ISSN: 2634 - 680X

Journal of Clinical Case Studies Reviews & Reports

SCIENTIFIC Research and Community

Case Report

Open Access

Intraventricular Pigmented Ependymoma with CSF Rhinorrhoea: A Rare Case with Review of Literature

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ABSTRACT

Intraventricular tumors present a diagnostic as well as surgical challenge to all the Neurosurgeons. Pigmented tumours in the Central nervous system are rare lesions and most commonly are meningeal melanocytic tumours or metastatic melanomas. Pigmented Ependymomas are much rarer and only 8 cases have been reported in the literature yet. We here present a case of 37-year-old female who had an unusual presentation with CSF Rhinorrhoea, on further evaluation she was found to have an intraventricular tumour. Post-operatively she was found to have a rare variety of Intraventricular tumour- Pigmented Ependymoma.

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Received: January 11, 2022; **Accepted:** January 17, 2022; **Published:** January 23, 2022

Keywords: CSF Rhinorrhea, Ependymoma, Hydrocephalus, Intraventricular Tumor, Pigmented Ependymoma

Abbreviations:
ADC: Apparent Diffusion Coefficient
CT: Computed Tomography
CSF: Cerebrospinal Fluid
FLAIR: Fluid attenuated inversion recovery
GFAP: Glial fibrillary acidic protein
IHC: Immunohistochemistry
MRI: Magnetic Resonance Imaging
PAC: Periodic acid–Schiff
PNS: Paranasal sinuses
T1W: T1 weighted
T2W: T2 weighted

Case Report

A 37-year-old female presented with history of spontaneous watery discharge from left nostril, there was no associated history of any fever, headache, nausea, vomiting or blurring of vision. She consulted an ENT specialist for the same and CT PNS was done. The CT scan was suggestive of a bony defect in the left cribriform plate and an incidental intraventricular lesion was detected. Further MRI Brain with gadolinium contrast was done to evaluate the intraventricular lesion, MRI findings revealed a solid cystic intraventricular heterogeneous lesion occupying the body of right lateral ventricle. The tumour was seen attached to the septum pellucidum with haemorrhagic foci within with contrast enhancement. The solid part of the lesion is appearing iso-intense on T2W images and hyperintense on T1W/FLAIR and showing restricted diffusion with corresponding low ADC values (Figure 1).

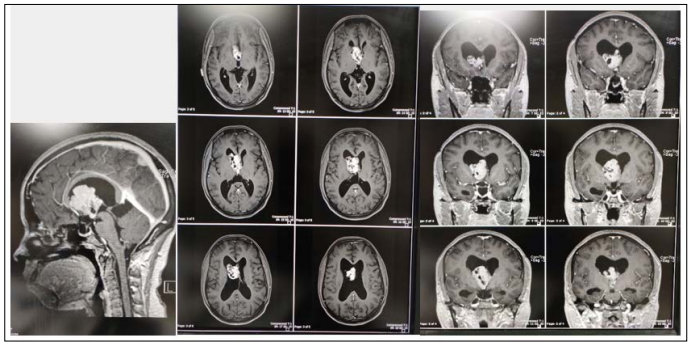

Figure 1: A well defined solid cystic intraventricular lesion is seen in the right lateral ventricle. It is causing mild dilatation of the right lateral ventricle and is seen attached to the septum pellucidum and is deviating to the left side by 6.3mm.On post contrast images, it is showing heterogeneous enhancement.

The features were suggestive of a Central Neurocytoma. She was admitted at our hospital for further management. After admission, a thorough clinical examination was done which revealed Grade 2 papilledema. There were no motor or sensory deficits.

After obtaining informed consents she under underwent Right fronto-parietal craniotomy for tumour excision along with placement of right frontal ommaya reservoir. She also underwent Left frontal craniotomy and Anterior Cranial Fossa repair for the CSF Leak (Figure 2).

