IgG4-Related Disease Presenting as Hypertrophic Pachymeningitis : A Diagnostic and Therapeutic Challenge

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INTRODUCTION

- IgG-4 related disease (IgG4-RD) is a lymphoproliferative disorder affecting many organs.
- Most common affected organs are pancreas, salivary glands, retroperitoneum and lymph nodes.
- CNS involvement is uncommon and mostly present with hypertrophic pachymeningitis (HP) and hypophysitis

METHODOLOGY

- Prospective study of 2 years of duration
- Patients were assessed with a detailed history and clinical examination
- 7 patients were included
- Investigations included – routine lab investigations, ANA, ENA, Vasculitis profile, CEMRI brain, CSF cytology-biochemistry & infective profile, PET-CT whole body, work up for TB & sarcoidosis

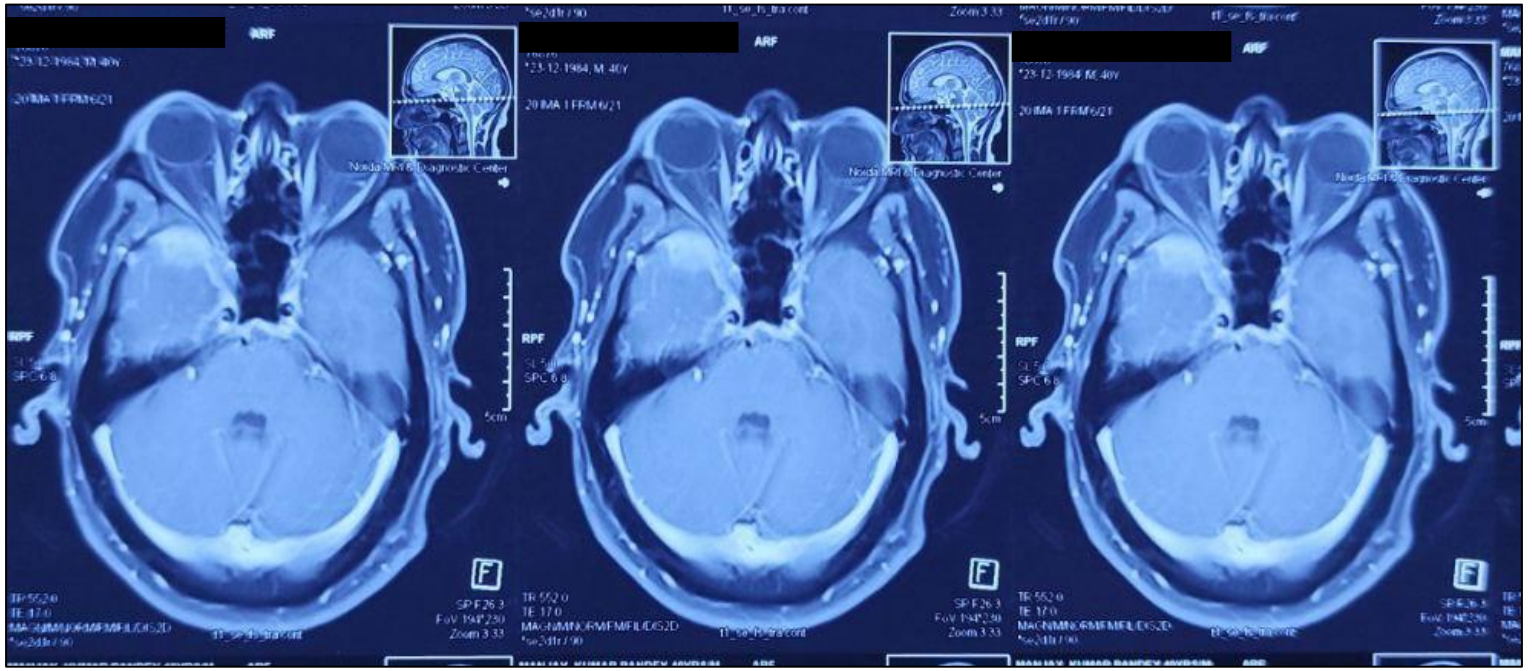
Comprehensive diagnostic criteria for IgG4- RD 2011

1. Clinical/Radiological evidence of diffuse/localized swelling or mass in single or multiple organ
2. Elevated levels of IgG4 (>135 mg/dL)
3. Histopathology study shows following findings :
   - Lymphoplasmacytic infiltration & fibrosis
   - IgG4 +ve plasma cell infiltration : IgG4/IgG +ve cells >40% and IgG4 +ve plasma cells/HPF >10

