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Case Study

A CASE STUDY ON NETHERTON'S SYNDROME

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ABSTRACT

Present case was diagnosed as "Netherton's Syndrome" in Sri Ramachandra Hospital (Sri Ramachandra University Porur Chennai-600116) and got treated in December 2010. Netherton's Syndrome is a severe, Autosomal recessive form of Ichthyosis associated with mutation in the SPINK 5 gene. It is characterized by chronic skin inflammation, universal pruritus, severe dehydration and stunted growth.

After observing signs and symptoms related to GI Tract such as Increased frequency of stool, Change in consistency of stool, Lower abdominal pain, reduced appetite, Vomiting and general presentation such as Dryness of skin, itching all over the body, stunted growth case was diagnosed and treated as Vataja Grahani (Vis-à-vis Mal absorption Syndrome). Patient had been administered with deepana, pachana, sangrahi (atisaraghna) drugs. After observing the signs and symptoms of proper functioning of agni and vata in GI Tract level, a course of takrabasti was administered for 16 days. Then after takrapana along with deepana, pachana, and sangrahiashadhis are continued. The results were assessed by grading of signs and symptoms at particular intervals and laboratory investigations before and after study. The results reveal the significant response over various subjective and objective parameters.

Keywords: Netherton's Syndrome, Vataja Grahani, Mal absorption Syndrome, Takrabasti.

INTRODUCTION

Grahaniroga described in Ayurvedic classics represents derangements of digestive System, caused by the impairment of agni. Its significance is emphasized by its inclusion among astamahagadas¹.

Vataja Grahani is a type of Grahani, which exhibits signs and symptoms like Tanuamamalapravritti (loose stools with mucus), Punapunamalaprivritti (Increased frequency of stool), Visuchika (Vomiting and diarrhoea), Vairasya (Tastelessness), Dukhapachana (difficulty in digestion of food), admana (bloating of abdomen), karshya (weight loss), dourbalya (weakness), Parushya (dryness or roughness of body parts like skin and hair).

However pathophysiology and symptoms of Vataja Grahani resembles with Mal absorption syndrome explained in contemporary science. Mal absorption constitutes the pathological interference with normal physiological sequence of digestion, absorption of nutrients.

Diarrhoea often steatorrhoea is the most common feature watery, diurnal and nocturnal bulky fragment stools are the clinical hallmarks of Malabsorption. Abdominal discomfort,

bloating of abdomen, weight loss, growth retardation, delayed puberty in children, failure to thrive are other general symptoms².

In present study though the case was diagnosed as Netherton's syndrome, patient had both GIT symptoms and general symptoms related to Vataja Grahani. Hence the patient was administered with deepana, pachana, sangrahi drugs and a course of takrabasti followed by takrapana.

CASE HISTORY

A male patient, From Tiruvanamalaidsttq, Tamil Nadu aged about 22yrs got admitted to Government Ayurveda Hospital, Mysore, In male 1st ward 15 bed number, With IP no 189 on 4/1/2013, with following complaints

- Increased frequency of stools 4-5times/day since 14 years.
- Change in consistency of stools (liquid) since 14 years.
- Lower abdominal pain immediately after intake of food which would reduce after passing stool since 14 years.
- Reduced appetite since 14 years.
- Vomiting occasionally after intake of food since 14 years.
- Dryness of skin since 14 years.
- Itching all over the body and scalp since 14 years.

FINDINGS ON GENERAL EXAMINATION

Built: hyposthenic
 Weight: 30 kg
 Height: 4.6 feet
 Nails: brittle

FINDINGS ON SYSTEMIC EXAMINATION

Abdomen;

Inspection: Shape: Scaphoid
 No visible scar, mass, dilated veins
 Palpation: Soft, No palpable mass detected
 Tenderness present at epigastric and hypogastric region
 Auscultation: bowel sound- heard 2-3/min
 Percussion: NAD

