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Avascular Necrosis (associated with Sickle Cell Anaemia and Thalassemia minor) Management through Ayurveda - A Case Report

Seema Gupta[1]*, Arun Kumar Mahapatra[2], Rajagopala S[3], Prasanth Dharmarajan[4]

MD (Ayu) Scholar[*1], Assistant Professor [2], Associate Professor[3] Dept. of Kaumarabhritya, Assistant Professor[4] Dept. of Panchkarma, All India Institute of Ayurveda (AIIA), Sarita Vihar, New Delhi, India

Abstract: Avascular Necrosis (AVN) of femoral head is one of the frequent and most debilitating complications in children suffering from Sickle Cell Anemia and ß-Thalessemia minor. AVN in Sickle Cell Anemia is prevalent in approximately 3% of children under age group 15, 8.7%-12.4% by age 21 and progressing to values higher than 50% in population over 35age group. AVN is associated with pain and reduction of Range of Motion. *Samprapti* (Pathogenesis) of this *anuktavyadhi (unmentioned disease)* in Ayurveda texts can be understood, based on Ayurvedic Principles as *Khavaigunya*→Avarana→*Raktakshaya*→*Vataprakopa*→*Asthimajjakshaya*, finally leading to the clinical condition similar to AVN. Limitation of AVN treatment is surgery, offers hip joint replacement of only 15-20 years life which is not so commonly available and expensive too. Ayurveda offers a suitable answer to AVN through *Panchakarma* procedures and *Shamana Chikitsa*, check the progression, reduce and reverse the disease process and improves quality of life of the patient. A male patient aged 16 years suffering from bilateral AVN with Sickle Cell Disease (SCD) and ß-Thalassemia minor was treated with *Deepana-Pachana*, *Virechana*, and *Kala Basti* followed by *Shamana* and *Rasayana Chikitsa*. The assessment was done on Harris Hip Scale (HHS),Visual Analogue Scale (VAS) and Depression Anxiety Stress Scale (DASS), patient had symptomatic improvements and specific changes on above scales. Ayurvedic Management provided marked relief from pain, tenderness, stiffness and improvement in gait.

Key words: Ayurveda Avascular Necrosis, ß-Thalassemia minor, *Panchakarma*, *Rasayana*, Sickle Cell Disease

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***CORRESPONDING AUTHOR:**
Dr. Seema Gupta
MD (Ayu) Scholar, Department of *Kaumarabhritya*, All India Institute of Ayurveda (AIIA), New Delhi, India
Email: seema.gupta177@gmail.com

Introduction:

Avascular Necrosis (AVN) is a progressive necrosis of bone, result transient/permanent loss of blood supply to the bone due to obturator arterial occlusion, thought to be due to the occlusion of Sickled Cells[1], when AVN is associated with Sickle Cell Disease. It affects predominantly weight bearing joints (particularly 60% hip joint) [2]. AVN in Sickle Cell Anemia is prevalent in approximately 3% of children under age group 15, 8.7%-12.4% by age 21 and progressing to values higher than 50% in population over 35[3]. The relative frequencies of the most common cause are alcohol intake (20-40%), corticosteroid therapy (35-40%) and idiopathic causes (20-40%). In a recent study, osteonecrosis (avascular necrosis) has been identified as one of the frequent and more debilitating complication of Sickle Cell Anemia. In Central India, Sickle Cell Disease has a very high prevalence and is the most common condition associated with osteonecrosis followed by alcohol abuse and corticosteroid use[4]. Patients are initially asymptomatic, femoral head Avascular Necrosis progress to destruction of the hip joint in a relatively short time, requiring arthroplasty[5].AVN (avascular necrosis) of femoral head is clinically characterized by gradual onset of pain and limitation of motion. Pain may be localized to groin area but may radiate down the affected limb or ipsilateral buttock, knee or greater trochanter region. Pain is exacerbated with motion or weight bearing and relieved by rest. Passive range of motion of hip is painful, especially forced internal rotation. A distinct abductor lurch and rotation with limitation of abduction and adduction is seen.

