

Suppl 1. Literature search on PubMed for published cases of IHSP since 2009

| Study | Age (years) | Sex | Acute/progressive | Ataxia | Weakness | Sensory symptoms | Bowel/bladder | Neck/back pain | UMN signs |
|-----------------------------|-------------|-----|-------------------|--------|----------|------------------|---------------|----------------|-----------|
| Alsulaiman A 2020 [4] | 38 | F | P | - | + | - | - | + | Yes |
| Bang JH et al 2015 [5] | 67 | F | P | + | - | - | - | + | Yes |
| Fakuda A et al 2017 [6] | 51 | M | P | - | + | - | - | + | NA |
| TK Jee et al 2014 [7] | 58 | F | P | - | + | - | - | - | No |
| Kim JH et al 2011 [8] | 55 | F | P | - | + | + | + | + | NA |
| | 45 | F | A | - | + | - | - | - | NA |
| Lowden MR et al 2009 [9] | 42 | F | P | - | - | + | - | - | Yes |
| Lucas Lodomiro AM 2020 [10] | 19 | F | A | + | + | - | - | - | NA |
| Olubajo F et al 2017 [11] | 51 | F | A | - | + | + | + | - | No |
| Chen et al 2012 [12] | 49 | M | P | - | - | - | - | + | No |
| Hsu et al 2015 [13] | 41 | M | P | - | + | + | - | - | NA |
| Van der pal et al 2015 [14] | 58 | F | P | - | + | + | + | + | Yes |
| Park JY et al 2020 [15] | 79 | F | P | + | + | - | - | + | No |
| Qin et al 2015 [16] | 44 | F | A | - | + | + | + | + | Yes |
| Kutty RK et al 2019 [17] | 29 | F | P | - | + | - | - | + | Yes |
| Ranasinghe et al 2011 [18] | 42 | F | P | + | - | + | - | - | Yes |
| | 77 | F | P | + | + | + | - | - | Yes |
| | 65 | M | P | + | - | + | - | - | No |
| Takahashi H et al 2010 [19] | 67 | M | P | - | + | + | + | - | Yes |
| Park TJ et al 2016 [20] | 51 | F | P | - | + | + | - | + | Yes |

| | | | | | | | | | |
|------------------------------|----|---|---|---|---|---|---|---|-----|
| Tosa M et al 2015 [21] | 63 | M | P | + | + | + | + | + | No |
| Maheshwari V et al 2016 [22] | 42 | M | P | - | + | + | - | + | Yes |
| Present case | 40 | M | P | - | + | + | - | - | NO |

Abbreviations: M: male, F: female, A (acute): disease onset to disability within 24 hours, P (progressive): disease onset to disability more than 24 hours, (+): present, (-): absent or not mentioned, NA: not available.

| Study | Protein | Cells | ESR | CRP | RA |
|-----------------------------|---------|---------------------------------|-----|-----|----|
| Alsulaiman A 2020 [4] | - | 60, lymphocyte predominant | - | - | - |
| Bang JH et al 2015 [5] | - | - | - | - | - |
| Fakuda A et al 2017 [6] | - | - | - | - | - |
| TK Jee et al 2014 [7] | - | - | + | - | - |
| Kim JH et al 2011 [8] | - | - | + | + | - |
| | - | - | + | + | - |
| Lowden MR et al 2009 [9] | - | - | + | - | - |
| Lucas Lodomiro AM 2020 [10] | - | - | - | - | - |
| Olubajo F et al 2017 [11] | - | - | - | + | - |
| Chen et al 2012 [12] | - | - | - | - | - |
| Hsu et al 2015 [13] | - | - | - | - | - |
| Van der pal et al 2015 [14] | - | - | - | - | - |
| Park JY et al 2020 [15] | - | - | + | + | + |
| Qin et al 2015 [16] | 6,160 | Lymphocyte predominant WBCs 150 | - | - | - |
| Kutty RK et al 2019 [17] | 150 | Lymphocyte predominant | - | - | - |
| Ranasinghe et al 2011 [18] | - | - | - | - | - |
| | - | - | - | - | - |
| | - | - | - | - | - |
| Takahashi H et al 2010 [19] | - | - | + | + | + |

| | | | | | | |
|------------------------------|-------|-----------------------------|--|---|---|---|
| Park TJ et al 2016 [20] | - | - | | + | + | - |
| Tosa M et al 2015 [21] | 243 | 20, lymphocyte predominant | | + | + | - |
| Maheshwari V et al 2016 [22] | - | - | | - | - | - |
| Present case | 1,620 | 188, lymphocyte predominant | | - | - | - |

Abbreviations: ESR: erythrocytic sedimentation rate, WBC: white blood cells, CRP: C-reactive protein, RA: rheumatoid arthritis factor.