LABORATORY FINDINGS

Hb% 13gms/dl
 TC- 5, 7000 cells/cu mm
ESR – 36mm/hr
 DC- P-61%,E-4%,M-1%,L-34%
 Urine-Albumin, Sugar-Nill, Micro-NAD
 Stool-ova, cyst-Absent, Micro-NAD, **Occult blood-Present**

SAMPRAPTI GHATAKAS

Dosha: Samanavata, Apanavata, Pachakapitaa

Dushya: annarasa, rasadhatu
 Agni: Jataragni
 Ama: Jataragnimandyajanyaama
 Srotas: Annavaha
 Srotodusti: Atipravritti
 Adhistana: Grahani
 Rogamarga: Abhyantara

MANAGEMENT

For first 7days patient was advised with kutaja Ghana vati³ 1tid After food with water and Jeerakadi churna⁴ 3grams tid after food with warm water , as deepana, pachana and grahi. In 3-4 days frequency of stool reduced from 4-5/day to 1-2times/day, pain Abdomen also got reduced markedly. From 8th day Yashadabhasma⁵ 125mg bid before food (As zinc supply) and Tuvaraka taila⁶ for external application (for itching) was added with previous medicines (Kutaja Ghana vatai and Jeerakadichurna).Same medicines were continued till 14th day. From 15th day all the above medicines were stopped. A course of takrabasti was advised for 16 days (in kalabasti pattern).

1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16
A	N	A	N	A	N	A	N	A	N	A	N	A	A	A	A

AnuvasnawithChangeryadi ghrita⁷-70ml
 Niruha with- Saindhavalavana 5grams
 Makshika 35ml
 Changeryadi ghrita-70 ml
 Putoyvanyadi kalka⁸ 20grams
 Takra 300 ml
 From 31st day after completion of basti, since patient had 2 episodes of vomiting and complaining about epigastric pain, he was advised with Pravalapanchamruta tablets 1tid before

food with ghrita (1tea spoon) Along with kapittastaka churna⁹ 6 grams bid before food with butter milk, kutaja Ghana vati 1tid After food with water. From 40th day he was advised to take 200ml of butter milk after food tid along with Pravalapanchamruta 1tid Before food with ghrita, Kapittastakachurna 6 grams bid before food with butter milk, Kutaja Ghana vati 1tid After food with water and 777oil for external application for dryness of skin.

Table 1: Showing the given treatment, duration and observation of results in the present case study

Treatment	Result	
	Before Treatment	After Treatment
Deepana, pachana, grahi with Jeerakadichurna 3gms tid A/F with warm water, kutaja Ghana vati 1tid A/F with water for first 7days	<ul style="list-style-type: none"> Frequency of stool 4-5times /day Abdomen pain after intake of food. 	<ul style="list-style-type: none"> Frequency of stool 1-2 times/day Reduction in severity of Abdomen pain after intake of food.
Yashadabhasma 125 mg bid B/F(zinc supply) and Tuvarakataila for external application along with above medicine from 8 th -15 th day	<ul style="list-style-type: none"> Dryness of skin and hair Itching all over the body scalp 	<ul style="list-style-type: none"> Mild relief from dryness of skin and itching
Takrabasti for 16 days in karma bastipattern. After basti course kapittastakachurna 6gms bd B/F with takra with above mentioned treatment.	<ul style="list-style-type: none"> Frequency of stool 1-2times/day Mild abdomen pain after intake of food 	<ul style="list-style-type: none"> Frequency of stool 1/day Complete relief from abdomen pain.
Sutashekara rasa 1tid A/F with water and tab Pravalapanchamruta 1 tid B/F with grita for 7days	<ul style="list-style-type: none"> Vomiting after intake of food Epigastric pain. 	<ul style="list-style-type: none"> Frequency of stool 1/day Complete relief from abdomen pain
Butter milk 200ml trice a day after food. Along with kutaja Ghana vati 1tid A/F with water, kapittastakachurna 6gms bd B/F with takra, pravalapanchamruta 1tid B/F with grita	<ul style="list-style-type: none"> Frequency of stool 1-2times/day Mild abdomen pain after intake of food Vomiting after intake of food Epigastric pain. Dryness of skin and hair 	<ul style="list-style-type: none"> Frequency of stool 1/day Complete relief from abdomen pain Dryness of skin –reduced No vomiting Complete relief from epigastric pain.

