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Neuropathology Education

Pigmented intraventricular tumor in a young adult

Kirti Gupta,[1] Balan Louis Gaspar,[1] Pravin Salunke[2] and Charan Singh Rayat[1]

Departments of [1]Histopathology, and [2]Neurosurgery, Post Graduate Institute of Medical Education and Research, Chandigarh, India

CLINICAL COURSE

A 22-year-old man presented with complaints of progressively worsening headache for the past 6 months. Clinical examination revealed papilledema. There were no sensory or motor deficits. Radiology showed an intraventricular heterogeneous lesion occupying mainly the body of right lateral ventricle with some growth into the left lateral ventricle (Fig. 1). The features suggested central neurocytoma. He was operated through an endoscopic approach that required conversion to open surgery due to intraoperative bleeding. The tumor was fleshy and vascular. Although near total excision was achieved, he succumbed to ventriculitis.

Pathological findings

The tumor demonstrated circumscribed margins on histology and was defined by a uniform population of small round cells with fine, stippled chromatin and small nucleoli with few interspersed pale islands of low cell density. Admixed with these, many tumor cells (constituting approximately a 30 % neoplastic population) revealed finely dispersed to coarsely granular blackish-brown pigment within their cytoplasm. Few large cells with ganglionic differentiation were also identified within the uniform small round cells. The tumor was supported by fine neuropil and a delicate capillary network (Fig. 2). No atypical features including mitoses, necrosis and endovascular proliferation or rosettes were identified. The pigment was positive with Schmorl and Masson Fontana stains (Fig. 3A,B) and was bleached with potassium permanganate, thus confirming its nature to be melanin. It was negative with Perl's and PAS stains. The tumor cells (including both pigmented and non-pigmented cells) revealed fine, granular immunoreactivity with synaptophysin and nuclear positivity with neuronal nuclear antigen (NeuN) (Fig. 3C,D). GFAP highlighted few entrapped reactive astrocytes (Fig. 3E) and tumor cells were negative with epithelial membrane antigen (EMA). Ultrastructurally, the tumor cells demonstrated numerous neurosecretory granules in their cytoplasm (Fig.4). Pigmented cells revealed presence of melanosomes, with some cells containing both melanosomes and dense core neurosecretory granules in their cytoplasm (Fig. 5A,B).

Diagnosis

Pigmented central neurocytoma (CN), WHO grade II.

DISCUSSION

CN are low-grade neuronal tumors arising in the lateral ventricles in young adults.[1,2] The supratentorial ventricular system is the frequent site; however, extraventricular examples are also on record.[2–4] Rare histopathologic variants of neurocytoma include ganglioneurocytoma, cerebellar liponeurocytoma and pigmented neurocytoma.[1,5] Pigment deposition with CN where the tumor cells contain melanin, lipofuscin and occasionally hemosiderin, is very rare with few cases illustrated in the literature.[6–9] Cytochemical stains including Masson's Fontana, PAS and Prussian blue stains are useful in highlighting and confirmation of nature of the pigment, which was found to be melanin in the present case based on its positivity with Schmorl, Masson Fontana stains and bleaching property with potassium permanganate. There was no difference in nature, morphology and immunophenotype of pigmented and non-pigmented tumor cells. Exceptional cases of pigment deposition have been described in other primary intracranial tumors, including choroid plexuses carcinoma, gliomas and ependymoma, which have to be particularly considered in the differential diagnosis at this site. Meningeal melanocytomas and melanomas (both primary and metastatic) are the classic examples of

Correspondence: Kirti Gupta, MD, Department of Histopathology, Postgraduate Institute of Medical Education and Research, (PGIMER), Chandigarh. India.
Email: kirtigupta10@yahoo.co.in

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