Neurological Case Reports

Case Report
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Intraventricular Pigmented Ependymoma with CSF Rhinorrhea: 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 tumors in the central nervous system are rare lesions and most commonly are meningeal melanocytic tumors 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 rhinorrhea; on further evaluation she was found to have an intraventricular tumor. Post-operatively she was found to have a rare variety of Intraventricular tumour- pigmented ependymoma.

Keywords: CSF rhinorrhea; Ependymoma; Hydrocephalus; Intraventricular tumor; Pigmented ependymoma

Case Presentation

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 tumor was seen attached to the septum pellucidum with hemorrhagic foci within with contrast enhancement. The solid part of the lesion is appearing iso-intense on T2W images and hyper intense on T1W/FLAIR and showing restricted diffusion with corresponding low ADC values (Figure 1).

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.

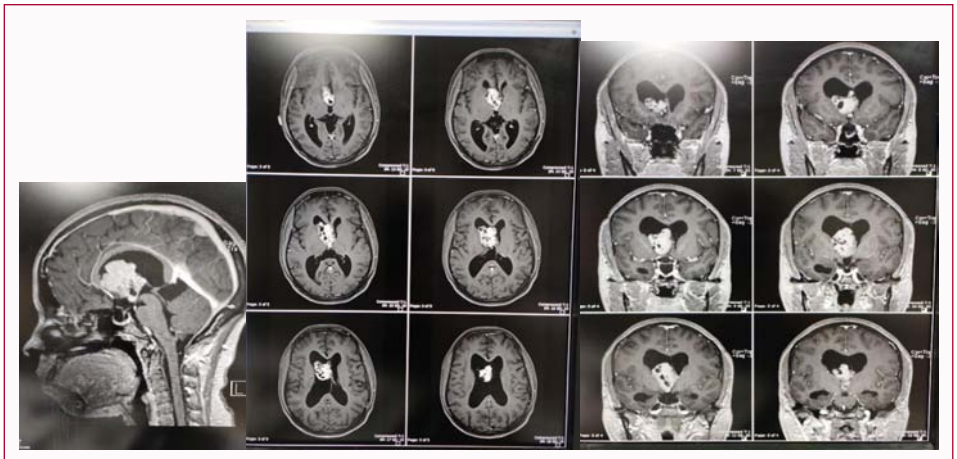

Figure 1: A well defined solid cystic intraventricular lesion is seen in the right lateral ventricle. It is causing mild dilatation of the right lateral ventricle and is seen attached to the septum pellucidum and is deviating to the left side by 6.3 mm. On post contrast images, it is showing heterogeneous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

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Table 1: The 8 cases of pigmented ependymomas have been reported in the literature.

Authors and year of report	Age and sex	Site of the lesion	Pigment in tumour
McCloskey et al. [6]	30/F	Posterior temporal lobe	Melanin
Rosenblum et al. [10]	13/F	Fronto-parietal lobes	Melanin
Rosenblum et al. [10]	52/M	Subependymoma in fourth ventricle	Melanin
Kirkpatrick et al. [4]	36/M	Fourth ventricle	-
Chan et al. [3]	52/M	Fourth ventricle	Neuromelanin
Ertan et al. [3]	35/F	Fourth ventricle	Lipofuscin + Neuromelanin
Ogawa et al. [8]	26/M	Sella turcica	Melanin
Malhotra et al. [5]	16/M	Fourth ventricle	Lipofuscin
Present Case. 2021	37/F	Right lateral ventricle	Neuromelanin

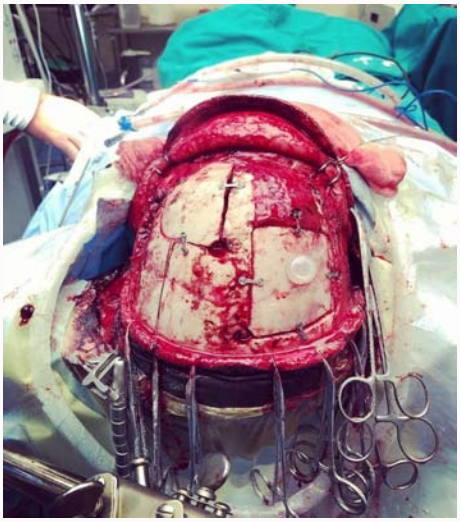

Figure 2: Right frontoparietal craniotomy for tumor excision, with right frontal ommaya reservoir. Left frontal craniotomy with autologous pericranial graft for CSF rhinorrhea repair.

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).

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

Histopathology

Formalin-fixed paraffin-embedded sections were stained with hematoxylin-eosin stain. Tumor 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 pseudo rosettes 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 4a, 4b).

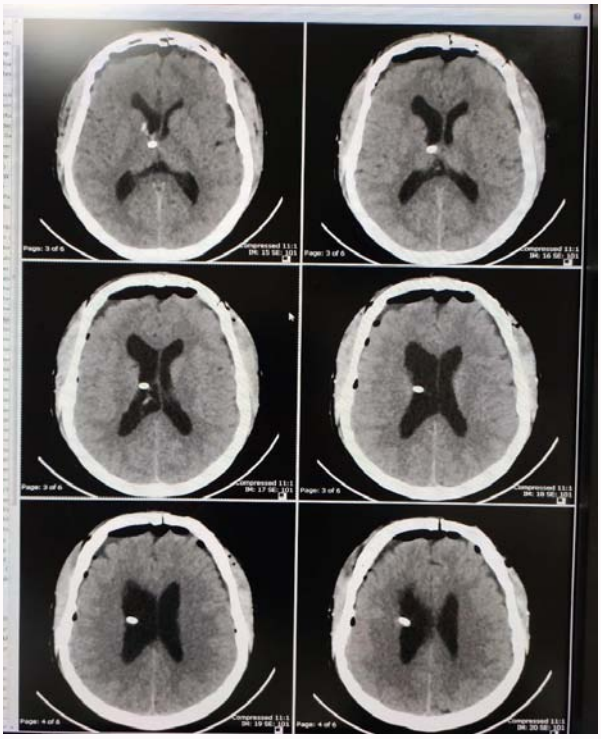

Figure 3: Post-operative NCCT Head was suggestive of satisfactory postoperative changes, with ventricular catheter *in situ*.