While discharging from the hospital on 50th day patient got complete relief from Pain abdomen, frequency of stool got reduced to once per day with normal consistency of stool (semisolid without associated with mucus), there was marked

improvement in his appetite. There was gain of 2kg body weight in patient (from 30 kg to 32 kg in 50 days). Other complaints like Dryness of skin and vomiting was reduced.

Table 2: Showing the method of assessment of treatment

Sl. No	Symptoms	Scoring
A	Constipation	
1	Normal once daily	B0
2	Alternative days	B1
3	Once in two days	B2
4	Once in three days	B3
5	Once in four days	B4
B	Changed consistency of stool	
1	Normal once daily	D0
2	Twice daily	D1
3	2-4 times daily	D2
4	4-6 times daily	D3
5	>6 times daily	D4
C	Frequency of stool	
1	Normal once daily	M0
2	Twice daily	M1
3	2-4 times daily	M2
4	4-6 times daily	M3
5	>6 times daily	M4
D	Abdomen pain/discomfort	
1	No abdominal pain	P0
2	Occasional/rarely abdominal pain	P1
3	Intermittent lower abdominal pain,relived by passage of flatus and stool	P2
4	Continuous pain not relived by passage of Flatus & stool	P3
E	Mucous in stool	
1	No visible mucous in stool	A0
2	Visible mucous stikled to the stool	A1
3	Passage of mucous with frequent stool	A2
4	Passage of large amount of mucous in stool	A3
F	Gas or Flatulence	
1	No abnormal gas / flatulence	G0
2	Occassional abdominal distention	G1
3	Frequent abdominal distention with increased flatulence & belching	G2
4	Rumbling/Gargling soundpresent	G3
G	Dryness of skin	
1	No dryness	R0
2	Milddryness	R1
3	Moderate dryness	R2
4	Sever dryness	R3
H	Vomiting	
1	No vomiting	V0
2	Occasional vomiting	V1
3	Vomiting daily once	V2
4	Vomiting 2-3/day	V3

Table 3: Showing the assessment chart

Lakshana	1st day	8th day	16th day	30th
Frequency of stool	M3	M1	M0	M0
Consistency of stool	D3	D1	D0	D0
Constipated stool	B0	B0	B0	B0
Abdominal pain	P2	P0	P0	P0
Mucous in the stool	A2	A1	A0	A0
Gas/ flatulence	G1	G0	G0	G0
Dryness of skin	R3	R2	R1	R1
Vomiting	V1	V1	V0	V0

Table 4: Showing the laboratory findings before and after treatment

FINDINGS	BT	AT
OCCULT BLOOD IN STOOL	Positive	Negative
E S R	36mm/hr	22mm/hr
WEIGHT	30kg	32kg
Hb%	13gm%	17gm%

Table 5: Showing the assessment during follow up period

Lakshana	15/4/13	20/5/13	17/6/13	15/7/13	5/8/13
Frequency of stool	M1	M0	M0	M0	M0
Consistency of stool	D0	D0	D0	D0	D0
Constipated stool	B0	B0	B0	B0	B0
Abdominal pain	P0	P0	P0	P0	P0
Mucous in the stool	A0	A0	A0	A0	A0
Gas/ flatulence	G0	G0	G0	G0	G0
Dryness of skin	R1	R0	R0	R0	R0
Vomiting	V0	V0	V0	V0	V0

DISCUSSION

Grahaniroga explained in Ayurvedic science is due to altered function of organ "Grahani and there by leading to disturbed pachanakriya at different level i.e at GIT level and at dhatu level¹⁰(digestion and absorption at different level).