Many scholars of Ayurveda have correlated the clinical features of AVN with *AsthimajjagataVata*[6] described in Ayurvedic Classics. In Ayurveda, vitiated *Vatadosha* in *Asthi* (bones) and *Majja* (marrow) leads to *AsthimajjagataVata*[7] which presents clinically with features as *Bhedo Asthiparvanam* (breaking type of pain in bones), *Sandhishoola* (joint pain), *Satataruka* (continuous in nature), *Mamsabalakshaya* (loss of strength and muscle weakness) and *Aswapna*[8](disturbed sleep), which can be correlated with the symptoms of AVN. As there is loss of blood supply and necrosis of bone and bone marrow, features of AVN like destruction of hip joint (*Asthishunyata*) and hip joint pain (*AsthiShoola*) closely resemble symptoms of A*sthimajjakshaya* due to vitiated *Vata Dosha. Samprapti* (Pathology) can be understood as *Asthimajjagatavata* leading to *Asthmajjakshaya* in this disease.

Case History: A 16 years male patient visited OPD of Kaumarabhritya, All India Institute of Ayurveda, New Delhi with chief complaints of pain in both hip joints, more in left hip joint, with restricted movements, difficulty in standing from lying position, stiffness in morning for 4-5 hrs, increase in weight, disturbed sleep due to pain and stressed since april 2018.

The patient claimed to be apparently healthy 2 years back. He had complaint of left knee swelling with pain since October 2017. Earlier patient was diagnosed for Reactive arthritis associated with Sickle Cell Anaemia and ß Thalassemia minor. Transfusion of one unit whole blood and

medicines like diclofenac sodium reduced pain and swelling of knee joint. After few months patient had pain in right knee joint along with swelling. Patient took treatment but not completely relieved. After six months patient had complaint of left hip joint pain. Pain was gradually increasing radiating to left knee joint. Patient started taking some Ayurvedic medicines. Mild relief in pain was obtained, but restricted movements with increased morning stiffness and difficulty in daily activities persisted. Patient was admitted in the IPD of AIIA under Department of *Kaumarabhritya* for the treatment.

Examinations:

Vitals: Temperature-98.9°F(armpit), P.R.-84 min, R.R.-20/min, B.P.-108/70mmhg.

Systemic Examination: Limping gait, Tenderness and reduced range of motion at left hip joint.

Investigations: In MRI bilateral Avascular Necrosis (AVN), Right hip-2nd Stage AVN and Left hip-3rdStage AVN was found. Laboratory findings were Hb-9.1 g/dl, WBC-9100/mm^3, Platelet Count-45000, Sickle Prep – positive, HbF-2.8.

Ayurvedic Pariksha revealed[9]–1.*Prakriti-Pittavata* 2.*Vikriti- Dosha:Vata* 3.*Dushya-Rakta, Asthi, Majja 4. Sara-Medoasthisara 5.Samhanana- Madhyam 6.Pramana- Madhyama 7.Satmya- Ekarasa Satmya 8.Satva- Avara 9. Ahara- Jarana Shakti-Avara, Abhyavaharana shakti-madhyam 10. Vyayama- Avara.*Patient was analysed as per *Ayurvedic* norms, based on which he was diagnosed as having *Asthimajjagatavata* resulting *Asthimajjakshaya*.

Assessment Criteria: Mentioned in table1.

Treatment given:

Deepana-Pachana [table 2]was given for 3 days. *Abhyantara Snehapana*[table-3] with *Go-Ghrita* was given to the patient on empty stomach and continued till the appearance of *Samyaksnehana lakshana* like *Vitsneha, Twakmardava, Mriduta Anganaam.* It took 7 days to observe these features. Dose was 30, 50, 100, 120,150, 180, 210 ml on 1st, 2nd, 3rd, 4th, 5th, 6th and 7th day respectively. This was followed by *Sarvanaga Abhyanga with Dhanvantara Taila* and *Sarvanaga Swedana* (Sudation in a steam chamber) with *Dashmoola Kwatha* for 3 days. Next morning empty stomach *Trivrit Avaleha*150mg with *Munnaka100g* was orally administered for *Virechana* and *Madhyam Shuddhi* was obtained. After *Virechana Samsarjana Karma* [table4] for 5 days two days gap was advised followed by *Kala Basti*[table -5]Patient was advised to consume warm water only and easily digestible light food items. Exposure to cold air, maintaining one particular posture for longer duration, frequent jerky movements and lifting weights were asked to be avoided.

