WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
SJIF Impact Factor 8.084
Volume 13, Issue 2, 657-666. Case Study ISSN 2277–7105

SUCCESSFUL AYURVEDIC MANAGEMENT OF ADULT-ONSET STILLS DISEASE (AOSD): A CASE REPORT

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Article Received on
29 November 2023,

Revised on 19 Dec. 2023,
Accepted on 09 Jan. 2024

DOI: 10.20959/wjpr20242-30964

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ABSTRACT

Adult-onset still's disease (AOSD) is a rare clinical entity with unknown etiology, typically characterized by a clinical triad of daily spiking fever, evanescent rash and arthritis. There are no specific diagnostic tests for AOSD, so the diagnosis of AOSD remains one of exclusion and the differential diagnosis may be lengthy. It has resemblance with *Gambira vatarakta* mentioned in ayurvedic classics. Conventional therapy used in this condition is unsatisfactory and is not free from side effects. A case of steroid dependent, young adult, having complaints of fever, inflammatory polyarthritis, joint stiffness, rashes were clinically diagnosed as AOSD and intervened successfully with Ayurvedic modalities is presented here.

KEYWORDS: Ayurveda, Adult-onset stills disease (AOSD), *Gambira vatarakta*.

INTRODUCTION

Adult-onset still disease (AOSD) is a rare systemic inflammatory disorder of unknown etiology. Its prevalence is less than 1/100,000 and it affects predominantly young people.[1] It is characterized by fever, skin rash, polyarthralgia's or polyarthritis, sore throat, hepatosplenomegaly, lymphadenopathy, leukocytosis, liver enzyme elevation, and high serum level of ferritin.[2]

According to *Ayurveda*, AOSD can be corelated to *Gambira vatarakta*. *Vatarakta* is a disease manifested by vitiated *vata* and *rakta* separately or combined.[3] According to Acharya Chakrapani, *Avagadha/Gambira* (Deep) and *Anavagadha/Uttana* (Superficial) classification is done for the purpose of treatment. "*Sarvaih lingaih*" insinuates combined clinical features of *Uttana* and *Gambira*.[4]

There are a number of diseases with different etiology that comes under the title of inflammatory arthritis, it is a challenging scenario to properly diagnose and provide appropriate management.

CASE REPORT

This is a case of steroid dependent, young adult, having complaints of fever, inflammatory polyarthritis, joint stiffness, rashes were clinically diagnosed and effectively intervened with complex ayurvedic modalities have been described. A substantial reduction in swelling, fever, stiffness was observed in 2 weeks. The improvement was assessed with diagnostic criteria and was remarkably evident through follow-up photographs.

Case discussion

Type of study

A case study

Clinical setup

Outpatient department of *kayachikitsa*, SDM Ayurveda Medical college, Udupi.

Age and Sex

A male patient of 29 years old.

Chief complaints

Patient presented with complaints of shifting type of pain, swelling and stiffness over bilateral foot, knee, and shoulder joints since one & half month. Associated with fever specially during early morning hours and red color rash over bilateral foot with dark red pimples.[figure 1][figure 2]

Medical history

- Flexura D
- Nervijen- NP

- Rabimor DSR
- Prednisolone 20 mg

Family history

No relevant family history

General examination

Bp-110/70 mmHg *Mala*: *Samyak Druk: Samyak*

Pulse: 70/min, regular *Mutra: Samyak Akruti: Madhyama*

Temp: 100 F *Jihwa: Lipta prakriti: Pitta-kapha*

Height: 168 cm *Shabda: Spashta Ahara shakti: Avara*

Weight: 56 kg *Sparsha: Ushna Vyayama shakti: Avara*

Nidra -Nidralpata patient was unable to sleep due to severe pain and fever.

Hetu observed

Consumption of eatables consisting of lavana (Salty), amla (Sour) rasa, virrudhashana (Incompatible food), divaswapna (Day-sleeping), prajagara (Night vigil), vega dharana (Suppression of natural urges).

Musculoskeletal examination

Inspection

- Swelling over B/L ankle and knee joints.
- Maculopapular rash over b/l feet (esp. during flare).

