J Clin Pathol 1996;49:764–765

Pontine neurocytoma

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Abstract

A case of neurocytoma arising in the rostral pontine region of an 18 year old man is reported. The patient developed a right trochlear nerve palsy and was shown to have a well circumscribed, contrast enhancing mass on magnetic resonance imaging. The tumour was characterised histologically by a uniform population of medium sized round nuclei and slightly eosinophilic cytoplasm or occasional perinuclear halos, with delicate branching capillaries, patches of fibrillary matrix, and occasional perivascular pseudorosettes. Immunohistochemical studies demonstrated strong reactivity for synaptophysin in the fibrillary processes and cytoplasm of tumour cells. The present tumour is an exceptional case of neurocytoma arising in the pons.

(*J Clin Pathol* 1996;**49**:764–765)

Keywords: neurocytoma, pons, synaptophysin.

The central neurocytoma, a well differentiated tumour of neuronal origin, is an uncommon primary central nervous system (CNS) tumour predominantly affecting young adults, typically located in the supratentorial ventricular system with characteristic radiological features, and associated with a favourable prognosis after surgical removal. Clinically, the tumour usually causes non-specific symptoms and signs related to raised intracranial pressure, as well as visual and mental disturbances. On magnetic resonance imaging (MRI), the tumour is well circumscribed and isointense with the cerebral cortex or slightly hyperintense on T1 and T2 weighted images. A discrete signal enhancement is demonstrated following the administration of Gadolinium-DTPA. The pathological diagnosis of neurocytoma is based upon immunohistochemical or ultrastructural, or both, features of neuronal differentiation, in addition to the histological characteristics on light microscopy, resembling the picture of oligodendrogliomas or ependymomas.[1] Although central neurocytomas are in most cases located in the lateral ventricle(s) or the third ventricle, or both, exceptional cases of spinal cord neurocytoma have been reported.[2–5] Ellison *et al*[6] described a midline cerebellar tumour with neuronal and lipocytic differentiation and named this unique neoplasm neurocytoma/lipoma (neurolipocytoma). To our knowledge, neurocytomas arising in the brain stem have not been described previously in the literature. This report presents a case of pontine tumour with the histological and immunohistochemical characteristics of central neurocytoma.

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Accepted for publication
4 June 1996

Case report

An 18 year old man presented with an acute onset of horizontal and vertical diplopia, and was found to have a right trochlear nerve palsy on physical examination. MRI obtained elsewhere demonstrated a round, well defined mass, 1.2 cm in diameter, in the region of right superior cerebellar peduncle of the rostral pons. The lesion appeared isointense on T1 weighted images and hyperintense on T2 weighted images, and was enhanced by contrast medium. At surgery, the mass was totally excised from the region of right superior cerebellar peduncle through a suboccipital craniotomy approach. The postoperative course was complicated by acute pyogenic meningitis with subsequent communicating hydrocephalus. The patient recovered gradually following the conventional treatment with antibiotics, as well as placement of a right ventriculoperitoneal shunt. No postoperative radiotherapy or chemotherapy was given. Follow up MRI studies revealed no evidence of tumour recurrence 14 months after surgery.

LIGHT MICROSCOPY

The tumour was composed of a uniform population of medium sized round cells, associated with delicate branching capillaries and patches of fibrillary matrix. An arrangement of the tumour cells around capillaries was occasionally observed, resembling the perivascular pseudorosettes of ependymoma (fig 1). The tumour cells had regular nuclei with a "salt and pepper" chromatin pattern, and slightly eosinophilic fibrillary cytoplasm with indistinct cell borders (fig 1, inset). Perinuclear halos were seen occasionally. There were scattered small foci of cystic degeneration, filled with bubbly basophilic material. Foci of vascular proliferation and accumulations of haemosiderin laden macrophages were identified. Mitotic figures were sparse, essentially limited to proliferating vascular structures, and tumour necroses were absent.

IMMUNOHISTOCHEMISTRY

Strong reactivity for synaptophysin (Dako, Glostrup, Denmark; polyclonal, dilution 1 in 50) was present in the vast majority of tumour cells, particularly in the fibrillary processes and focally in the cytoplasm (fig 2). The small number of tumour cells (up to 4%) showed cytoplasmic reactivity for glial fibrillary acidic protein (GFAP; Biomeda, Foster City, California, USA; polyclonal, dilution 1 in 160; and Biogenex, San Ramon, California, USA; monoclonal, dilution 1 in 5).

Discussion

The pathological diagnosis of the present case was based on the histological and immunohistochemical findings that revealed the pathological characteristics of central neurocytoma:

