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A Case Study on Wilson's Disease - An Ayurvedic Perspective

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Abstract

A 32 year old male patient came up with complaints of reduced strength in right upper limb,-tremors in both upper limb,slurred speech,difficulty in swallowing and generalized stiffness since 6 months. His USG investigations showed chronic parenchymal disease in the liver and splenomegaly.And MRI brain impression with altered signal intensity and mild to moderate cerebellar atrophy.This case of generalized stiffness with hepatomegaly and splenomegaly merely correlated with Agnimandya at the Dhatu level that led to Vatavyadhi, Yakrutodara, and Plihodara. In Ayurvedic classics treatment modality mentioned for Vatavyadhi is Snehana (oleation), Mrudu Swedana (mild sudation) and Anuvasana Basti (oil enema).For Yakrutodara and Plihodara is Virechana(Purgation) and Niruha Basti (Decoction Enema). The case was treated with Abhyanga, Swedana,Virechana,Basti and oral medication. After treatment, the symptoms were reduced. His ceruloplasmin serum level at the time of admission was 12.67mg/dl, came to normal 21.76mg/dl. After treatment, the symptoms were reduced.Strength in rt.upper limb improved.Tremors of both upper limb ,difficulty in swallowing and generalized stiffness also reduced.Hence it can be concluded that the Ayurvedic approach and diet modifications in such patients may help in providing supportive care and improving the quality of life.

Wilson's disease can't be correlated directly with any disease as per Ayurveda due to its diverse manifestation. In the present case, the generalized rigidity, hepatomegaly, and splenomegaly may be correlated to Vatavyadhi, Yakrutodara, and Plihodara. Wilson disease is a genetic disorder that prevents the body from getting rid of extra copper.Copper builds up in the liver,brain,eyes & other organs.Serum Ceruloplasmin (Normal value is 14 to 40 mg/dL) -Serum ceruloplasmin level less than 14 mg/dL is considered diagnostic for Wilsons disease.Kayser-Fleisher rings(KF ring) result from a buildup of copper in the eyes and are the unique sign of Wilsons disease.Wilsons disease first attacks the Liver,the central nervous system or both.A buildup of copper in the central nervous system may result in neurologic symptoms including-Problems with speech,swallowing,physical coordination,tremors,muscle stiffness.

Keywords: Ayurveda,Vatavyadhi,Yakrutodara,Plihodara,Wilson's disease

1. Introduction

Wilson's disease (WD) is an inherited disease of fallacious copper metabolism caused due to mutation

in ATP7B, a copper-binding protein, which leads to deviant accumulation of copper in hepatocytes that further spills into circulation and eventually gets deposited in other organs. WD may present as hepatic, neurologic or psychiatric problems.

An inherited disorder that causes too much copper to accumulate in the organs.
Treatment often includes medication that can prompt the organs to release copper into the bloodstream. Once it's in the bloodstream, it can then be eliminated from the body through the kidneys.
Wilson's disease, also known as hepatolenticular degeneration and progressive lenticular degeneration, is a rare genetic disorder that causes copper poisoning in the body. It affects about 1 in 30,000 people worldwide.

In a healthy body, the liver filters out excess copper and releases it through urine. With Wilson's disease, the liver cannot remove the extra copper properly. The extra copper then builds up in organs such as the brain, liver, and eyes.

Early diagnosis is crucial for stopping the progression of Wilson's disease. Treatment may involve taking medication or getting a liver transplant. Delaying or not receiving treatment can cause liver failure, brain damage, or other life-threatening conditions.

Kayser-Fleischer rings and sunflower cataract

Kayser-Fleischer (K-F) rings (Fig-1) and sunflower cataract in the eyes. K-F rings are abnormal golden-brown discolorations in the eyes that are caused by deposits of excess copper. K-F rings show up in about 97 percent of people with Wilson's disease.

Sunflower cataracts show up in 1 out of 5 people with Wilson's disease. This is a distinctive multicolored center with spokes that radiate outward.

Fig 1

Lab tests

For blood tests:-

- Abnormalities in liver enzymes
- Copper levels in the blood
- Lower levels of ceruloplasmin, a protein that carries copper through the blood
- A mutated gene, also called genetic testing
- Low blood sugar

Imaging tests

Magnetic resonance imaging (MRI) and computerized tomography (CT) scans may help show any brain abnormalities, especially if any neurological symptoms. These findings can't diagnose the condition, but they can help determine a diagnosis or how advanced the condition is.