Palpation

- Warmth +++ [B/L ankle and knee joints]
- Pitting edema +++ [B/L ankle]
- Tenderness +++ [B/L ankle, knee,& shoulder]

Range of motion

- Shoulder joint- Possible with pain (Bilaterally)
- Knee joint- Restricted with severe pain ++(Bilaterally)
- Ankle joint-Not possible bilaterally (Pain & stiffness)

Diagnosis

The diagnosis of ASOD remains a clinical one. Unlike other systemic rheumatic diseases, it is not associated with rheumatoid factor (RF), antinuclear antibody (ANA) & cyclic citrullinated peptide (CCP). Common hematological abnormalities include leukocytosis, raised ESR & C-Reactive Protein (CRP) levels. Recently serum ferritin and glycosylated ferritin is also considered as a diagnostic tool and disease activity marker.[5]

For the diagnosis of AOSD, several sets of classification criteria have been published. They all have been developed from retrospective data and classify criteria as major and minor. Among them Yamaguchi's criteria[6] were shown to have more sensitive (93.5%). Diagnosis requires at least five features, with at least two of these being major diagnostic criteria.

Table 1: Yamaguchi's criteria.

Major criteria	Minor criteria
Fever at least 39C for at least one week	Sore throat
Arthralgias or arthritis for at least two weeks.	Lymphadenopathy
Nonpruritic salmon colored rash (Usually over trunk or extremities while febrile)	Hepatomegaly or splenomegaly
Leukocytosis (10,000/microL or greater), with granulocyte predominance.	Abnormal liver function tests.
	Negative tests for antinuclear antibody and rheumatoid factor.

Based on the above criteria.

Table 2: Assessment of Yamaguchi's criteria.

Fever at least 39C for at least one week	>39C
Arthralgia's or arthritis for at least two weeks	one & half month
Nonpruritic salmon colored rash (Usually over trunk or extremities while febrile)	Present
Leukocytosis (10,000/microL or greater) with granulocyte predominance.	Present
Abnormal liver function tests.	Present
Negative tests for antinuclear antibody and rheumatoid factor.	Negative

Hence, diagnosed as AOSD based on 4 major criteria and 2 minor criteria.[Table 2]

Table 3: Laboratory investigations.

1. TC WBC	13,900 cells/cumm
2. ESR	130 mmUhour
3.RA Quantitative(Serum)	Negative (2.40)
4.C-Reactive Protein (CRP)	7.89 mg/dl
5.SGOT(Ast)	39.51 U/L
6.SGPT(Alt)	48.70 U/L
7.Anti nuclear antibody	Negative
8.CCP Antibody	Negative
9. Serum Ferritin	397.9 ng/mL

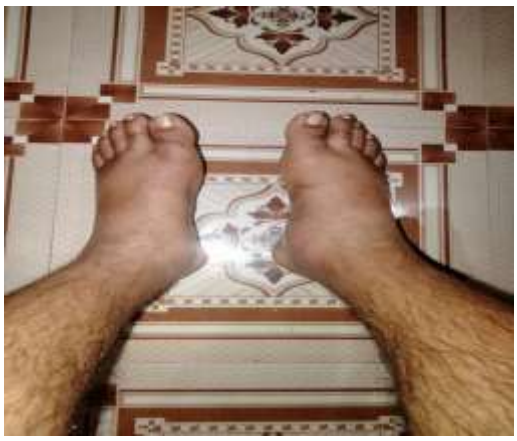

Figure 1: [Before treatment].

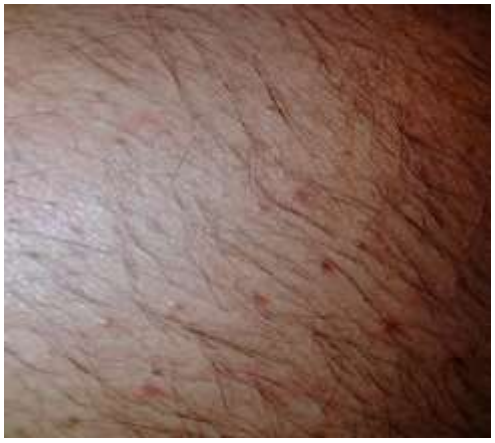

Figure 2: [Before treatment].

Case Conception and Selection of treatment

In this case, patient was under corticosteroid and analgesics for a period of 1 month but patient did not find any relief with the symptoms and also, he became aware of the disadvantages of corticosteroids from other sources, so he had chosen ayurvedic intervention for his condition. As there wasn't proper diagnosis initially, ruling out disease was of primary concern and later when patient was diagnosed with AOSD with the help of clinical and laboratory findings, *vatarakta* line of management was adopted. *Vatarakta* is a disease in which aggravated *vayu* gets covered by aggravated or increased *rakta* (Blood) on its way, thus obstructed *vata* vitiates entire blood.[7] The word "*Agnimaruthatulya*" describes the status of disease, that it attacks and develops very fast and is difficult to eradicate.[8]