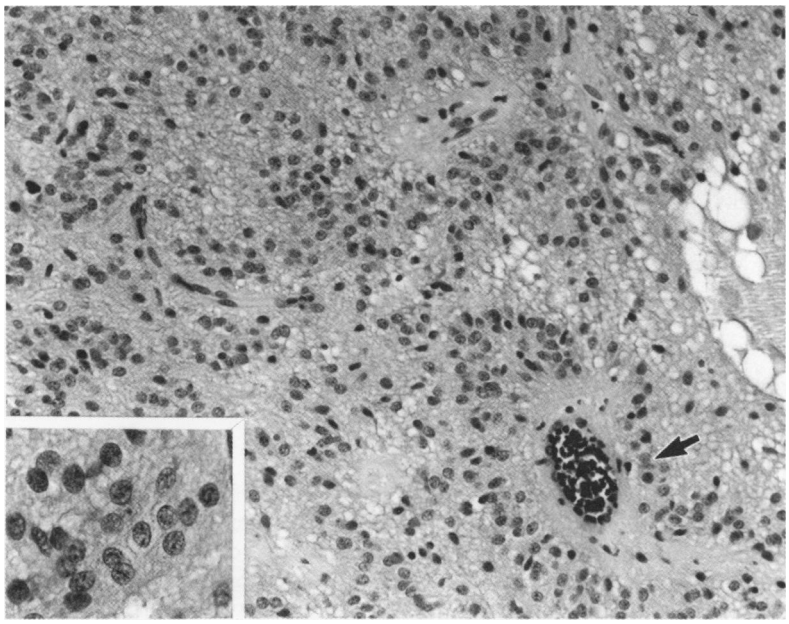

Figure 1 Light microscopy showing uniform round cells with patches of fibrillary matrix, cystic degeneration, and perivascular pseudorosette formation (arrow) (haematoxylin and eosin). Inset: higher magnification of tumour cells demonstrating a "salt and pepper" chromatin pattern.

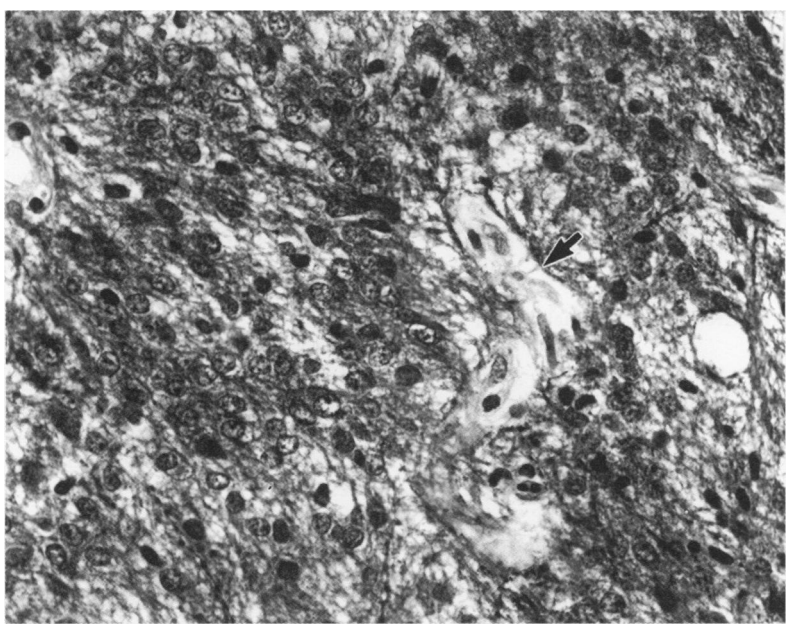

Figure 2 Synaptophysin immunohistochemical reactivity in tumour cells; note the absence of synaptophysin reactivity in vascular endothelial cells (arrow).

(1) the tumour cells showed uniform, medium sized round nuclei and slightly eosinophilic cytoplasm or perinuclear halos, in association with delicate branching capillaries, patches of fibrillary matrix, and perivascular pseudorosettes; (2) the vast majority of tumour cells expressed immunoreactivity for synaptophysin. According to the anatomical location in the pontine region and the histological features of this case, a differential diagnosis with ependymoma and oligodendroglioma was considered, although the latter is rare outside the cerebral hemispheres.[7] Nevertheless, the presence of synaptophysin immunoreactivity, which has been documented to be the definitive evidence for neuronal or neuroendocrine differentiation, strongly supported the diagnosis of neurocytoma in the present case.[8] Expression of synaptophysin has not been reported in astrocytomas, ependymomas, or oligodendrogliomas and it has been considered to be the most suitable and reliable immunohistochemical marker for the diagnosis of central neurocytoma.[1 9 10] In two reports by von Deimling *et al*, one described two neurocytomas which contained a considerable percentage of GFAP positive tumour cells by immunohistochemistry.[10] In the other report, coexpression of synaptophysin and GFAP by a high percentage of individual tumour cells was demonstrated by immunohistochemistry on adjacent histological sections of one neurocytoma; and by western blot analysis all four tumours studied were found to express both synaptophysin and GFAP.[11] GFAP expression by the small proportion of tumour cells in the present case was considered exceptional. Unfortunately, the appropriate tissue was not available for immunoblotting analysis or ultrastructural examination. The presence of vascular proliferation together with high mitotic activity and necrosis of the tumour cells in central neurocytomas has been considered to be histological evidence of anaplasia or malignancy; however, the prognostic significance remains to be determined.[1 10 11] In the present case, rare mitotic figures were seen in the proliferating vascular structures but not in the tumour cells and there was no evidence of tumour necrosis.

We consider that the present tumour is an exceptional case of neurocytoma arising in the rostral pontine region. Although this patient developed postoperative meningitis, he seemed to have a favourable prognosis after complete surgical excision alone, as did the patients with a supratentorial central neurocytoma reported previously.[1] This observation suggests that neurocytomas can occur in the infratentorial location and be included in the differential diagnosis of primary CNS neoplasms in this location.

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