| Study | T1 | T2 | Contrast enhancement | Location | Spinal segments |
|-----------------------------|------|------|----------------------|-----------------|-----------------|
| Alsulaiman A 2020 [4] | NA | Hypo | + | Ant and post | C2-C7 |
| Bang JH et al 2015 [5] | Hypo | Hypo | + | Posterior | occiput to C2 |
| Fukuda A et al 2017 [6] | Hypo | Hypo | + | Posterior | C2-T1 |
| TK Jee et al 2014 [7] | Hypo | Hypo | - | Posterior | C5-C7 |
| Kim JH et al 2011 [8] | Hypo | Hypo | NA | NA | C6-T7 |
| | Hypo | Hypo | NA | Posterior | C1-C4 |
| Lowden MR et al 2009 [9] | NA | Hypo | NA | Anterior | T2-T5 |
| Lucas Lodomiro AM 2020 [10] | NA | NA | NA | Posterior | C7-T4 |
| Olubajo F et al 2017 [11] | Hypo | Hypo | NA | Ant and post | T3-T8 |
| Chen et al 2012 [12] | NA | NA | + | Anterior | T1-T3, L4 |
| Hsu et al 2015 [13] | Hypo | Hypo | + | Circumferential | T2-T4 |
| Van der pal et al 2015 [14] | Hypo | Hypo | + | Circumferential | C7-T7 |
| Park JY et al 2020 [15] | Hypo | Hypo | NA | Posterior | C4-T4 |
| Qin et al 2015 [16] | Iso | Hypo | + | Anterior | T2-T6 |
| Kutty RK et al 2019 [17] | Hypo | Hypo | + | Anterior | C7-T1 |
| Ranasinghe et al 2011 [18] | Hypo | Hypo | + | Circumferential | T1-T6 |
| | NA | NA | + | Circumferential | C4-T11 |
| | NA | NA | + | Circumferential | T4-T9 |
| Takahashi H et al 2010 [19] | Iso | Iso | + | Anterior | C3-C7 |

| | | | | | |
|------------------------------|------|------|---|-----------------|---------|
| Park TJ et al 2016 [20] | Hypo | Hypo | + | Ant and Post | T2-T9 |
| Tosa M et al 2015 [21] | Hypo | Hypo | + | Circumferential | T11-T12 |
| Maheshwari V et al 2016 [22] | NA | NA | + | Circumferential | C2-C5 |
| Present case | Hypo | Hypo | + | Circumferential | C2-T2 |

Abbreviations: Hypo: hypointense, Iso: isointense, Ant and Post: anterior and posterior.

| Study | Drugs | Surgery | Relapse (in weeks) | Prognosis |
|-----------------------------|------------------|---------|--------------------|-----------|
| Alsulaiman A 2020 [4] | MPS**+PRED** | NO | 24 | I |
| Bang JH et al 2015 [5] | NO | YES | - | PI |
| Fukuda A et al 2017 [6] | PRED | NO | - | I |
| TK Jee et al 2014 [7] | MPS | YES | - | PI |
| Kim JH et al 2011 [8] | DEXA+MPS*+PRED** | YES | 4 | I |
| | DEXA | YES | - | NI |
| Lowden MR et al 2009 [9] | NA | NA | - | NA |
| Lucas Lodomiro AM 2020 [10] | NO | YES | - | I |
| Olubajo F et al 2017 [11] | IVIG+ DEXA | YES | - | PI |
| Chen et al 2012 [12] | STEROIDS | YES | - | I |
| Hsu et al 2015 [13] | NA | NA | - | NA |
| Van der pal et al 2015 [14] | PRED*+MTX* | YES | 12 | I |
| Park JY et al 2020 [15] | DEXA | YES | - | NI |
| Qin et al 2015 [16] | MPS+PRED | YES | 1 | NI |
| Kutty RK et al 2019 [17] | MPS*+PRED** | YES* | 6 | I |
| Ranasinghe et al 2011 [18] | DEXA+PRED | YES | - | PI |
| | DEXA+PRED | YES | - | PI |
| | DEXA | YES | - | NI |
| Takahashi H et al 2010 [19] | PRED | YES | - | I |

| | | | | |
|------------------------------|----------------|-----|---|----|
| Park TJ et al 2016 [20] | STEROID**+MTX* | YES | 8 | PI |
| Tosa M et al 2015 [21] | PRED | YES | - | I |
| Maheshwari V et al 2016 [22] | NO | YES | - | I |
| Present case | PRED | NO | - | I |

*Indicate drug used while relapse only, **Indicate drugs used while initial treatment and during relapse.

Abbreviations: MPS: methylprednisolone, PRED: prednisone, DEXA: dexamethasone, IVIG: intravenous immunoglobulin, MTX: methotrexate, I: completely improved, PI: partial improvement with some remaining neurologic disability, NI: no improvement.