The chief complaints of patient were severe pain and swelling over bilateral foot initially, which further developed to other joints. This presentation can be considered to be similar to that of "*akhuvisha*"[9] Other symptoms include *shwayathu sthabdatha* (Fixity and Hardness of swelling), *antharbrisharthiman* (Deep seated severe pain), *shyava tamra varna* (Reddish/Coppery color). Distinctive feature present in this patient was he had limping gait; this can be considered as "*karothi ganjam pangu va*".[10]

The principle of management mentioned in vatarakta is based on the presentations, i.e., *Bahya* or *Uttana* and *Gambira*. *Acharya charaka* has mentioned *vatarakta* as "*antarasrayam*" connotes that deeper *dhatus* leaving skin and muscles are involved in *Avagadha* or deep *vatashonitha*.[11] According to *Acharya sushruta*, just like *kushta* becomes deep, on passage of time *Uttana* (Superficial) can change to *Avagadha* (Deep).[12] In this case Uttana and Gambira line of management was adopted, which includes *Bahya alepa* , *virechana* and *samshamana aushadas*.[Table 4]

Table 4: Ayurvedic drugs prescribed to AOSD case.

Drugs	Dosage	Frequency	Duration
1.*Amavatari rasa*	1 tablet	TID before food	1-7th day
2.*Amruthotharam kashaya +eranda taila*	AK 15 ml+ ET 5ml	Twice a day before food	1-7th day
3.*Tab Trishun*	1 tablet	TID after food	1-7th day
4.*Tab mrityunjaya rasa*	1 tablet	TID after food	1-15thday
5.kokilaksha kashaya + eranda taila	KK 15 ml +ET 10ml	Twice a day after food	8-15th day
6. *Tab vatanashaka*	2 tablets	Twice a day before food	8-15th day
7.*Panchakola phanta*	30 ml	Twice a day before food	8-15th day
8.*Upanaha with grahadoomadi choorna +marmani vati+dhanyamla*	-	Twice a day	5-15th day

AK: Amruthotharam kashayam, ET: Eranda taila

OBSERVATIONS

During discharge following improvement was noted. fever was completely resolved with no recurrence. Joint pain and swelling over bilateral knee and shoulder was completely relieved. Swelling over bilateral foot was reduced remarkably and pain over ankle was reduced. Maculopapular rash and skin redness over bilateral foot was completely cured. Limping gait of patient changed to normal walking pattern.[figure 3]

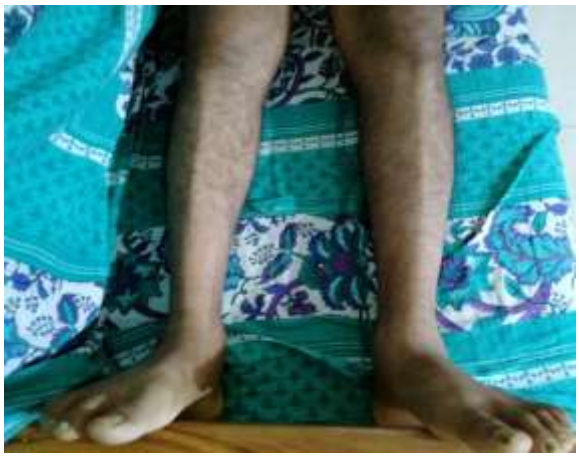

Figure 3: [After treatment].

DISCUSSION

"*Yadihasti tadanyatram*" according to *acharya charaka*, any concept told in *Samhita* we won't get anywhere and concepts told anywhere will be found in *Samhita's*. It is rightly proved through this rare case of AOSD. Based on the symptoms presented, AOSD almost mimics *Gambira vatarakta* and with the help of treatment modalities mentioned in *vatarakta*

chikitsa, we could eliminate the pathogenesis of disease from its root and prevent its progression.

Vata-rakta is an exquisite paradigm of *avarana janya vatavyadi*. Here pathology of *vatarakta* revolves around *avarana*. *Vata dosha* has *prasaranasheela guna* that helps the *vata dosha* to circulate even in the subtle channels of the body. On the other hand, *rakta dhatu*, also called as *Jivana* because of its *sara* and *drava guna*, is responsible for circulation. In the present case, due to the consumption of *lavana* (Salty), *amla* (Sour) *rasa*, *virrudhashana* (Incompatible food), *divaswapna* (Day sleeping), *prajagara* (Night vigil), *vega dharana* (Suppression of natural urges) both *vata dosha* and *rakta dhatu* got vitiated. *Dooshitha rakta dhatu* does the *avarana* of aggravated *vata dosha*, thus obstructed *vata* vitiate blood. Vitiated *vata* and *rakta* moves through *siramarga*, and gets obstructed in *sandhi* (Joint) because of the complex nature of its anatomical structure. Hence vitiated *vata* and *rakta* lodged in joints produce different pain sensations in association with either *pitta* or *kapha dosha*. Since it is a disorder of *vata*, it should be controlled by *vata* alleviating treatment. It is usually only in *Gambira vatarakta* that *vata* overpowers *rakta*. In this case, patient was advised to take medicines as per ayurveda principles and to taper the dose of corticosteroid and other analgesics and gradually corticosteroid and analgesics were totally ceased.