Since general line of treatment mentioned for "Grahani" rogain Ayurvedic classics is deepana, pachana and grahi depending on the stage of the disease¹¹. Jeerakadi churna and kutaja Ghana vatai was advised for first seven days .Both acted as deepaka, pachaka and grahi.

It has been mentioned in classics that Kutaja possess kashaya, tikta rasa, laghuruksaguna, sheetaveerya and katu vipaka¹². Because of its Kashaya rasa and sheetaveerya it acted as stambhaka and grahi, which is especially use full in grahani. By its katuvipaka it does deepana and pachana. Kutaja is Kaphapitta hara grahi hence indicated in Grahani and Atisara. The gut stimulant and relaxant activity of kutaja are mediated possibly through activation of histamine receptors and calcium channel blockade respectively and this study provides sound mechanistic background for its usefulness in gut motility disorders such as constipation, colic and diarrhea. kutaja is proved as anti-inflammatory¹³. Jeerakadichurna mentioned in Bhaishajyaratnavali "Grahanirogadhikara" contains ingredients Jeeraka, Tankana, Musta, Pata, shatapushpa. etc)¹⁴ with katu, tikta, kashayarasa, deepana and grahiguna, hence acted as deepaka, pachaka and grahi¹⁵. Jeeraka the main ingredient of jeerakadichurna is best Carminative and it

reduces superficial inflammation and pain. Apart from kindling appetite, digests food and normalizes digestive system, Reduces pain experienced during indigestion¹⁶.

Since basti has been considered as best for vataja vikara¹⁷ and takra is also tridosahara and good remedy in grahanivikara. takrabasti was administered in karma basti pattern.

Takrapana in grahani serves the purpose of pathya and oushadha. By its Amla rasa and sandraguna it pacifies vata, Because of its swaduvipaka its considered as pittahara, Kapahahara by its Kashyarasa, ushna, roukshya, vikasi qualities. Best in Grahaniroga because of its sangrahi (anti-diarrhoeal) and Laghu(light for digestion) guna¹⁸.

Probiotics in takra introduce healthy bacteria into the body, particularly into the digestive tract, where bacteria are essential for the breakdown and absorption of foods and their nutrients. A regular source of probiotics like buttermilk is important, as it will help to maintain normal gut flora to support optimal digestion and overall health.¹⁹. Hence takrapana was advised after a course of basti.

Other symptomatic treatment such as using sutasheskara rasa and pravalapanchamruta for managing udarashula and chardi is considered as per the seat of grahaniroga is grahaniavayava and sthanikadosha is pachakapitta. Both the formulations are indicated in Grahani Roga as per "Yogaratanakara". Both the drugs serve the purpose of symptomatic treatment and thereby giving symptomatic relief in chardi and udarashula, also acted on grahaniroga.

The Ingredient of Pravalapanchamruta–Pravalabhasma, Muktabhasma, shankabhasma, arkaksheera, muktashukti-bhasm, all these possess kshareeya, swadu, sheetala, laghu, deepana, pachana, qualities²⁰. Sutashekara rasa is mentioned in Yogaratnakara Amlapittadhikara. Its indicated in GIT disorders such as Amlapitta, Chardi, grahani, tridoshaja atisara²¹.

FOLLOWUP AFTER TREATMENT

After discharging from the hospital patient is advised to continue with Kutaja Ghana vati 1tid A/F with water, jeerakadichurna 3gms tid A/F with warm water, Pravalapanchamruta 1 tid B/F with grita, 777 oil for external application²².

CONCLUSION

According to charakasamhita sutra sthana 18th chapter 44thshloka,Its not possible to name each and every disease. The same dosha which get vitiated due to different causes ,may get localizes at different level and cause different disorders. Hence after analysing prakruti, adhistana and samuttana (Rogajanakavisheshakarana) physician should treat disorders. One who treat the disorder after analysing avasta of roga and rogi, desha, kala, dosha will definitely get success. In present case patient was managed with grahani line of treatment according to the observation of dosha-vatapradhanatridosha, Adhistana-Grahani, andlinga-lakshana of vatajagrahani and patient got marked relief.

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