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Case Study

AN AYURVEDIC APPROACH IN THE MANAGEMENT OF *HIRAYAMA* DISEASE

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ABSTRACT

Hirayama disease is a self limiting condition affecting the motor component of distal upper limbs without any sensory impairment. It mainly affects young males of Southeast Asia with an increased prevalence in India and Japan. **Clinical findings and Diagnosis:** A 32 year old male patient approached the OPD with a known case of *Hirayama* disease for the past 5 years. By analyzing the condition and *Dosha* involved Ayurvedic treatment, both internal medication and external treatments were developed. **Outcome:** After a course of 28 days treatment divided into 9 and 19 days with a gap of 1 week, the patient showed significant improvement in the clinical signs and symptoms. **Conclusion:** *Hirayama* disease can be correlated to *Vatavyadhi* spectrum disorders like *Bahusosha* (a *Vataja nanatmaja vyadhi*). The main aim of treatment is to relieve the vitiated *Vata* and nourish the depleted *Mamsa dhatu*.

INTRODUCTION

Hirayama disease is a rare cervical myelopathy seen in young males in their teens and twenties characterized by muscular weakness and atrophy of distal upper limbs unilaterally or bilaterally without sensory loss which progresses for 1 or 2 years before plateauing and eventually showing an abrupt arrest.[1] The disease was first brought to attention by Keizo *Hirayama* and his colleagues. In 1959 they published 12 cases distinguishing a new entity from what was originally thought to be a type of progressive and degenerative motor neuron disease.[2] The underlying pathology of the disease was brought to light after the first autopsy case of Hirayama disease done in 1982. The exact cause of the disease is unknown. Several mechanisms were proposed. *Hirayama* suggested a disproportional growth between the vertebral column and the contents of the spinal canal, especially the dural sac, during juvenile growth spurt leads to forward dural sac displacement during neck flexion. The posterior epidural ligaments are present between posterior dura mater and ligamentum flavum. These ligaments have a tendency to be abundant at C1 through C2, decreased below C2 and sparse at C6 and C7. It is assumed that these ligaments may contribute to resistance against the separation of the posterior dura mater from the ligamentum flavum. Unequal distribution or lack of these ligaments may be the essential cause of asymmetric cord compression. Strenuous activities that requires sustained or repeated neck flexions such as writing at a desk or playing a musical instrument were frequently noted in patients.[2]

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Patient Information

A 32 year old male patient reported to out-patient department with insidious onset of weakness of bilateral upper limbs, left side since 5 years followed by right side since 1 year. The patient was apparently healthy till the age of 27 years. He was working as an interlock tile worker from the age of 18 years. 5 years back, he started to experience weakness of left thumb that lasted for about 2 minutes which gradually increased up to 1 hour. Gradually the weakness progressed to hand and forearm muscles and faced difficulty to perform daily activities like washing hands, eating food, holding nail, buttoning of clothes etc. He had also

noticed wasting of muscles of hand and forearm which was gradually progressive in nature. He also experienced tremor of left hand and worsening of symptoms when exposed to cold. Later he started to experience similar symptoms in right hand and forearm also. Routine laboratory investigations were within normal limits. While undergoing evaluation, the patient received the diagnosis of Hirayama Disease or Monomelic amyotrophy (MMA) based on the clinical and radiological characteristics seen in MRI. He took allopathic medications for around one year and got no significant improvement. Then he was advised to undergo surgical intervention and since he was reluctant to undergo surgery he came for Ayurvedic treatment, first from outpatient department and then he got admitted in the inpatient department for better management. He had no significant past medical and surgical histories. No history of exposure to toxins/allergies/addictions reported. Family history was not significant.