Even though etiology of AOSD is unclear, there is evidences of various mechanisms that contribute to the pathogenesis of AOSD. The activation and amplification of inflammation by innate immune cells, characterized by cytokine storm is the hallmark of AOSD. The formation of cytokine storm leads to “suicide attack” that not only contributes the elimination of pathogenic microorganisms, but also causes tissue toxicity. Adopting immunoregulatory therapies in order to inhibit hyper-activated inflammatory responses can resist cytokine storms. Initially the patient was administered *jwara chikitsa* along with *ama hara chikitsa* as fever was one of the major presentations. *Amruthotharam kashaya*, *amavatari rasa* and *mrithyunjaya rasa* were administered. *Amruthotharam kashaya* was given along with *eranda taila* to act as mild *virechana*. It also helps in *amapachana* and *vatanulomana*. *Amruthotharam kashayam* enhances *Deepana, pachana* and has effects on inflammatory reactions in body. *Amavatari rasa* is a combination of *chitraka, guggulu, triphala, eranda, Shuddha parada* and *Shuddha gandaka*. Maximum drugs in this formulation have *ushna veerya* and *Madhura vipaka*, which acts as *amapachana* and *vatanulomana.*

Trishun tablet which contains *sudharshana choorna* and *tribhuvana keerti rasa* also relieves pain, fever and also enhance immunity. In *sudharshana choorna*, *Swertia chirayita* is 50% of the total quantity of the remaining ingredients. It possesses good antioxidant potential and can be used as beneficial therapeutic agent for diseases associated with oxidative stress. Tribuvanakeerti rasa also has analgesic, antipyretic, antioxidant, anti-inflammatory properties. Eventually fever was reduced and there were remarkable changes in redness and swelling. *Upanaha* (External application of medicated paste) was applied to all affected joints, a combination of *grahadoomadi choorna*, *marmani vati* and *dhanyamla* was used. *Grahadoomadi choorna* which is *kaphavatahara*, *srothoshodana* and *shophahara* has a pioneer role in *kaphapradhana vatarakta*. *Marmani vati* which is *tridosha shamaka*, *pittagna* and *dahashamaka* acts in inflammatory pathology. *Marmani vati* and *grahadoomadi choorna* applied in the medium of *dhanyamla* which is having penetrating quality increases the potency of other ingredients. Later after a week, internal medications were changed to *vatanashaka vati*, *kokilaksha kashaya* and *panchakola phanta*. Key ingredients of *vatanashaka vati* are *devadaru* (*Cedrus deodara*), *guduchi* (*Tinospora cordifolia*), *mustaka* (*Cyperus rotundus*), *rasna* (*Pluchea lanceolata*), *pippali* (*Piper longum*), *shunti* (*Zingiber officinale*), *eranda* (*Ricinus communis*). Majority of these drugs are *jwaragna* (Alleviates fever), *kaphavatahara*, *hridya* and *ushna veerya*. Amrutha is the drug of choice for vatarakta according to charaka *agrya aushadha* and *Bhavaprakasha Nighantu*. *Vatarakta* being a *raktavahasroto vyadi*, *raktavahasrotogami* property of *guduchi* may be helpful here. *Tikta rasa* of *guduchi* subsides *rakta prakopa* and *Madhura vipaka* subsides *vata prakopa*. *Kokilaksha* is having property of *vatapittahara*. It has analgesic, anti-inflammatory, anti-arthritic activity. *Panchakola phanta* contains *shunti*, *pippali*, *pippalimula*, *chavya*, *chitraka* and when taken with *ushnodaka* is indicated in *mandagni* and *amakaphaja vikara*.

CONCLUSION

Adult-onset still's disease is a rare systemic autoinflammatory disorder of unknown etiology. AOSD still remains as a diagnostic dilemma for physicians as it presents with combination of nonspecific symptoms. The conventional treatment options are also not satisfactory and are not free from systemic side-effects. This observation endorses a step toward the practice of ayurvedic intervention in adult-onset stills disease.

ACKNOWLEDGEMENT

We are thankful to the participant for providing informed consent to publish the result of the study.

Conflicts of interest

There are no conflicts of interest.

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