A. Possible – 1 + 2
B. Probable – 1 + 3
C. Definite – 1 + 2 + 3

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7
Age/Gender	74/Female	56/Male	61/Female	56/Male	40/Male	36/Female	43/Female
Presenting complaints	• Headache • Left eyelid drooping • Blurring of vision in left eye • Numbness over left face	• Headache • Diplopia • Right eyelid drooping	• Headache • Blurring of vision in left eye	• Left eyelid drooping • Double vision	• Headache	• Headache • Numbness over left face • Decreased hearing in left ear	• Decreased hearing b/l • Left facial weakness • Double vision • Speech & swallowing difficulty
Past History	Similar complaints in Feb 2024 – improved without treatment	NA	Left eye ptosis in Jan 23, Feb 24, July 24 – improved with steroids	NA	• Vitreous hemorrhage in 2007, 2011 • Right Cavernous sinus syndrome in 2020 – improved with oral steroids	Left eyelid drooping and diplopia – improved without treatment	• Sept'20 – Rt. Ear tinnitus • Jan'24 – b/l hearing loss • April'24 – left facial weakness • June'24 – speech and swallowing difficulty • Partial recovery with steroids

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7
On examination	Orbital apex syndrome	Right Cavernous sinus syndrome	Left cavernous sinus syndrome	Left Orbital apex syndrome	Headache with h/o cranial nerve palsy	Cranial nerve palsy (5^{th}, 8^{th}) & h/o 3^{rd} CN palsy	Recurrent multiple cranial nerve palsy
Serum IgG4	2.27	5.24	3.66	2.03	3.20	3.23	2.5
CEMRI Brain	Enhancement of left cavernous sinus, orbital apex and enhancement of left trigeminal nerve	Enhancement of Right cavernous sinus	Dural thickening	Left cavernous sinus and Left orbital apex enhancement	Extra-axial enhancement over right anterior middle cranial fossa along greater & lesser wing of sphenoid, orbital apex	Dural enhancement along left CP angle, left tentorium cerebelli, left falx cerebri along left temporal lobe	Pachymeningeal thickening over bilateral temporal lobe
PET-CT					FDG avid LN in left inguinal region		Increased FDG uptake in caecum and ascending colon