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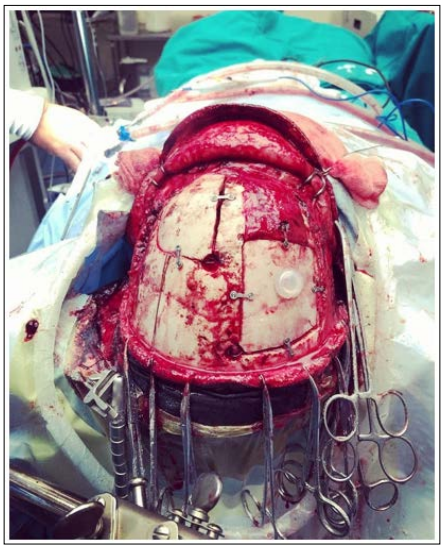

Figure 2: Right fronto-parietal craniotomy for tumour excision, with right frontal ommaya reservoir. Left frontal craniotomy with autologous pericranial graft for CSF Rhinorrhea repair

Intraoperatively the tumour was dirty brown coloured, soft, fleshy, vascular lesion. Gross total tumour excision was done. Post-operative NCCT Head scan (Figure 3) showed satisfactory post-operative changes. She did not have any CSF Rhinorrhoea and she recovered well. She was discharged in stable condition.

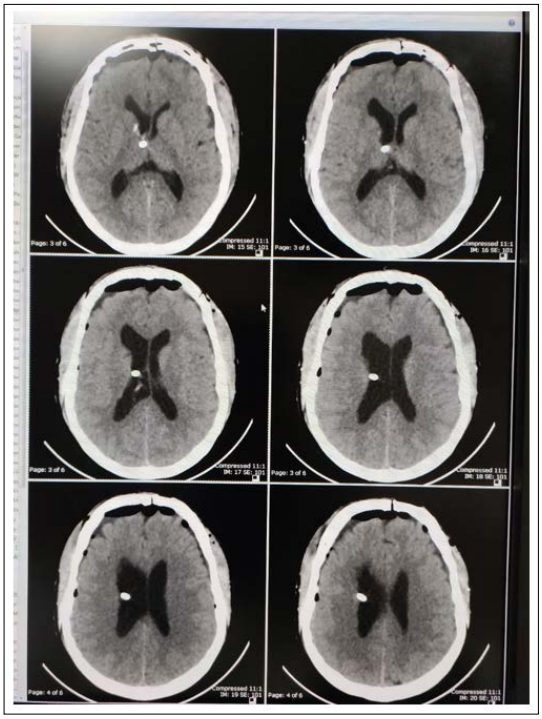

Figure 3: Post-operative NCCT Head was suggestive of satisfactory postoperative changes, with ventricular catheter in situ

Histopathology

Formalin-fixed paraffin-embedded sections were stained with hematoxylin-eosin stain. Tumour was sharply demarcated from the glial tissue. The sections showed a tumour composed of cells having a pleomorphic to round –oval nuclei. The nuclei had salt and pepper chromatin with moderate amount of eosinophilic cytoplasm. The cells were arranged in sheets with prominent perivascular pseudorosettes and true ependymal rosettes. Seen throughout along with inclusion bodies. Most of the cells contained abundant brownish pigment in their cytoplasm. No mitosis or any necrotic tissue was seen. Background was fibrillary. Cytoplasmic processes radiating out from central vascular cores were seen, typical of an Ependymoma (Figure 4 a &b).

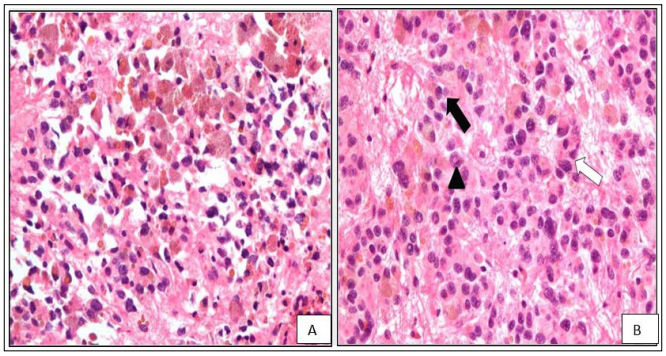

Figure 4: a) showing true rosette (white arrow), pseudo rosette (black arrow), intranuclear inclusion (arrowhead) **b)** showing abundant intracellular pigment [H & E, 400x].

Histochemistry

We performed a battery of stains to identify the pigment. The tumor must be distinguished from meningeal melanocytic tumours and metastatic malignant melanomas as these entities have different clinical, therapeutic and prognostic implications. The stains included, Masson-Fontana and melanin bleach (melanin), Sudan black B, Oil red O and, periodic acid–Schiff (PAS; glycogen). The pigment in our case was melanin positive (Figure 5 a) not acid-fast positive, periodic acid–Schiff (PAS) negative (Figure 5 b). It was Masson-Fontana Positive. The positivity for Masson-Fontana was abolished after pre-treatment with potassium permanganate for 10 mins. This duration of pre-treatment is not adequate to bleach lipofuscin, hence this pigment was thought to be neuromelanin/ melanin or a mixture of both.