Clinical Findings

Neurological examination revealed a fully conscious, alert with normal higher mental functions and cranial nerves. Motor examination showed reduced grip strength in both hands and reduced muscle power in left forearm. Muscle atrophy-thenar, hypothenar, interossei, wrist and forearm flexors were more in left side. Muscle bulk in forearm and upper arm were decreased in left side when compared to right side. Adduction of thumb was not possible in left hand and opposition of thumb was not possible in both hands (Table 3). The sensory examinations showed normal sensory functions and superficial and deep tendon reflexes of both upper and lower limbs were normal. Muscle tone of lower limb was also normal.

Diagnostic Assessment

MRI showed anterior displacement of the posterior wall of dural sac from C4 to D2 level with expanded crescentic enhancing posterior dural space, maximum at the C6 level on flexion.

Therapeutic Intervention

We can identify the involvement of *Vata dosha* from the clinical features exhibited in *Hirayama* disease. Combining the principles of management of *Vatavyadhi* and *Bahusosha*, the management was designed with both internal and external treatments (Table no.2). Patient got admitted in the hospital IPD in two schedules with a gap of 1 week in between.

During the first course of treatment he was treated with *Takrapana* for 2 days, followed by *Udwartana* for 5 days and *Snehapana* for 2 days to begin with initial *Abhyantara snehana*. (Table 1) In the second course of treatment, he was treated with *Udwartana* and *Valuka sveda*. It was then followed by *Abhyangam* and *Ushmasveda* for a day to begin with the initial *Bahya snehana*. Afterwards, *Patra potala sveda* was done for 4 days followed by *Shashtika pinda sveda* for 5 days. During the course of SPS, on the last 3 days *Ksheeravasti* was administered. (Table 2)

Table 1: Therapeutic intervention (1st course)

Days	Treatment	Medicines
1 - 2 days	*Takrapanam*	*Takram + Brihath Vaisvanara Churnam*
3 - 7 days	*Udvartanam*	*Kulatha Churna*
9 - 10 days	*Snehapanam*	*Vidaryadi ghrita*

Table 2: Therapeutic intervention (2nd course)

Days	Treatment	Medicines
1 - 3 days	*Udvartana*	*Kulatha churna*
4 - 8 days	*Valuka svedam*	
9th day	*Abhyangam* and *Ushma sveda*	*Dhanvantaram tailam*
10 - 14 days	*Patrapotala sveda*	*Dhanvantaram tailam*
15 - 19 days	*Shashtika pinda sveda*	*Panchatiktakam kashaya*
16 - 19 days	*Ksheera vasti*	*Tiktaka ghrita*

Follow-Up and Outcomes

After the treatment of 29 days, the patient showed significant improvement in symptoms and motor functions of bilateral hand and forearm. Hand grip, muscle power of forearm, adduction of fingers and opposition of thumb got significantly improved (Table 3).

Table 3: Assessment and Outcome

	Before treatment		After treatment	
	Right	**Left**	**Right**	**Left**
Hand grip	4/5	3/5	5/5	4/5
Muscle power-forearm	5/5	4/5	5/5	5/5
Muscle power-upper arm	5/5	5/5	5/5	5/5
Muscle atrophy-thenar, hypothenar, interossei, wrist and forearm flexors	+	++	+	++
Muscle bulk-forearm(5 inches from olecranon process-in cms)	25.5	22.5	25.5	23
Muscle bulk-upper arm(5 inches from olecranon process-in cms)	31.2	31	31.2	31
Adduction of fingers	Possible	Not possible	Possible	Possible
Opposition of thumb	Not possible	Not possible	All possible	Possible upto middle finger