Treatment	IV MPS f/b oral steroids	IV MPS f/b oral steroids	IV MPS f/b oral steroids	Oral steroids	IV MPS f/b oral steroids + Rituximab	IV MPS f/b oral steroids	IV MPS f/b oral steroids + Rituximab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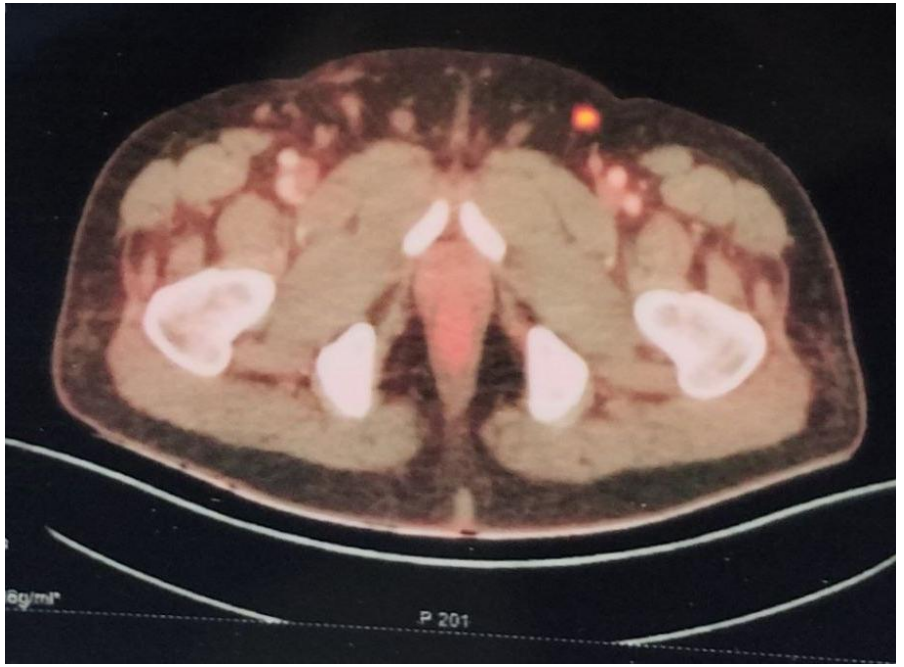
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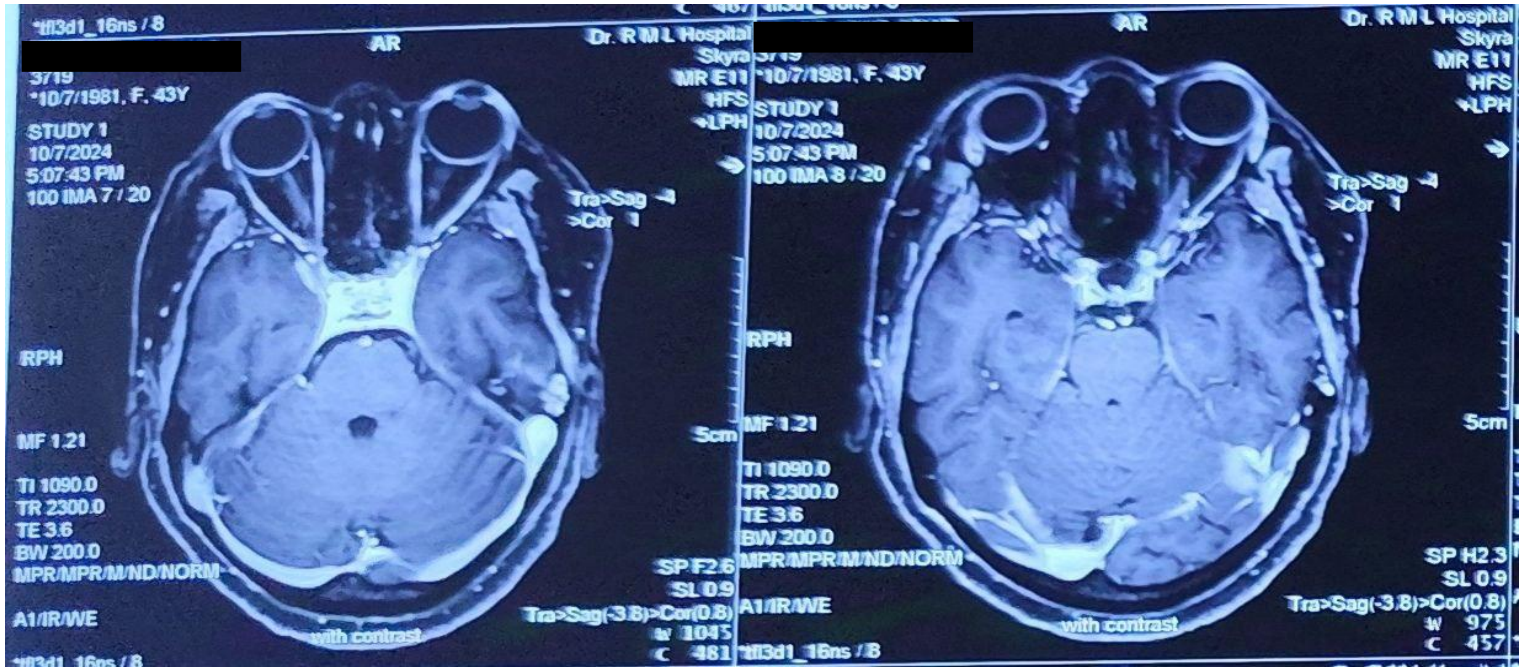
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STUDY 1
10/7/2024
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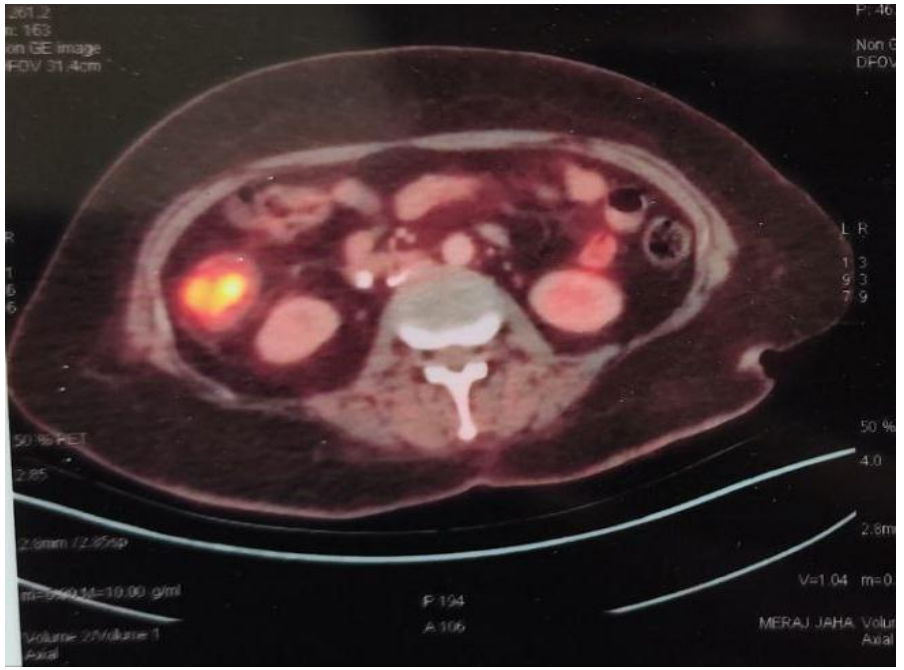
RPH
MF 1.21
TI 1090.0
TR 2300.0
TE 3.6
BW 200.0
MPR/MPR/M/ND/NORM
A1/R/WE
with contrast
SP F2.6
SL 0.9
Tra>Sag(-3.8)>Cor(0.8)
5cm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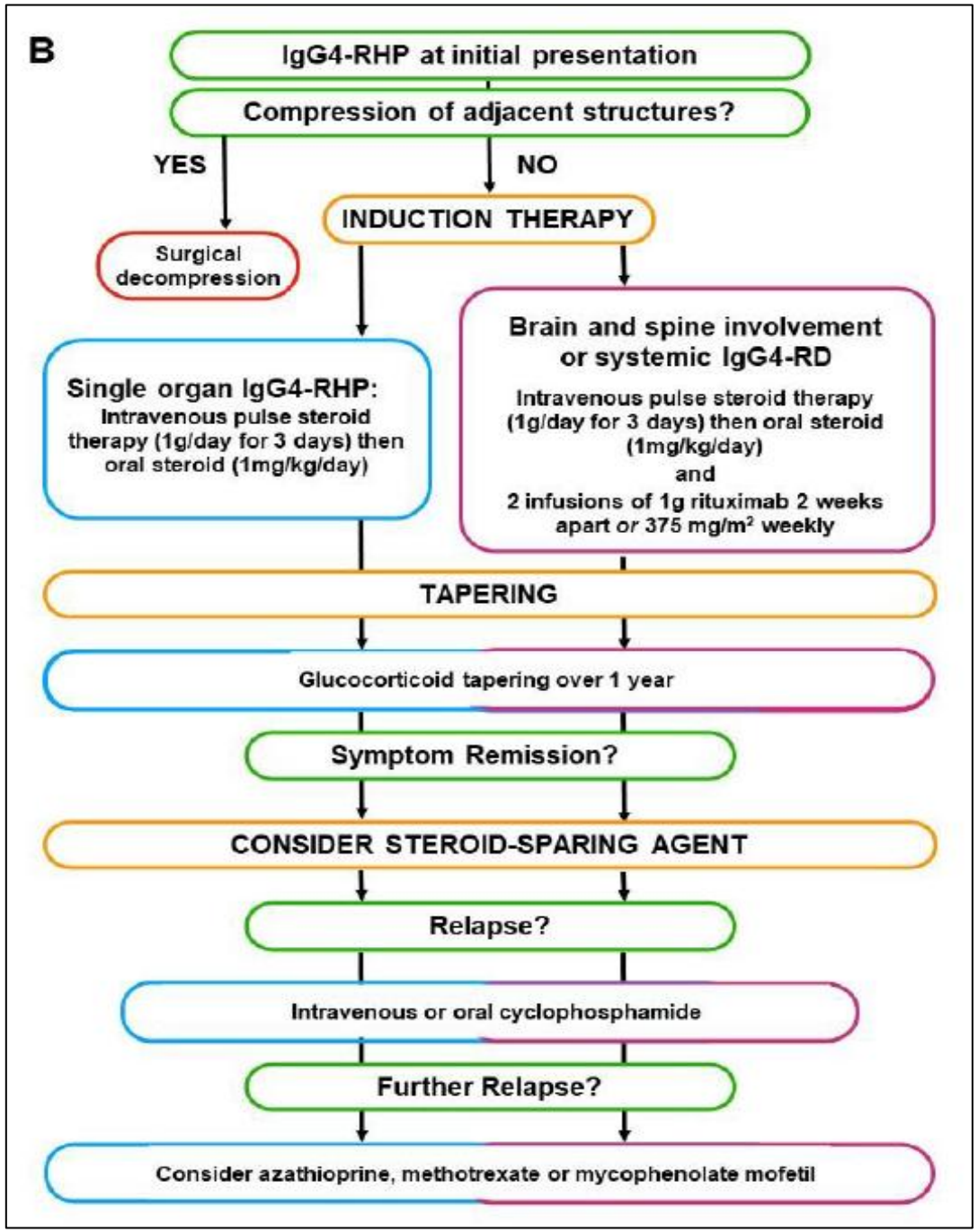

CONCLUSION

- Corticosteroid responsiveness is characteristic of IgG4-RD
- It can mimic infection, inflammation and malignancy
- IgG4-RHP characterized by the lack of extra-neurological organ involvement and systemic signs
- Differential diagnosis include – lymphoma, sarcoidosis, tuberculosis, granulomatosis with polyangiitis and Langerhans-cell histiocytosis.

REFERENCES

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