Idiopathic Oedema: A Lesson in Differential Diagnosis

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Summary
This paper outlines our approach to the diagnosis of Idiopathic Oedema. The patient presented illustrates some of the pertinent clinical and laboratory pointers one has to take into consideration before labelling a person as suffering from idiopathic oedema. The discussion also includes a brief review of the literature on the patho-physiology and management of this benign disorder.

Key Words: Idiopathic oedema, Water-load test, Diuretics

Introduction
Peripheral oedema is a common problem. A wide range of systemic and regional disorders can result in fluid retention in the peripheries.

Idiopathic oedema is a diagnosis of exclusion. It is characterised by excessive accumulation of interstitial fluid in the absence of cardiac, renal, hepatic or thyroid disease; venous or lymphatic obstruction; angioedema; or other causes including, hypoalbuminemia, side effects of drugs, hypersensitivity reactions and premenstrual syndrome. Simple clinical assessments in the form of diurnal weight charts and a positive water loading test help to place the diagnosis on a more firmer ground.

The patient presented here highlights some of the salient features of the syndrome.

Case Presentation
A 40-year-old woman presented with a ten-year history of generalised body swelling.

She developed painless swelling of legs, about ten years prior to consultation. This was accompanied by puffiness of hands and a bloating sensation in her abdomen. The symptoms were somewhat worse in late afternoons and evenings. On certain days, she was unable to remove her rings prior to her evening baths. She had not noticed any facial swelling.

The systemic review was normal. She denied any cardio-respiratory, renal, hepatic or thyroid-symptoms.

There was no history of drug, alcohol or tobacco abuse. She had not taken nonsteroidal anti-inflammatory agents (NSAIDS) or oral contraceptive pills.

She was married with 4 children, the youngest being 15 years old. She was not noted to have high blood pressure or “dirty urine” during her pregnancies.

Physical examination revealed a pleasant but anxious looking lady who was alert and cooperative.

The most striking feature was the presence of gross pitting bilateral pedal oedema. Her fingers too looked puffy. The examination was otherwise normal.

Laboratory evaluations including complete blood counts, erythrocyte sedimentation rate, fasting and postrandial blood sugar, liver function tests and renal profile were normal. The thyroid function tests (serum T₃, T₄ and
thyroid stimulating hormone) and lipid profile were normal. The electrocardiogram, urinalysis, chest X-ray and ultrasound examination of abdomen were unremarkable.

The patient was then asked to record her weight twice a day for a week - i.e. once as soon as she woke up in the morning and second reading at 7.00 pm. Her weight chart showed a striking diurnal variation with the evening weight being more than her morning recording on an average of 1.8 kg.

She was then subjected to a water-load test under strict supervision as a lodger in the ward. (Figure 1). She excreted 90% of the water-load in the recumbent position, but only 53% when the test was repeated in the orthostatic position on the following day.

The patient was advised on moderate salt and carbohydrate restriction, and told to avoid tight fitting garments.

She was then started on spironolactone 50 mg in the morning. Two weeks later, the dose of spironolactone had to be stepped up to 100 mg/day.

The above measures helped to relieve the patient of her oedema. She felt much better a month later.

**Discussion**

Oedema represents an excess accumulation of fluid in the interstitial tissues which can be the result of an interplay of a host of factors including increased capillary permeability, obstruction of the venous or lymphatic flow from the involved part and the increased accumulation of fluid in the tissues because of lowered oncotic pressure of the plasma. One needs to exclude an extensive list of possibilities before diagnosing idiopathic oedema.

A detailed history and thorough physical examination narrow the field of possibilities. In the patient presented here, the duration of symptoms alone eliminated several systemic causes of peripheral oedema. She had been oedematous for at least ten years at the time of consultation without any features to suggest a systemic cause. Thus the possibility of her oedema being of cardiac, renal, hepatic or hypoproteinemic origin was remote on the basis of her clinical presentation alone. This clinical impression was reinforced by the normal or negative results of the extensive investigations done on her.