References

4. Alsulaiman A. Idiopathic Hypertrophic Spinal Pachymeningitis: A Diagnostic Challenge: A Case Report and Review of the Literature. *J Neurosci Rural Pract.* 2020 Jan;11(1):175-177.
5. Bang JH, Cho KT, Kim EJ. Idiopathic Hypertrophic Pachymeningitis in the Craniocervical Junction. *Korean J Spine.* 2015 Sep;12(3):169-72.
6. Fukuda A, Punaro E, Rogério F, de Souza Queiroz L, Reis F. Idiopathic hypertrophic pachymeningitis as a rare cause of cervical compressive myelopathy. *J Craniovertebr Junction Spine.* 2017 Oct-Dec;8(4):387-389
7. Jee TK, Lee SH, Kim ES, Eoh W. Idiopathic hypertrophic spinal pachymeningitis with an osteolytic lesion. *J Korean Neurosurg Soc.* 2014 Aug;56(2):162-5.
8. Kim JH, Park YM, Chin DK. Idiopathic hypertrophic spinal pachymeningitis : report of two cases and review of the literature. *J Korean Neurosurg Soc.* 2011 Oct;50(4):392-5.
9. Lowden MR, Gill D. Teaching NeuroImage: idiopathic hypertrophic spinal pachymeningitis. *Neurology.* 2009 Feb 3;72(5):e27
10. Araújo Melo LL, Daher MT, Gonçalves MVM, Freitas MB. Idiopathic Hypertrophic Spinal Pachymeningitis: A Case Report. *Rev Bras Ortop (Sao Paulo).* 2021 Aug 13;57(3):521-523
11. Olubajo F, Yermakova T, Highley JR, Arzoglu V. Concomitant idiopathic hypertrophic spinal pachymeningitis and Guillain-Barré syndrome in a patient: coincidence or a triggering mechanism? *J Neurosurg Spine.* 2017 Sep;27(3):335-340.
12. CHEN H, LI Y, MEHRA S et al Idiopathic Hypertrophic Pachymeningitis as a Rare Cause of Spinal cord compression Journal of Medical Cases, North America3, 2012 Aug 3;4 :267-269
13. Hsu HT, Hsu SS, Chien CC, Lai PH. Teaching NeuroImages: Idiopathic hypertrophic spinal pachymeningitis mimicking epidural lymphoma. *Neurology.* 2015 Mar 3;84(9):e67-8

14. van der Pol CB, Chakraborty S, Côté I, Humphrey-Murto S, Michaud J. Case 216: hypertrophic spinal pachymeningitis. Radiology. 2015 Apr;275(1):303-7.
15. Park JY, Choi I, Khil EK, Kim WJ, Shin IY. Idiopathic Hypertrophic Spinal Pachymeningitis with Spinal Cord Lesion: A Case Report. Korean J Neurotrauma. 2020 Jun 5;16(2):367-373.
16. Qin LX, Wang CY, Hu ZP, Zeng LW, Tan LM, Zhang HN. Idiopathic hypertrophic spinal pachymeningitis: a case report and review of literature. Eur Spine J. 2015 May;24 Suppl 4:S636-43.
17. Kutty RK, Sreemathyamma SB, Sivanandapanicker JL, Peethambaran A. Idiopathic Hypertrophic Spinal Pachymeningitis: A Rare Cause of Spinal Cord Compression. Neurol India. 2019 Sep-Oct;67(5):1380-1385
18. Ranasinghe MG, Zalatimo O, Rizk E, Specht CS, Reiter GT, Harbaugh RE, Sheehan J. Idiopathic hypertrophic spinal pachymeningitis. J Neurosurg Spine. 2011 Aug;15(2):195-201.
19. Takahashi H, Wada A, Yokoyama Y, Ishii M, Shibuya K, Suguro T. Idiopathic hypertrophic spinal pachymeningitis: a case report. J Orthop Surg (Hong Kong). 2010 Apr;18(1):113-7
20. Park TJ, Seo WD, Kim SY, Cho JH, Kim DH, Kim KH. Effective Response of Methotrexate for Recurrent Idiopathic Hypertrophic Spinal Pachymeningitis. Korean J Spine. 2016 Dec;13(4):200-203.
21. Tosa M, Hara M, Morita A, Ninomiya S, Ebashi M, Kamei S, Maseda M, Tokuhashi Y, Hemmi A, Nemoto N. Idiopathic Hypertrophic Spinal Pachymeningitis. Intern Med. 2015;54(15):1923-6.
22. Maheshwari V, Gill M, Kumar M, Srivastava C. Idiopathic hypertrophic cervical pachymeningitis - A rare report. Neurol India. 2016 Sep-Oct;64(5):1038-40.