Immunohistochemistry

Immunohistochemistry was performed to confirm the diagnosis of ependymoma. GFAP- Glial fibrillary acidic protein was positive in the tumour cells with a perivascular accentuation (Figure 5 c) and S100 (Figure 5 d). diffusely positive. Epithelial membrane antigen showed numerous cells with a Para nuclear dot-like positivity (Figure 5 g) corresponding to microlumina seen in an ependymoma. HMB45 IHC was negative in our case, thus ruling out the possibility of a metastatic malignant melanoma. Immunohistochemical profile confirmed the tumour to be a pigmented ependymoma. Thus, Identification of this rare entity requires the awareness about this variant.

Special stains

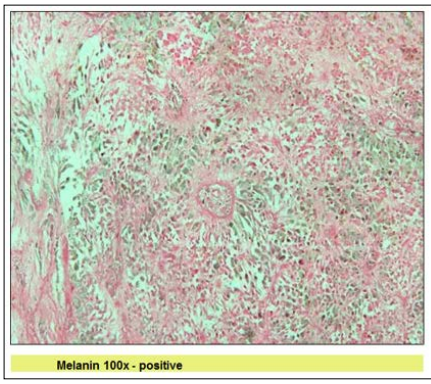

Figure 5: a

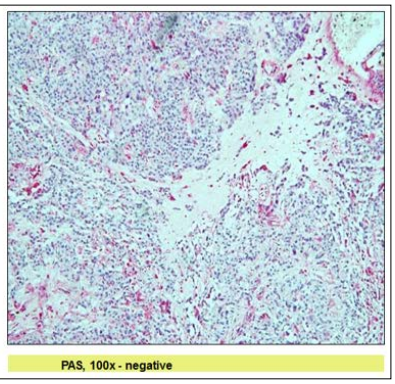

Figure 5: b

Immunohistochemistry

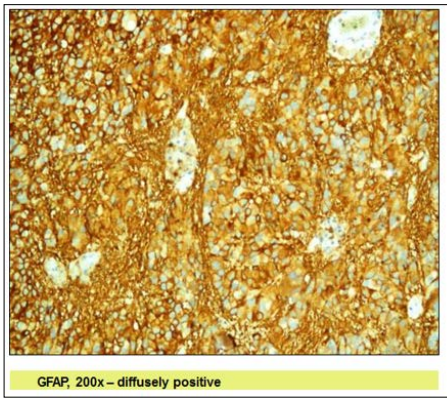

Figure 5: c

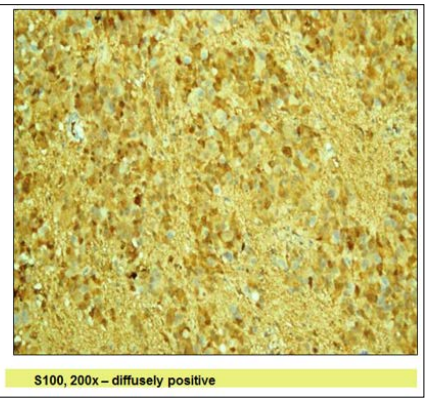

Figure 5: d

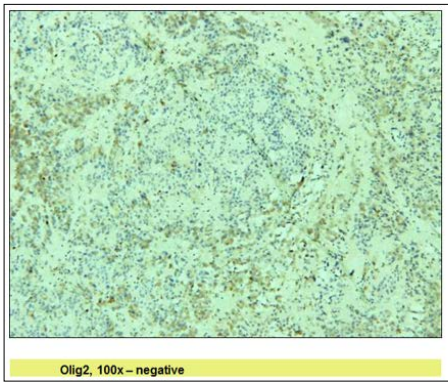

Figure 5: e

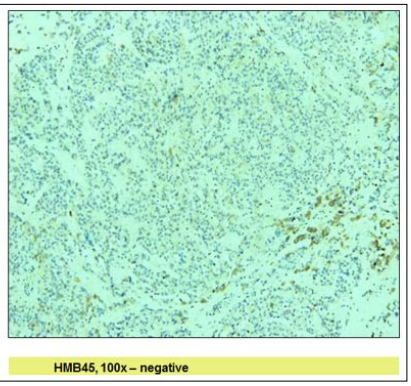

Figure 5: f

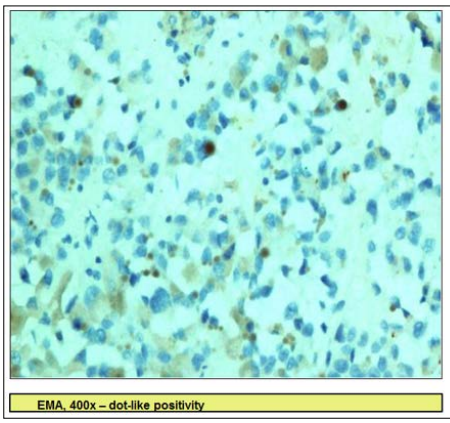

Figure 5: g

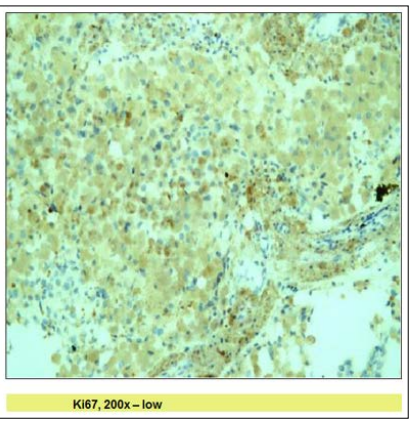

Figure 5: h

Discussion

There are 3 types of pigments which have been described in central nervous system. Neuromelanin, Melanin, and Lipofuscin. Pigment production has been seen in circumscribed gliomas, neurocytomas, ependymomas, subependymomas, and choroid plexus tumors [1-3]. Melanin, is derived from tyrosine. Inside specialised structures called melanosomes the tyrosine substrates are acted upon by tyrosinase. Neuromelanin is distinctly different from melanin, which is believed to be produced by nonenzymatic autooxidation of dopamine. Lipofuscin, is derived from iron-catalyzed peroxidation of membrane lipids and lipoproteins inside lysosomes. It further undergoes crosslinking and polymerization of its proteins with addition of saccharides over the period of time to mature into this non