Dynamic Magnetic Resonance (DMR) Study of the Hypothalamic-Pituitary Region in Patients with Langerhans Cell Histiocytosis (LCH)

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Purpose

- Children with Langerhans cell histiocytosis (LCH) are at risk of developing either central diabetes insipidus (DI), or multiple pituitary hormone deficiency.
- Maghnie M et al reported the use of dynamic magnetic resonance (MR) study in the assessment of pituitary vasculopathy in children with hypopituitarism and DI caused by various diseases including LCH.
- The natural history of progressive damage to the pituitary gland that leads to multiple hormone deficiency in LCH patients has never been clarified.
- We would like to report our experience on the use of magnetic resonance (MR), including study after dynamic gadolinium infusion, in the assessment of changes in the pituitary glands in LCH patients with or without pituitary deficiency.
Methods and Materials

- 13 patients (established or newly diagnosed) with biopsy proven LCH were prospectively recruited for the current study.
- Non-contrast MR study of the pituitary region was performed (1.5T GE Signa Advantage MR System), followed by a dynamic sagittal fast gradient echo (FSPGR) examination with 30 frames and 4 seconds interval over 111 seconds after intravenous injection of Magnevist (0.2ml/kg).
- Computerized reconstruction of the pattern of enhancement of straight sinus, posterior and anterior pituitary were done by measuring the mean pixel density of the structures of interest.
- The enhancement pattern was reconstructed and displayed in graphic form, with the mean pixel density plotted against time (second).
- The straight sinus was used as reference.
- The initial time of enhancement of straight sinus and posterior pituitary lobe, and the time duration from the onset to maximal enhancement of the entire posterior pituitary lobe were studied.
- 10 children without endocrine dysfunction or LCH, matched for age, were studied as controls.
- Two patients with proven panhypopituitarism due to other causes were also included as controls.
- Differences in enhancement time between patients and control subjects were analysed.
- Urinary concentration test was performed in all patients.
- Other endocrinological workup were applied as clinically indicated.
Results

- 7 out of 13 studied patients (7 months to 29 years old) had endocrinopathies and MR changes
- Absent posterior pituitary hyperintense signal in T1W images (7/7)
- Thickened pituitary stalk (3/7).
- All had diabetes insipidus (6 complete and 1 partial)
- 2 cases had panhypopituitarism
- 3 had growth hormone deficiency.
- All 7 patients (7/7) with either partial or complete DI showed delay in onset of enhancement (> 7.5 second) and exceptional long enhancement time (>120 seconds)
- 5 cases of mild to moderate delay in enhancement
- Pattern of enhancement of the posterior pituitary followed the pattern of the straight sinus and was similar to the controls
- 2 cases of panhypopituitarism showed atrophic pituitary gland and stalk, and the enhancement pattern were totally abnormal.
- This finding was comparable to the control with pituitary atrophy.
- 6 LCH patients without endocrinopathy had normal MR findings and were comparable to the control (normal).
- 10 control group patients without LCH had normal pituitary stalk and no delay in contrast enhancement on dynamic study.
- The normal pattern of enhancement of the posterior pituitary lobe was the same as the straight sinus.
- There should be a rapid initial rise followed by a peak, plateau and then gradual decrease in contrast enhancement.
- Initial enhancement of the posterior pituitary lobe occurred within 4.7 seconds after the initial enhancement of the straight sinus was noticed in the normal control group.
- Atrophic pituitary gland showed totally abnormal pattern of enhancement.
- From the mean pixel density plots of contrast enhancement, no notable discrepancy in enhancement patterns of straight sinus and posterior pituitary lobe was observed in the normal control group.
- However, the enhancement time varied from one patient to another.
- When using enhancement time of posterior pituitary lobe (Tpp) only as a variable in the Kruskal Wallis test, p<0.001.
- But specificity of this test is only 74% when a monitoring time interval of 20 seconds is used as a cut-off value.
- By increasing the cut-off value to 60 seconds, specificity was found to be 95%.
• The sensitivity dropped from 100% to 85%.
• Varying the cut-off value does not get rid of the effect of blood flow on the enhancement time measurement.
• In order to minimize the effect of blood supply, it is necessary to have a reference site to indicate the speed of blood flow; and straight sinus is chosen in our study.
• Because of the relationships between blood flow and endocrinopathies are not known, for the sake of simplicity, it is an attempt to assume the enhancement time to be a sum term or product term.

• In the sum term's hypothesis, a monitoring index known as Authentic Enhancement Time (AET) is put forward and is defined as a difference between Tpp and the enhancement time of straight sinus (Tss). i.e. AET = Tpp-Tss.
• In the Kruskal Wallis test, AET demonstrates itself to be a good indicator in predicting endocrinopathies (p<0.001) in the entire patient group.
• Using a cut-off value of 7.5 seconds, the specificity is improved and has a value of 95% without degrading the sensitivity.

• In the product term's hypothesis, another monitoring index known as Enhancement Time Ratio (ETR) is put forward in order to make a comparison between the two hypotheses. i.e. ETR is defined as a ratio of Tpp to Tss.
• ETR also proves itself to be a good indicator in predicting endocrinopathies (p<0.001).
• Sensitivity and specificity of ETR are found to be 100% and 95% respectively when using 1.7 as a cut-off value.
Images for this section:

Fig. 1

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Dynamic enhancement pattern of LCH patient with DI

Fig. 2

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Dynamic enhancement pattern of Control Group

Fig. 3

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Conclusion

- All 7 patients with pituitary hormone deficiency have MR changes in contrast to none in those without.
- Absence of posterior bright spot is the most consistent MR findings but is non-specific, occasionally seen in normal subjects.
- Presence of thickened stalk and/or delayed contrast enhancement are more specifically associated with endocrinological manifestation in LCH.
- In our study, delay in onset of enhancement is even more sensitive than thickened stalk in DI cases.
- Both ETR and AET are statistically significant in indicating whether or not LCH patient has endocrinopathies.
- ETR (>1.7) gives a better sensitivity (100%) and specificity (95%).
- Dynamic study may therefore offer additional evidence of hypothalamic-pituitary involvement in LCH.
- Longitudinal study of newly diagnosed LCH patients with/without DI would be needed to further clarify whether such MR changes can predict subsequent development of endocrinopathy.
References