After *Kala Basti, Pizichil* was done with *Karpaasthyadi Taila for 5 days* followed by *Shashtikashali Pindasweda* for 5 days. Along with *Kala Basti, Pizichil* and *Shashtikashali Pindasweda, Samshamana Chikitsa* was also given[table 6], After *Panchakarma* procedures patient was kept for 2 months on *Rasayana Chikitsa* with *Brihatvata Chintamani Rasa*125mg tds with honey and *Samshana Chikitsa* for 2 months [table- 7].

The assessment of result was done as shown in table 9.

Table-1: Assessment Criteria:

1. H.H.S. – Harris Hip Score[10]

<70Poor	70-79 Fair	80-89 Good	90 -100Excellent

2. V.A.S. – Visual Analogue Scale[11]

0	2	4	6	8	10
No hurts	Hurts little bit	Hurts little more	Hurts even more	Hurts whole more	Severe

3. Weight Assessment
4. 4. DASS-Depression Anxiety StressScale[12]

Table-2 : Shodhana and ShamanaChikitsa planned for managing AVN

<table>
<tr><th>S.N.</th><th>Procedure</th><th>Days</th><th>Drugs</th></tr>
<tr><td>1</td><td>Deepana-Pachana</td><td>3</td><td>Chitraktadi Vati[13] 2 tab tds, Gandharvahastadi Kwath[14] 20 ml bd, Triphala Churna 1tsf</td></tr>
<tr><td>2</td><td>Snehapana</td><td>7</td><td>Goghrita</td></tr>
<tr><td>3</td><td>Virechana</td><td>-</td><td>Trivrit Avaleha[15]and Munnaka</td></tr>
<tr><td>4</td><td>Kala Basti</td><td>16</td><td>
Anuvasana Basti(A)
<table>
<tr><th>S.N.</th><th>Drug</th><th>Qt</th></tr>
<tr><td>1</td><td>Sahacharadi Taila[16]</td><td>40 ml</td></tr>
<tr><td>2</td><td>Tiktaka Ghrita[17]</td><td>60 ml</td></tr>
<tr><td>3</td><td>Shatpushpa Churna</td><td>15 gm</td></tr>
<tr><td>4</td><td>Saindhava (Rock Salt)</td><td>10 gm</td></tr>
</table>
Niruha Basti(N)
<table>
<tr><th>S. N.</th><th>Drug</th><th>Qt.</th></tr>
<tr><td>1</td><td>Saindhava(Rock Salt)</td><td>5g</td></tr>
<tr><td>2</td><td>Shatpushpa</td><td>15g</td></tr>
<tr><td>3</td><td>Gomutra</td><td>50 ml</td></tr>
<tr><td>4</td><td>Imli Rasa</td><td>50 ml</td></tr>
<tr><td>5</td><td>Manjishthadi Kwatha[18]</td><td>150 ml</td></tr>
</table>
</td></tr>
<tr><td>5</td><td>Pizichil</td><td>5</td><td>KarpasasthyadiTaila[19]</td></tr>
<tr><td>6</td><td>Shashtikashali PindaSweda</td><td>5</td><td>Shashtikashali Rice and Bala Moola Kwatha with milk</td></tr>
</table>

Table-3 Snehapana:

Day	Dose	Symptoms/Observations	Complications
1	30ml	*Deeptaagni*	None
2	50	*Deeptaagni*	None
3	100	*Deeptaagni* ,Slight *Snigdha Mala*	None
4	120	*Vatanulomana*, Slight *Snigdha Mala*, *Asamhat Mala* (Slight loose stool)	None
5	150	*Vatanulomana*, Slight *Snigdha Mala*, *Asamhat Mala*	None
6	180	Above *Lakshanas* (Symptoms) +*MriduTwak* (Soft Skin)	None
7	210	Above + *Snigdha*Twak	None