Successful treatment of Wilson's disease depends upon timing more than medication. Treatment often happens in three stages and should last a lifetime. If the patient stops taking the medications, copper can build back up again.

First stage

The first treatment is to remove excess copper from body through Detoxification therapies (Panchakarma).

Second stage

The goal of second stage is to maintain normal levels of copper. Internal medications are given to prevent the condition from worsening or slow its progress.

Third stage

This includes continuation of internal medications along with periodic detoxification therapy. Also manage copper levels by avoiding copper utensils and foods with high levels, such as:

- Dried fruit
- Liver
- Mushrooms
- Nuts
- Shellfish
- Chocolate
- Multivitamins

Case Study

Presenting complaints

- A 32 year old male, diagnosed with Wilson's disease, reported on December 2016 with com plaints of reduced strength in rt.upper limb associated with slurred speech, tremors of both upper limb, difficulty in swallowing and generalized stiffness since 6 months.

Etiology

- Patient's parents were copper smiths and patient had exposure to copper since childhood.

History of present illness:

Initially symptoms started with reduced strength in right upper limb. Gradually developed tremors in both hands. Later slurred speech along with difficulty in swallowing and generalized stiffness.

Investigation

MRI Brain (on 05/11/2015)
Impression: Altered signal intensity. Mild to moderate cerebellar atrophy
USG abdomen: Cirrhosis of liver, Spleenomegaly
Hydrouretronephrosis due to V-U junction calculus.
Blood test (on 22/04/2016):
Ceruloplasmin(serum) -12.67mg/dl
LFT: SGOT: 49 U/L, SGPT: 31U/L

Diagnostic focus and assessment

The patient was diagnosed with Wilson's disease. After a thorough clinical examination, the condition seemed to be Vatavyadhi with Yakrutodara and Plihodara according to Ayurveda.

Therapeutic focus and assessment

Internal medications:

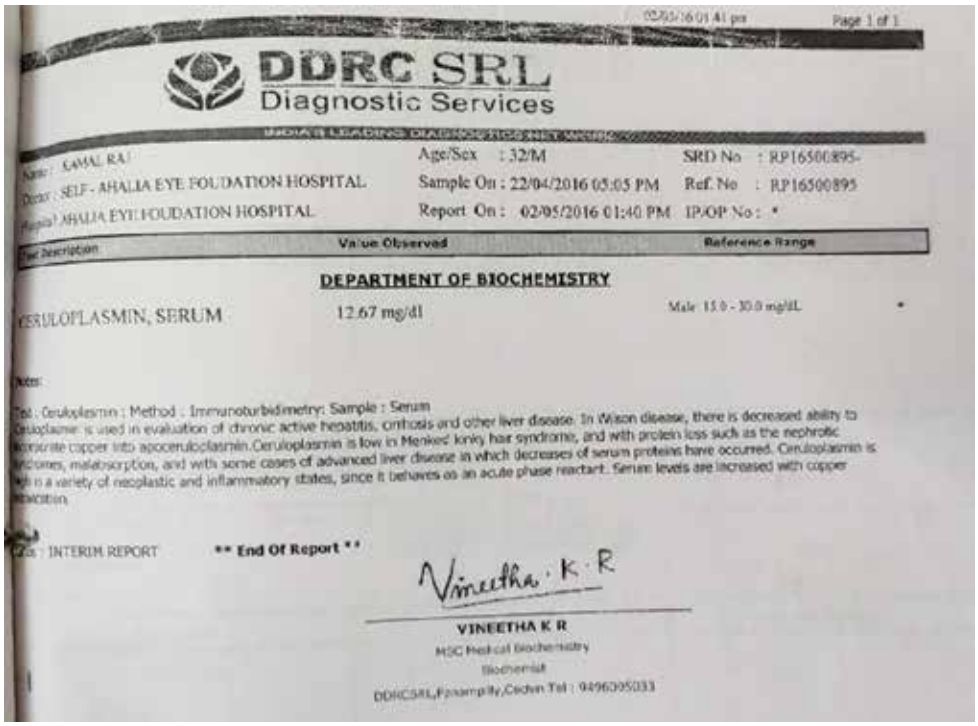
Sl.no.	Medicine	Dose	Duration	Therapeutic effect
1.	Chitrakaadi vati	1 TID	2 Days	Deepanapaachana
2.	Drakshadi kashayam	20ml kashayam+80ml luke warm water BID	For 3 months	Vatapitta shamanam , kamala
3.	Patolakaturohanyaadi ks	20ml kashayam +80ml luke warm water BID	For 1 month	Kamala, Vishagnam
4.	Abhayarishtam +Punarnavaasavam	15 ml each (BID)	For 3 months	Agnideepanam, udaram, pleeha(Yakritodaram, pleehodaram)
5.	Vilvadi gutika	1 TID	3 months	Ajeerna, Vishagnam
6.	Goskuradi guggulu	2 BID	1 month	Ashmari, mootra vikara
7.	Dooshivishari gutika	1 BID	3 months	Dooshivisha, sthavarvisha
8.	Dashamoolaharitaki lehyam	1 tsp HS	3 months	Pleeha,mootra roga
9.	Liv 52 tab(patent drug)	1 BID	3 months	Yakrutprasadakaram, immunomodulatory action