DISCUSSION

Hirayama disease is a rare cervical myelopathy which is self limiting. This condition is best diagnosed with flexion MRI of cervical spine. In Ayurveda this disease condition can be better related to diseases of *Vata*. *Vatavyadhi* spectrum disorders like *Bahusosha* (a *Vataja nanatmaja vyadhi*) should be considered while planning the treatment. In Hirayama disease, due to *Nidana* like *Ativyayama* (repeated neck flexion), *Dhatukshaya* (loss of anterior horn cells) and *Margavarodha*, *Vata* gets vitiated leading to symptoms like *Mamsa kshaya, Bala kshaya, Kampa* etc. *Takrapana*, *Udwartana* and *Valuka sveda* in the initial phase of treatment helps to mitigate the *Margavarodha* of *Vata*. *Takra* possess qualities like *Vikasi* and *Ruksha guna*, *Amla rasa* and *Deepana* property which aid in mitigating the *Margavarodha*.[3] *Udvartanam* is *Vata kapha hara* and hence provides *Rukshana* as well as *Vataharatva*.[4] After *Rukshana*, *Snehapana* with *Vidaryadi ghrita* provides *Brihmana* and alleviation of *Vata* localised in *Urdhva bhaga*.[5] Subsequent *Bahya snehana* will also provide the same effect. Treatments like *Patra potala sveda* and *Shashtika pinda sveda* help to mitigate the *Vata dosha* and nourish the *Dhatus* which results in *Pushti* of *Mamsa* and *Bala*. *Ksheera vasti* is a *Niruha* in *Mridu* form which possess qualities like *Vatasamana*, *Bala vardhana* and *Snigdha guna*.[6] When done with *Tiktaka ghrita* it possess excellent *Brihmana* property and may help to revive the damaged anterior horn cells and thus provide improvement in motor functions. Also the *Laghu guna* of *Tikta rasa dravyas*[7] allow penetration into deeper *Dhatus*. The internal medicines administered during the treatment course include *Bhadradarvadi kashaya*, *Dhanvantaram* 101 *Avarti*, *Gandha taila*, *Balarishtam*. These medicines also help in relieving the vitiated *Vata dosha* and provides *Dhatu brihmana*.

CONCLUSION

Hirayama disease is a self limiting motor neuron disease which lacks a definite conservative management. Ayurveda has the potential reserve of treatment methods that can be utilized for the effective management of Hirayama disease. This patient was treated on the basis of *Vatavyadhi* management focusing on *Bahusosha* with both internal and external treatment. After the two schedules of treatment courses with a gap of 1 week, the patient got significant symptomatic improvement in symptoms and motor functions. It is evident that Ayurveda can be effective for the treatment of chronic neurological diseases like *Hirayama* disease.

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REFERENCES

1. Hirayama K. Non-progressive muscular atrophy of the distal upper extremity (Hirayama disease). Intern Med. 2000 Apr; 39(4): 223-9.
2. Huang YL, Chen CJ. Hirayama disease. Neuroimaging Clin N Am. 2011 Nov; 21(4): 939-

50, ix-x. doi: 10.1016/j.nic.2011.07.009. PMID: 22032508.

3. Sharma R, Dash VB, editors. Agnivesa's Caraka Samhita: Text with English translation and critical exposition based on Cakrapani Dutta's Ayurveda dipika. Reprint ed. Varanasi: Chaukambha Sanskrit Series; 2009. Chikitsa Sthana chapter 15/117-118, P.46.
4. Vaidya Yadavji Trikamji Ācharya, Narayan Ram Ācharya 'Kavyatirtha', editors. Suśruta samhitha. Varanasi: Chaukhamba Sanskrit Sansthan; 2017. Chikitsa sthana chapter 24/51, P.497.
5. Hari Sadasiva Sastri Paradakara (editor), Astangahrdaya. Varanasi: Choukhamba publications; 2009, Sootra sthāna chapter 15/10, P.234.
6. Sharma R, Dash VB, editors. Agnivesa's Caraka Samhita: Text with English translation and critical exposition based on Cakrapani Dutta's Ayurveda dipika. Reprint ed. Varanasi: Chaukambha Sanskrit Series; 2009. Siddhi sthana chapter 8/4, P.310.
7. Hari Sadasiva Sastri Paradakara (editor), Astanga hrdaya. Varanasi: Choukhamba publications; 2009, Sootra sthāna chapter 10/16, P. 156

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