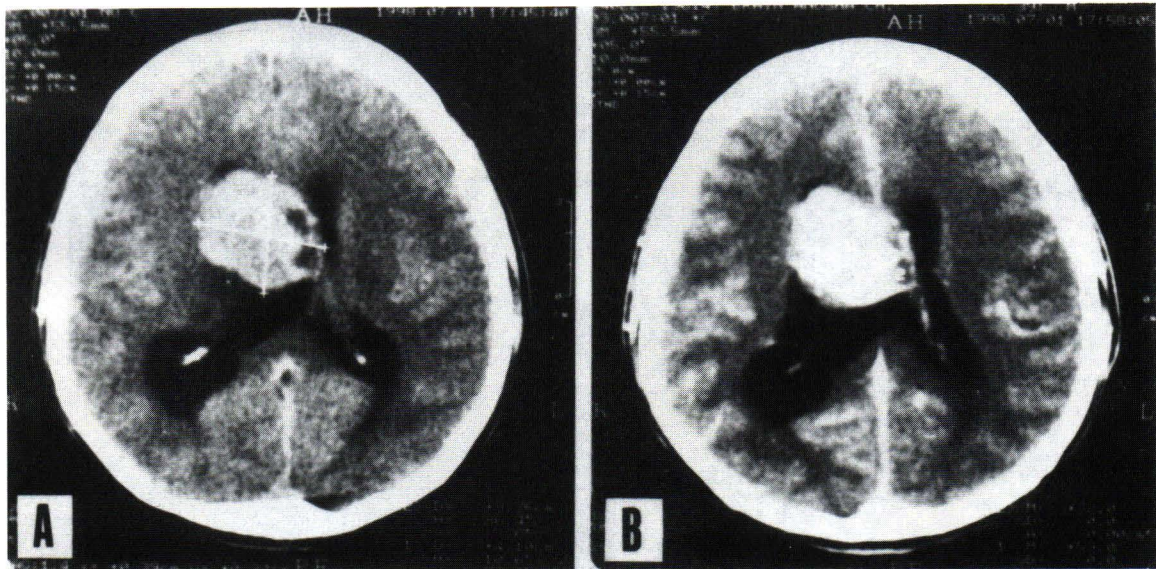


Figure 1. CT scan appearance of central neurocytoma. Unenhanced CT

(A) Shows high density mass, 3.75 x 4.10 cm, with dense enhancement

(B) Occupying frontal horn and body of the right lateral ventricle. The lesion displaces the adjacent mid-line structure to the contralateral side. There are two small areas of hypodensity in medial aspect.

Fragments of the lesion were fixed in 10 % formalin, embedded in paraffin and stained with hematoxylin and eosin. Sections of paraffin - embedded tissue were also processed by peroxidase - antiperoxidase indirect immunohistochemical methods using antibodies to glial fibrillary acidic protein (GFAP), CD 57 (Leu-7), S-100 protein, neurofilament (NF), chromogranin A, and synaptophysin.

Microscopically, section revealed a hypercellular tumor composed of monotonously small round cells with fine granular nuclear chromatin and perinuclear halo. There were many delicate branching capillaries with some perivascular clear zones of fibrillary

matrix scattered in the lesion (Fig 2 A & B). Mitoses were very infrequent. Neither vascular proliferation nor tumor necrosis was encountered.

The tumor cells were immunoreactive for synaptophysin (Fig 2C), Leu-7 and S-100 protein but negative for chromogranin A as well as NF. A few reactive astrocytes were highlighted by GFAP. The pathological diagnosis was central neurocytoma.

In follow-up CT scan, two months after the operation, a small residual tumor was detected in the right lateral ventricle and the patient still had left hemiparesis.