Procedures:

Plan of action	Procedure	Therapeutic effect	Medicine used	Medicine action
(1st sitting) Rookshanam	Udvartanam	srothorodhahara, amapaachanam	kolakulathadi choornam	VataKaphahara
Snehanam(Bahyam)	Abhyangam	Balyam	Dhanvantaram tailam	Vata roga, kshataksheena
Snigdha sweda	Shiro dhara (taila dhara)	Buddhi prasada,Indriya prasada	Ksheerabala tailam	Vata vyadhi
Swedanam	Bashpa swedam	Dosha vilayanam stamba gourava hara	Balamoola kwatha	Vatavyadhi
Virechanam	Snigdha virechana	Dosha shodhanam	Eranda thailam(30ml) + goksheeram(30ml)	Vatavyadhi
(2nd sitting) Rookshanam	Dhanyaaamla dhara	srothorodhahara, amapaachanam	Dhanyaamlam	Vata kapha hara
Swedanam	Pizhichil	Snigdha swedanam	Dhanwantharam tailam	Vata roga, kshataksheena
Virechanam	Hrudya virechana	Dosha shodhanam	Truvrit lehyam(40gm) + drakshakashyam (as anupaanam)	Yakrutprasadakara
Vasthi	Sneha vasti	Vatanulomana,Indriyaprasadam bala vardhanam	Sahacharadi Mezhukupakam	Kampam,Sthamba m
	Nirooh vasti	Vatanulomana, rogopashanti, bala vriddhi	Dashamoola niruha vasthi	Sarva vatavyadhihara
Snigdha Swedanam	Shashtika shaali pinda sweda	Balyam	Shastikashaali, Ksheerabala taila	Vatanulomana

Follow-up and outcomes

• After the first sitting of 15 days tremors of both upper limb, difficulty in swallowing and generalized stiffness were reduced. His ceruloplasmin serum level at the time of admission was 12.67mg/dl(-Fig-2).After the second sitting, underwent investigation of ceruloplasmin serum report noted level was 21.76mg/dl.(Fig-3)



DDRC SRL
Diagnostic Services

Name : KAMAL RAJ | Age/Sex : 32/M | SRD No : RP16500895-
Doctor : SELF - AHALIA EYE FOUDATION HOSPITAL | Sample On : 22/04/2016 05:05 PM | Ref. No : RP16500895
Hospital: AHALIA EYE FOUDATION HOSPITAL | Report On : 02/05/2016 01:40 PM | IP/OP No : *

Test Description	Value Observed	Reference Range
DEPARTMENT OF BIOCHEMISTRY		
CERULOPLASMIN, SERUM	12.67 mg/dl	Male 15.0 - 30.0 mg/dL

Notes:
Test : Ceruloplasmin ; Method : Immunoturbidimetry; Sample : Serum
Ceruloplasmin is used in evaluation of chronic active hepatitis, cirrhosis and other liver disease. In Wilson disease, there is decreased ability to incorporate copper into apoceruloplasmin.Ceruloplasmin is low in Menkes' kinky hair syndrome, and with protein loss such as the nephrotic syndromes, malabsorption, and with some cases of advanced liver disease in which decreases of serum proteins have occurred. Ceruloplasmin is high in a variety of neoplastic and inflammatory states, since it behaves as an acute phase reactant. Serum levels are increased with copper ingestion.