Table-4: Samsarjana Karma

Day	*PratahAnnakala (Morning diet)*	*SandhyaAnnakala (evening diet)*
1	-	*Peya*
2	*Peya*	*Vilepi*
3	*Vilepi*	*Akrita Yusha*
4	*KritaYusha*	*Akrita Mamsa Rasa*
5	*KritaMamsa Rasa*	*Light Diet*

Table-5: Sequence of Basti:

Day of Basti	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16
Anuvasana(A)/Niruha(N)	A	A	N	A	N	A	N	A	N	A	N	A	N	A	A	A

Table-6: Shaman Chikitsa(during panchakarma procedures):

Drug	Dose
AbhaGuggulu[20]	2 tab bds
TriphalaGuggulu	2 tab bds
GugguluTiktaka Kashaya	10ml bds
GandhaTaila[21] Capsule	1 cap OD

Table-7: Shaman Chiktsa(during follow up):

Drug	Dose
Brihatvata Chintamani Rasa[22]	125mg tds with honey
Kaishore Guggulu[23]	1tab bds
Lakshadi Guggulu[24]	1tab bds
Guggulu Tiktaka Ghrit[25]	1 tsf bds empty stomach
Gandha Taila	1 cap OD
Manjishthadi Kwath	30 ml bds
Lakshadi Tail[26]	For local application

Table-8: Assessment of results

Scales		B.T.	After *Snehpana*	After *Virechan*	After *Basti*	After *pizichil*	After s.s.p.s.	After follow up of 2 months
Harris Hip Score(H.H.S.)		34.55	44.7	44.55	45.55	63	68.05	71.35
VAS Score		6	4	6	4	4	4	2
Weight(in kg)		87	80	79.6	79	78	78	75
DASS Score	Depression	17	14	15	14	12	9	7
	Anxiety	6	6	7	6	5	5	3
	Stress	18	12	15	13	12	10	9

Results and Discussion:

Patient who was initially unable to walk without support and unable to bear whole body weight on left leg was able to walk without support with mild limping and was also able to bear his weight on left leg for 10seconds. Significant reduction in pain, stiffness, weight, stress and improvement in gait and range of motion were obtained after the follow up [Table 8].

Avascular necrosis is a degenerative condition of the bone which is progressive in nature, due to lack of blood supply[27]to the particular part of the bone due to injury or any occlusion[28] in the blood vessels nourishing the bone tissue.

Prakriti of the patient was *Pittavataja* and weight on admission was 87 kg. *Aama* symptoms such as *Aruchi, Gaurava, Anilamudhata,Stabdha Gatra* and *Malasanga* were observed in the patient. So A*mapachana* was first targeted by adopting *Deepana Pachana.*

DeepanaPachana:

ChitrakadiVati and G*andharvahastadi Kashaya* were administered for *Amapachana* and *Vata-Anulomana* as they contain ingredients predominate in *Katu-Rasa and Ushna-Veerya*. These Gunas (qualities) increase *Jatharagni* and help to achieve *Vatanulomana*.

AvaranaChikitsa:

TriphalaChurna has *Lekhana* effect helps in reducing *Meda* (as patient was overweight) and relieving *Malasanga* following the principal "*Brimhamastumridulanghayet*", can help in resolving *Avarana* of *Kapha* and *Meda*..

Srotodushti Chikitsa:

Raktavaha Srotorodh[29] became the prime cause leading to *Asthidhatu Kshaya* in this patient, as in Sickle Cell Disease the cause of ischemia is supposed to be the sequestration of sickled cells (*Khavaigunya)* in the branch of obturator artery supplying femoral head leading Avascular Necrosis of hip joint. To counter this *SrotoDushti, Virechana* was planned for *Shodhana* to produce detoxifying effects, to provide *Dhatu Sthairya* (stability) and more chances for better absorption of *Basti Dravya. Goghrita* was selected for S*nehapana* which can be easily taken by *Avara Satva* patient, favours normal functioning of *Dhatvagni*, thereby facilitating increased nutrition of A*sthimajja Dhatu*. *Ghrita* is *Vatapitta Shamaka, Balya, Agnivardhaka, Madhura,*

Sheetaveerya and helps to improve the *Dhatu Upachaya*

As the patient was *Kroora Koshthi* and *AvaraSatva, Trivritavaleha* with *Munnaka* was given for *Virechana*. Increase in pain was reported which could be due to discontinuation of analgesics and anti inflammatory drugs because *Virechana* flushes all the toxins and removes the suppressing effect of previously given drugs.

