A Child with Prolonged Fever, Chronic Raised ICP and Vision Loss-No its not TBM!

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Introduction

Primary diffuse leptomeningeal melanomatosis (PDLM) is an extremely rare, aggressive malignant neoplasia of the central nervous system (CNS). We report the **first case of pediatric PDLM** from India, with a review of literature. To date, only 15 pediatric cases have been reported in the literature.

Methodology

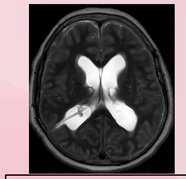
Our case is a description of a rare pediatric neurological condition.

History and Examination

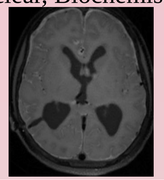
- A 13years old male child presented with complaints of intermittent **fever, vomiting and headache** for last 2years and now had **progressive visual impairment**, seizures and abnormal behaviour for last 5months.
- There was no other significant history and the child had no tuberculosis contact.
- On examination child had normal vitals and anthropometry but **impaired vision**. Formal visual assessment revealed finger counting at 1metre in left eye and only perception of light present in the right eye.
- There was **hearing impairment** present in bilateral ears R>L
- Other systemic examination- WNL

Investigations

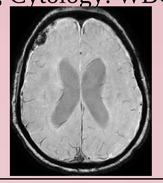
CSF examination: Appearance- clear, Biochemistry: S 66 **P 527**, Cytology: WBC-3 (predominant LMN) RBC-25



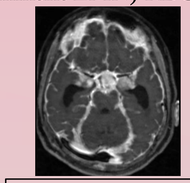
T2 hypointense leptomeningeal+ pachymeningeal thickening



T1 hyperintense leptomeningeal+ pachymeningeal thickening



Blooming of SWI



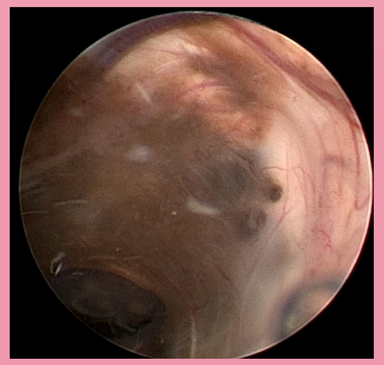
Diffuse thick leptomeningeal and pachymeningeal enhancement

MRI Brain

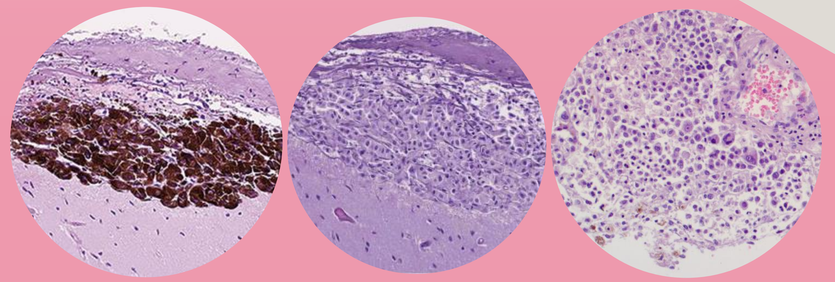
- ❑**No tuberculomas**
- ❑**Start of disease with convexity predominant leptomeningitis.**

FDG-PET Scan- Metabolically active nodular enhancing lesion along the sella and bilateral perisylvian region with pachymeningeal and leptomeningeal enhancement and diffuse left hemicerebral hypometabolism

Endoscopy guided brain biopsy



Histopathology-HMB-45 and MelanA



Final Diagnosis
Diffuse Leptomeningeal melanomatosis

Conclusion

- **Diagnosing PDLM can be daunting in light of its slow but malignant progression mimicking TBM**
- **However, the absence of any supportive microbiological evidence and failure to respond to the standard ATT with subsequent progression of the symptoms should prompt the need for finding an alternative diagnosis and raise suspicion of a condition like leptomeningeal melanomatosis.**
- **A targeted molecular diagnosis and precision medicine may provide a favorable outcome in children with PDLM**

References

Syoko Yamauhci, Tetsuhiro Fukuyama, Yuji Inaba, Hideko Nakajima, Yuka Hattori, Yosuke Miyairi, Yoshihumi Ogiso. Diagnostic challenges of primary diffuse leptomeningeal melanomatosis in early adolescence: A case report, Brain and Development. Volume 44, Issue 6,2022;421-425,