Status : INTERIM REPORT ** End Of Report **

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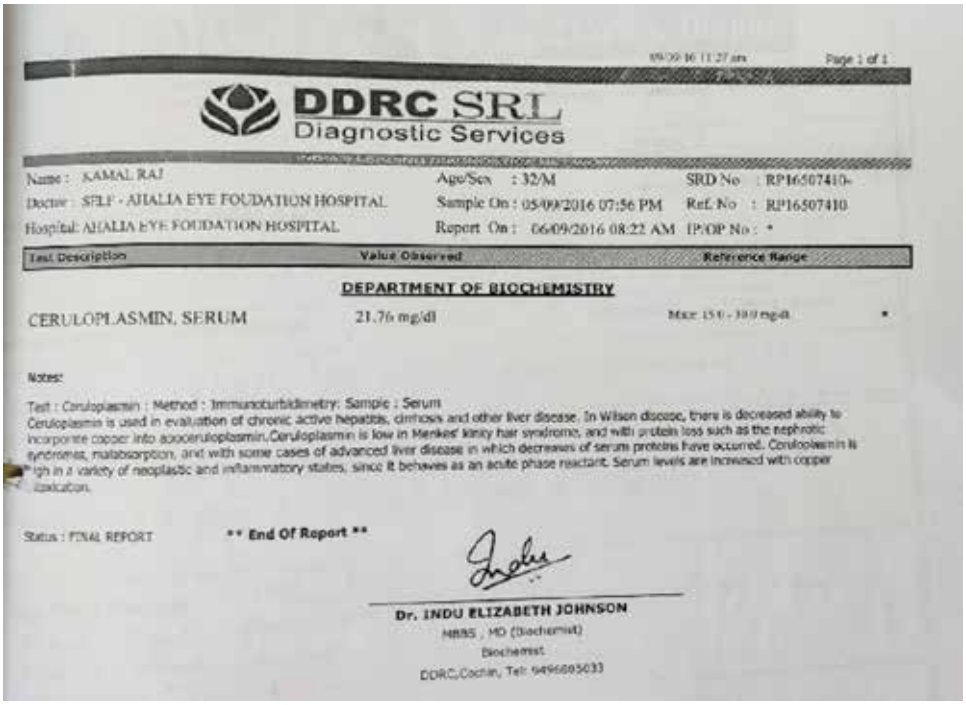
Fig 2



DDRC SRL
Diagnostic Services

Name : KAMAL RAJ | Age/Sex : 32/M | SRD No : RP16507410-
Doctor : SELF - AHALIA EYE FOUDATION HOSPITAL | Sample On : 05/09/2016 07:56 PM | Ref. No : RP16507410
Hospital: AHALIA EYE FOUDATION HOSPITAL | Report On : 06/09/2016 08:22 AM | IP/OP No : *

Test Description	Value Observed	Reference Range
DEPARTMENT OF BIOCHEMISTRY		
CERULOPLASMIN, SERUM	21.76 mg/dl	Male 15.0 - 30.0 mg/dL

Notes:
Test : Ceruloplasmin ; Method : Immunoturbidimetry; Sample : Serum
Ceruloplasmin is used in evaluation of chronic active hepatitis, cirrhosis and other liver disease. In Wilson disease, there is decreased ability to incorporate copper into apoceruloplasmin.Ceruloplasmin is low in Menkes' kinky hair syndrome, and with protein loss such as the nephrotic syndromes, malabsorption, and with some cases of advanced liver disease in which decreases of serum proteins have occurred. Ceruloplasmin is high in a variety of neoplastic and inflammatory states, since it behaves as an acute phase reactant. Serum levels are increased with copper ingestion.