Vata Dosha Chikitsa:

According to the *Ayurveda* there is no direct correlation with avascular necrosis but clinical presentation shows there is dominance of *Vata Dosha* and *Kshaya* of *Rakta, Asthi, Majja Dhatu*. This patient had S*roto Sanga* (occlusion of blood vessels) along with *Medo Dhatu Samavriddhi* symptoms signify *Avarana* which leads to reduction in the blood supply to the femoral head ultimately necrosis. *Margavarodha* further aggravates *Vata Dosha* finally resulting into progressive loss of A*sthimajja Dhatu*. Main symptoms present were pain, stiffness, decreased Range of motion which is indicating *Vata Dosha* involvement. Therefore *Kala Basti* was planned.

Rakta Kshaya Chikitsa:

After *Virechana*, *Kala Basti* for 16 days followed by *Pizichil* + *Basti* for 5 days was planned. *Manjishthadi Kshara Basti as Niruha Basti* was given because patient had *Rakta Kshaya* and *Sirasanga*. *Tiktaka Ghrita* is *Tikta Rasa Pradhana* and is effective in *Raktadushti Chikitsa* and *Sahacharadi Taila* is useful in *Vata Dosha Shamana*. Pain and stiffness were mildly reduced. As patient was suffering from Sickle Cell Anemia and ß Thalessemia minor *Rakta Shodhana* for a long term was required along with *Vata Shamana* to initiate the process of reversing the *Shadvidha Kriyakala*, from *Vyakti→Sthanasamshraya→Prasara→Prakopa→ to Sanchaya*, and reduction of symptoms. *Pizichil* was given with *Karpaasthyadi Taila* having *Vata Shamaka* properties.

Asthimajjakshaya Chikitsa:

With the administration of *Kala Basti, Shaman Chikitsa* was also started included *Abha Guggulu, Gandha Tailam* Capsules for *Asthi Dhatu* nutrition, *Triphala Guggulu* to stop recreation of *Avarana* and *Guggulu Tiktaka Kashaya* for *Vata Dosha Shamana* and improving *Asthimajjakshaya*. In the end of the procedures *Shashtikashali Pinda Sweda* was done, *Balya* and effective for *Asthimajja Dhatu*. Pain and stiffness reduced significantly, range of motion improved, tenderness was no more, gait improved, weight reduced, *Jatharagni* (appetite) was good, *Raktavaha Srotoshudhhi* was obtained. Throughout the whole procedure depression, stress and anxiety scores were consistently reducing.

***Shamana* with *Rasayana Chikitsa* for improvement of Quality of life:**

Follow up period of 2 months was planned. *Brihatvatachintamani Rasa (Rasayana), KaishoraGuggulu (Shothahara), LakshadiGuggulu* (*AsthiPoshak* and *Shothahar*), *Gandhataila Casules* (*AsthiPoshak* and *Shothahar*) and *ManjishthadiKwatha* (*Raktashodhaka*and *Prasadaka)* along with *Lakshadi Taila* (*Vata Shamaka)* were advised to the patient with *Pathya* and *Apathya* for preserving the effect of *Panchakarma* and

to prevent *Samprapti* and progression of the disease.

Conclusion:

The adopted therapy in the current case provided relief symptomatically from pain, tenderness, stiffness, range of motion, stress, general debility and improvement in the gait. Further clinical trials and extended therapy in the present case are needed to establish a standard management of Avascular Necrosis associated with Sickle Cell Anemia and ß thalessemia minor

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