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Glial hamartoma presenting as obstructive hydrocephalus in a patient with Neurofibromatosis-1

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INTRODUCTION

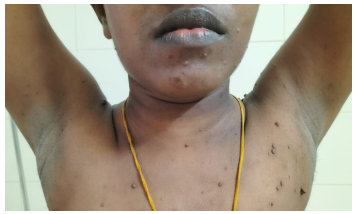
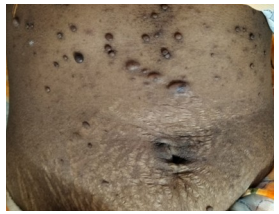
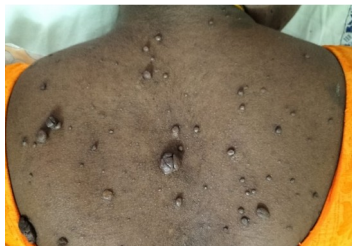
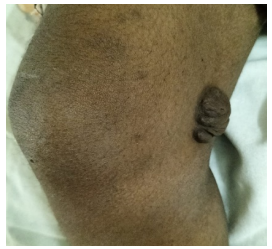
- NF-1 is a tumor disorder that is caused by the malfunction of a gene on chromosome 17 that is responsible for control of cell division.
- Cranial manifestations usually include glial tumors of the central nervous system, mainly optic pathway gliomas and astrocytomas.
- Other CNS manifestations of NF-1 are “hamartomas” or focal areas of high signal intensity on T2-weighted images.
- We present a patient with NF-1 who had a symptomatic, pathologically verified hamartoma of the third ventricle.

CASE REPORT

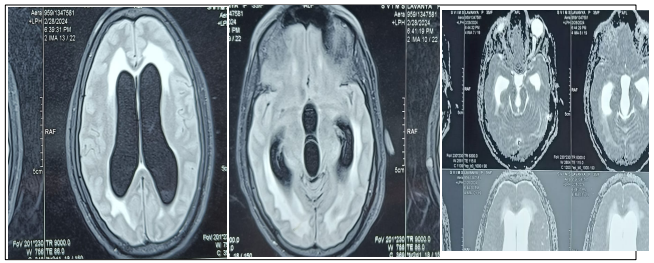
- This is a 35-year-old woman with NF-1 presented with giddiness for 4 months associated with holocranial headache, gait abnormality in the form of swaying for past 1 month, 2 episodes of generalised tonic clonic seizures 1 month ago and 3 days prior to admission. She had no family history of neurofibromatosis.

EXAMINATION

- NF-1 with multiple subcutaneous nodules,axillary freckling and café-au-lait spots. Her Visual acuity was bilateral 6/9 and fundus exmination showed grade 3 papilloedema. Motor power and sensory examination yielded normal results.

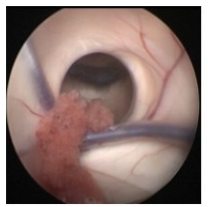
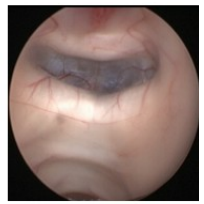
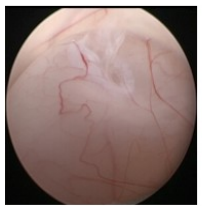
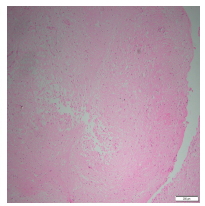
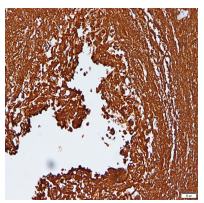
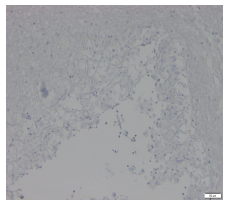





IMAGING



INTRA OP AND HPE

- At surgery, endoscopic third ventriculostomy along with puncture of cyst and biopsy was done.
- Through foramen of monro entered into third ventricle and Lilliquist membrane is fenestrated with fogatry catheter and balloon inflation done, CSF entered into the cisternal space clearly from third ventricle
- On IHC, it showed GFAP, OLIG2, S100- positive. TTF1- negative. CD 68- highlights admixed macrophages, negative in atypical cells. EMA- negative. Mib-1 labelling index-1%.

CONCLUSION

- In the present case, the lesion had produced symptoms of obstructive hydrocephalus consisiting of giddiness, headache and ataxia. Magnetic resonance imaging revealed CSF intensity cyst measuring 2.6x 2.4x1.6 cm in posterior third of 3rd Ventricle. Pathological examination showed glial hamartoma.

REFERENCES

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2.Balestrazzi P, de Gressi S, Donadio A, et al. Periaqueductal gliosis causing hydrocephalus in a patient with neurofibromatosis type 1. Neurofibromatosis 1989;2:322–25.