Status : FINAL REPORT ** End Of Report **

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Fig 3

Discussion

Wilson's disease can't be correlated directly with any disease as per Ayurveda due to its diverse manifestation. In the present case, the generalized rigidity, hepatomegaly, and splenomegaly were correlated to Vatavyadhi, Yakrutodara, and Plihodara. The root cause of the manifested disease was Agnimandya. Agnimandya at the Jatharagni level further led to Agnimandya at the Dhatvagni level. This led to the blockage of channels and ultimately caused Vata vitiation and metabolic disorders. Vatavyadhi is Vataja disorder (occurred due to Vata Dosha) in which there is a contraction in body parts, stiffness, pain in the joints, rigidity at limbs, back, neck and head, insomnia, tremors, etc.

Initially to treat Agnimandya Chitrakadi vati was given. Thereafter, Abhyanga with Dhanwantaram Taila was planned along with Dashamoola niruha Basti and Sahacharadi mezhukupakam Anuvasana Basti in Yoga Basti pattern because for Vataja disorder Snehana (Oleation), Swedana (sudation) and Basti (Enema) have been mentioned as the best treatment. For Swedana mild Sweda opted in the form of Shastikashali Pinda Sweda.

Ksheerabala Taila and Shastikashali provide nourishment to the body and reduce vitiated Vata. Stabdhata (stiffness) was due to Sheeta (cold) Ruksha (dry) properties of Vata, Shastikashali is having Snigdha (unctuous) and Bruhmana (nourishing) effect, so, along with Ushna (hot) property of Swedana (sudation), it helped in pacifying Vata Dosha thus helped in relieving Stabdhata (stiffness) of the patient. Niruha Basti (Decoction enema) and Anuvasana Basti (oil enema) opted with the drugs that were having Bruhmana effect so it nourished the body and pacified Vata Dosha.

Ayurveda attributes a great role to diet as part of treatment. A proper diet containing milk, Ghee and Shalishashti rice was advised. The patient had advised to avoid dietary sources enrich in copper such as seafood (especially shellfish), whole grains, organ meats (e.g. liver), legumes (e.g. beans and lentils), cereals, potatoes, peas, mushrooms, chocolate and nuts (including peanuts). Ayurvedic literature describes kāmāla as a Pitta dominating liver disease. Acārya Caraka describes Kāmāla as next stage of untreated Pāṇḍu. This case serves as a perfect example wherein the classical line of approach which includes mṛdu śodhana and prescribing tikta (bitter) medicines gave the desired result. Paṭolamūlādi Kaṣāya, a tikta auṣadha (Bitter medicine), is a potent ayurveda medicine used in Kāmāla because tikta rasa reduces pitta. This medicine, especially due to its drug combination consisting of Paṭola, Triphalā, Śuṇṭhi, Kaṭukarohini etc. performs detoxificatory action on liver. Vilwadi gulika is a drug that reduces āma, improves and sustains agni. Abhayarishtam along with punarnavasavm is most commonly used Ayurvedic medicine and is known to address most of the symptoms which arise during Kāmāla like, aruci (Lack of taste or appetite), pāṇḍu (anaemia), agnimāndya (reduced hunger) and general weakness. Gandharvahastādi Eraṇḍa taila was used in this patient initially for doṣa anulomana. Being a snigdha mṛdu virecana dravya, gandharvahastādi eraṇḍa made sure the purgative action did not antagonize vāta doṣa and removed excessive systemic pitta. Liv 52 is a propriety medicine contains Bhūmyāmalakī (Phylanthus niruri). Bhūmyāmalakī is known to have hepatoprotective and hepato corrective property. It reduces pitta and detoxifies the liver.

Conclusion

From the case study it can be concluded that the drugs which correct the metabolism as a major role to play in wilson’s disease. Ayurveda therapies like Basti and Virechana also plays an important role in the management of the disease.Ayurvedic approach and diet modification have shown good results in reducing symptoms. Wilson disease is a rare, autosomal recessive inherited disorder of copper metabolism and comes under Sahaja Vyadhi or Vata Vyadhi in Ayurveda. Ayurvedic line of management gives satisfactory physically and mentally improvement. Patient got relief in many abnormal

symptoms i.e. Strength in rt.upper limb improved.Reduced symptoms such as tremors of both upper limb, difficulty in swallowing and generalized stiffness.

After treatment, the symptoms were reduced so, it can be concluded that the Ayurvedic approach and diet modifications in such patients may help in providing supportive care and improving the quality of life.

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