degradable pigment of a post mitotic cell. Pigmented ependymomas are rare tutors. To the best of our knowledge only 8 cases of pigmented ependymomas have been reported in the literature. (Table 1), out of which 4 cases deduced the pigment to be melanin. They postulated that melanin in these tumours is due to the fact that ependymal lining cells derive from primitive ciliated neuroepithelium, which has an inherent capacity for melanin production, as is the case for optic cup lining.

Table 1

Authors and Year of report	Age and Sex	Site of the Lesion	Pigment in Tumour
McCloskey et al 1976 [4]	30/F	Posterior Temporal Lobe.	Melanin
Rosenblum et al 1990 [5]	13/F	Fronto-parietal lobes	Melanin
Rosenblum et al 1990 [5]	52/M	Subependymoma in fourth ventricle	Melanin
Kirkpatrick 2000 [6]	36/M	Fourth ventricle	-
Chan et al 2003 [7]	52/M	Fourth ventricle	Neuromelanin
Ertan et al 2010 [8]	35/F	Fourth ventricle	Lipofuscin + Neuromelanin
Ogawa et al 2016 [9]	26/M	Sella turcica	Melanin
Malhotra et al 2020 [10]	16/M	Fourth ventricle	Lipofuscin.
Present Case. 2021	37/F	Right Lateral Ventricle.	Neuromelanin

In 5 out of the 8 reported cases the tumour was located in the Fourth Ventricle. Medulloblastomas with melanotic differentiation also must be excluded in cases arising in the fourth ventricle. In one case it was located in the fronto-temporal lobe, 1 in posterior temporal horn and another in sella turcica. Our case is the first case of a pigmented ependymoma located in the lateral ventricle. The presentation with CSF Rhinorrhoea also makes this case unique as none of the other cases had similar presentation. The case is reported for its unique and rare features. Our workup concludes that the pigment is neuromelanin/melanin or mixture of both. The pathological significance of this finding is yet to be determined taking into conjunction similar reports of neuromelanin rich ependymomas.

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