Thus, by a process of elimination, our patient was thought to have idiopathic oedema, which is an ill defined, poorly understood syndrome. The exact aetipathogenesis is not known. Several postulates have been put forward including increased capillary leak during upright posture, excessive renin-aldosterone activity, increased antidiuretic hormone secretion and diuretic abuse.

As illustrated by our patient, idiopathic oedema occurs almost exclusively in women during their reproductive years. It is a benign condition. Even on prolonged follow up, patients with idiopathic oedema do not develop features of an underlying organ damage.

The *sine qua non* of idiopathic oedema is the excessive weight gained by the patient by the end of the day. The weight would have returned to its baseline level by the following morning as a result of excessive nocturnal enuresis during recumbency position. Outpatient’s diurnal weight chart elegantly demonstrated this classical phenomenon of the disease.

Though, there are no specific laboratory tests to confirm idiopathic oedema, the water-loading test helps to substantiate the diagnosis.

As shown in Figure I, the water-loading test is simple, non invasive and can easily be performed on an outpatient basis. It is based on the physiological principle that normal persons would be able to excrete more than 65% of water ingested independent of posture. Patients with idiopathic oedema however have a substantial decrease in their ability to excrete water taken during upright position but handle the water load effectively during supine position.

There is no specific treatment for idiopathic oedema. These patients need repeated reassurance of the benign nature of their condition.

Patients should be advised on moderate salt (2gm/day)
IDIOPATHIC OEDEMA

Figure 1

WATER-LOADING TEST.
(Results of patient’s test are given in brackets)

Preparation;
1. Calculate ideal body weight (50 kg)
2. Discontinue diuretics at least for two weeks prior to the test.

Procedure.

Day 1
1. Patient is instructed to drink 20 ml/kg of water (calculated at ideal body weight) over 15 minutes at 8.00 am following an overnight fast (50 x 20 = 1,000 ml of water)
2. Empty the bladder immediately
3. Collect all urine during the next 4 hours (900 ml)
4. Patient is to remain supine throughout the test.
5. No alcohol/tobacco is allowed during the test.

Day 2
Repeat the test as Day 1 with the patient remaining upright during the procedure.

(Volume of urine voided at the end of 4 hours = 530 ml)

Interpretation-of-test.

Normal;
More than 65% of the water consumed can be excreted regardless of patient’s position during the procedures.

Idiopathic Oedema:
Less than 65% of water ingested is excreted when the patient remains upright in Day 2.

False Positives.
1) Obesity
2) Adrenal insufficiency
3) Autonomic insufficiency
4) Thyroid disorders
5) Syndrome of inappropriate antidiuretic hormone secretion (SIADH)

NB: It is not necessary to admit the patient for the test.
It is sufficient to lodge the patient during the procedures for adequate supervision.

and carbohydrate restriction. Avoidance of excessive standing and tight garments too may be of help. Elastic support stockings may be useful1-3.

Currently, the agent of the preference in idiopathic oedema is spironolactone followed by thiazide diuretics3. Both these drugs are used in a stepped-care fashion.

Treatment is usually commenced with spironolactone 50mg in the morning. Depending upon the patient’s clinical response it can be stepped up to 150mg/day. If a patient becomes refractory to spironolactone, then a small dose of thiazide diuretic (e.g. hydrochlorothiazide 50mg) can be added to the regimen, usually given at 7.00pm or 8.00pm. This is to ensure the patient has a brisk diuresis by 11.00pm and so that her sleep is not disturbed3.

Potent loop diuretics should be avoided. Long term use of frusemide in idiopathic oedema has been known to damage renal tubules and/or interstitum resulting in acute renal failure.

Sympathomimetics, ACE inhibitors, Betablockers and Bromocriptine too have had their advocates in the management of idiopathic oedema3.

The current therapeutic approaches admittedly do not address the basic issues involved in idiopathic oedema. They, however bring symptomatic relief to patients with this enigmatic condition.
CASE REPORT

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Reference