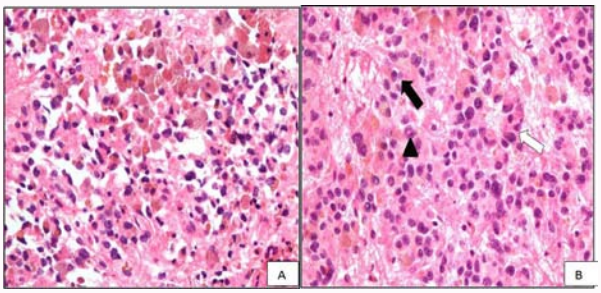

Figure 4: a) Showing true rosette (white arrow), pseudo rosette (black arrow), intranuclear inclusion (arrowhead) b) showing abundant intracellular pigment [H&E, 400x].

Histochemistry

We performed a battery of stains to identify the pigment. The tumor must be distinguished from meningeal melanocytic tumors and metastatic malignant melanomas as these entities have different

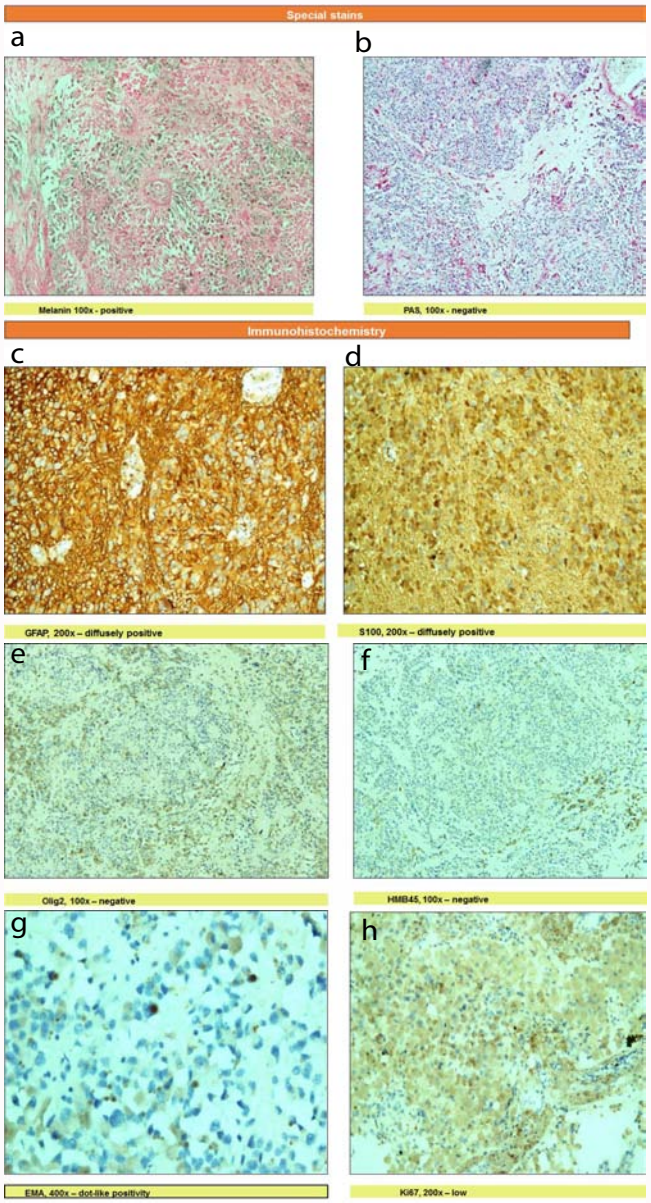

Figure 5a, 5b: Special stains.
Figure 5c, 5d: Immunohistochemistry.
Figure 5e, 5f: Special stains.
Figure 5g, 5h: Special stains.

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 in Figure 5a not acid-fast positive, Periodic Acid-Schiff (PAS) negative in Figure 5b. It was Masson-Fontana Positive. The positivity for Masson-Fontana was abolished after pre-treatment with potassium permanganate for 10 min. 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 5c and S100 Figure 5d diffusely positive. Epithelial membrane antigen showed numerous cells with a Para nuclear dot-like positivity Figure 5g 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.

Discussion

There are 3 types of pigments which have been described in central nervous system. Neuromelanin, melanin and lipofuscin [1-6]. Pigment production has been seen in circumscribed gliomas [7,8], neurocytomas [9], ependymomas, subependymomas, and choroid plexus tumors [2]. Melanin is derived from tyrosine. Inside specialized structures called melanosomes the tyrosine substrates are acted upon by tyrosinase. Neuromelanin is distinctly different from melanin, which is believed to be produced by non enzymatic auto oxidation of dopamine. Lipofuscin is derived from iron-catalyzed peroxidation of membrane lipids and lipoproteins inside lysosomes. It further undergoes cross linking 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 [10]. They postulated that melanin in these tumors 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. 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 rhinorrhea 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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