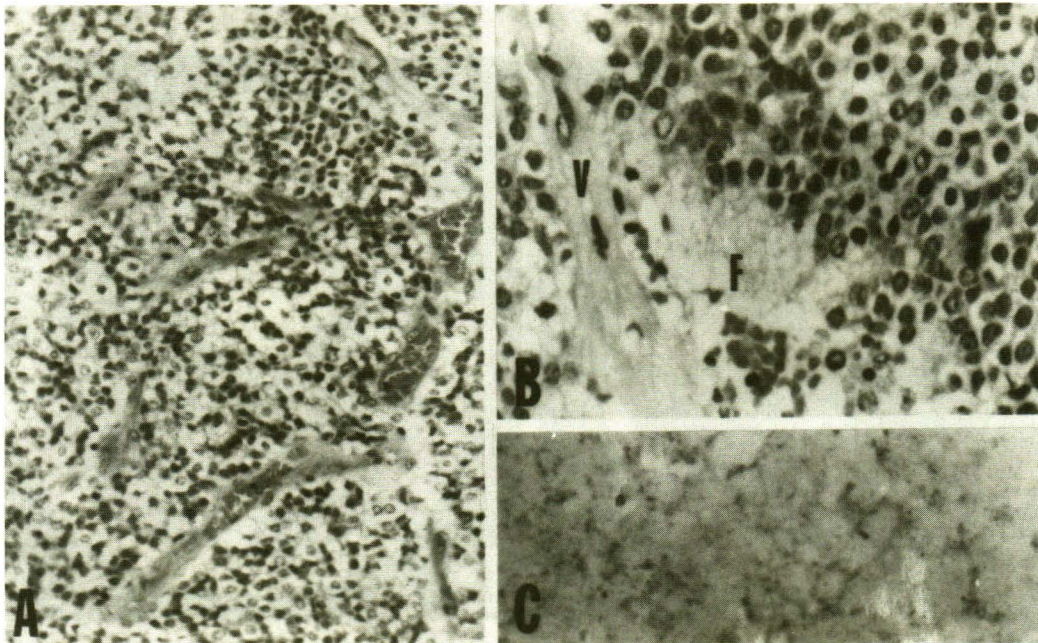


Figure 2. Pathology of central neurocytoma.

- (A) Photomicrograph reveals monotonously small round tumor cells with fine granular nuclear chromatin and perinuclear halo. There are delicate vessels scattered in the lesion (H & E).
- (B) Higher magnification discloses clear zone of fibrillary matrix (F) around a blood vessel (V) (H & E).
- (C) The tumor cells show granular immunoreactivity to synaptophysin.

Discussion

Central neurocytomas are uncommon tumors of the central nervous system recently recognized as a separate pathological entity.⁽⁵⁾ The lesions predominantly affect young adults with no significant sex preference. The most frequent site is the anterior portion of one lateral ventricle with some exceptional instances of spinal, pontine and other location.^(1,5,6) Typically, they are slow growing neoplasms which are surgically curable and are generally associated with excellent prognosis.⁽⁵⁾

Regarding the histopathologic features of central neurocytoma especially the perinuclear halo (fried egg appearance), several differential diagnoses including oligodendroglioma, clear cell ependymoma and intraventricular meningioma should be considered.

Oligodendroglioma has long been known for the characteristic fried egg appearance caused by a delayed fixing artifact of round tumor cells; therefore, it is not surprising that oligodendroglioma may be confused with central neurocytoma.⁽⁷⁾ The intraventricular location of the latter together with synaptophysin immunoreactivity favors the correct diagnosis.

Ependymoma enters the differential diagnosis because it is one of the common intraventricular neoplasms. Furthermore, a clear cell variant displays monotonously round cells with clear cytoplasm resembling central neurocytoma.⁽⁸⁾ However, the absence of the ordinary features of ependymoma such as true (ependymal) and perivascular rosettes as well as GFAP negativity excludes this possibility.

Meningiomas can occur in the ventricles since, normally, there are clusters of meningotheial cells in the stroma of choroid plexus.⁽⁹⁾ These tumors are

notable for various histologic appearances. Some contain uniform round tumor cells with perinuclear halo superficially mimicking oligodendroglioma and also central neurocytoma.^(7,10) The lack of typical characteristics of meningioma, namely cellular whorls, syncytial appearance, intranuclear inclusions, and psammoma bodies in company with synaptophysin positivity of the current tumor is evidence against meningioma.

A variety of other neuronal markers have been demonstrated in the central neurocytoma, including synapsin, neuron-associated class III β -tubulin, MAP-2, tau, calcineurin, the neuron-specific antigen LI and the isoform 180 of the adhesion molecule N-CAM.⁽¹⁾ Immunoreactivity to NF, a marker of advanced neuronal differentiation, is seldom identified⁽¹¹⁻¹³⁾ whereas chromogranin A is constantly negative.^(1,14) Failure to express these markers is probably due to the immaturity of the tumor cells. As detected in our example, S-100 protein and Leu-7 are usually positive but diagnostically insignificant because they lack specificity.^(1,11) To date, synaptophysin is the most reliable immunohistochemical marker for the central neurocytoma.⁽¹⁴⁾ However, its absence does not exclude the diagnosis and, in such circumstance, an additional electron microscopical study is required.⁽¹⁾

The ultrastructural appearance of microtubules and their terminations containing clear vesicles and dense-core neurosecretory granules confirm the neuronal nature of central neurocytoma.⁽⁷⁾ The potential for astrocytic differentiation is still somewhat controversial but is supported by the observation that neurocytoma cells co-express synaptophysin and GFAP after one day *in vitro* cell culture.⁽¹¹⁾ This evidence suggests that the tumor cells retain a bipotential differentiation capacity toward both neuronal and

neuroglial lineages although the exact histogenesis of central neurocytoma remains enigmatic.⁽¹¹⁾

In conclusion, we described herein a case of central neurocytoma, a relatively rare recent entity of nervous system tumor. In addition to accurate clinical information and routine staining, the definite diagnosis requires immunohistochemical study. Pathologists should regard central neurocytoma as a differential diagnosis when dealing with brain tumors containing round cells with fried egg appearance. When such an instance occurs in locations other than the central one, the term central neurocytoma becomes inappropriate and the lesion should be called neurocytoma.

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