

Hyperstomy syndrome

Síndrome de hiperostomia

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Abstract

Hyperstomy syndrome is defined as a dysfunction of microcirculatory arteriolo-venular flow which stimulates ischemic diseases without arterial occlusion and causes precocious venous backflow. This paper presents a review of hyperstomy syndrome since this entity was first described. It also presents six cases of patients with hyperstomy syndrome who were successfully treated.

Key words: ischemia, lymphedema, microcirculation.

Resumo

A síndrome de hiperostomia é definida como uma disfunção do fluxo microcirculatório arteriovenoso que provoca doenças isquêmicas sem oclusão arterial e causa refluxo venoso precoce. Este artigo apresenta uma revisão da síndrome de hiperostomia desde a sua primeira descrição. Além disso, são apresentados seis casos de pacientes com síndrome de hiperostomia tratados com sucesso.

Palavras-chave: isquemia, linfedema, microcirculação

Hyperstomy syndrome is defined as a dysfunction of microcirculatory arteriolo-venular flow which stimulates ischemic diseases without arterial occlusion and causes precocious venous backflow. Currently, it is considered that intense lymphatic repercussion is possible in some cases, turning malignant some ulcers of the lower leg.

Pratesi and Malan¹⁻⁴ were the first to describe this syndrome, which was called hemometakinesis by De Bakey.⁵ In 1968, Amir-Jahed^{6,7} presented as angiodyskinesia a similar syndrome associated mainly with venous problems and affecting young females after the use of contraceptive pills. Based on these findings, Dramez et al.^{8,9} have successfully treated the syndrome by intraarterial injection of procaine. At the same time, Haimovici^{10,11} observed similar findings in

angiographic studies of arteriovenous communication. In 1976, Mayall¹²⁻¹⁵ devoted an entire monograph to the treatment of this entity, and called attention to the syndrome by describing lymphedemas with dysregulated arteriolo-venular flow on angiography and angiographic findings in 56 patients, in addition to some cases of malignant Marjolin ulcers by cancer invasion.

Etiopathogenesis and pathophysiology

Microcirculation (Zweifach-Neleman) is responsible for maintaining the blood flow through the capillaries, both at rest and during muscle work. Using electronic microscopy, Redish & Tangco¹⁶ have shown this arteriolo-venular microcirculation. It is controlled by the neurovegetative system, which regulates the closure and opening of the channels responsible for venular and lymphatic hypertension. Arteriolo-venular microcirculation can be observed using high speed arteriography, vein oxymetry, or by observing the venous pressure on clinostatism, which can be measured by Doppler up to 30 mmHg. In addition, microcirculation can also be observed by the increase in skin temperature due to the arterialization of the veins

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warmed to evaluate increased oxygen tension and reduced CO₂ tension.

Using arteriography, after injecting 60 ml of dye in the inguinal artery one must wait 6 seconds for the warm venous backflow to fill the femoral vein, 7 to 8 seconds for the knee, and 10 to 12 seconds for the leg all the way to the feet; to clearly see the arteries of the legs, it is fundamental to have the leg on internal and external rotation, so as to see the three arteries clearly separated from the two bones. In addition, to be able to observe the venous backflow on the radiograph plates, it is important to see the abnormal branching of the main arteries, the femoral artery and the three arteries running along the leg up to the foot. It is also important to observe the direction, if it is horizontal or vertical, instead of following the acute angle that is normal in most cases, and to observe if the flow is delayed or not, especially when hyperstomy is due to the effect of closure or opening of the preferential microvessel channels and/or of the glomus by the neurovegetative sphincter effect on alpha- adrenergic control.

Etiologic factors

The most important are: trauma of the soft tissues in the extremities; bone fractures; postphlebotic syndromes; venous insufficiency; arterial occlusion; reduction of arterial pressure in muscle arterioles; vasomotor syndromes without arterial lesion; arterial syndromes following Raynaud's phenomenon; and also obstruction of lymph nodes closer to the proximal roots.

Symptoms

The symptoms of hyperstomy syndrome depend on several factors and on the intensity of each case.

Pain: pain is manifested only as paresthesia or light pain, distension of the skin, and intermittent claudication not quickly improved by rest, as in venous or arterial insufficiency.

A slight increase in local temperature. If left without treatment, hyperstomy syndrome may develop into elephantiasis, as occurs in venous or lymphatic diseases; hyperstomy syndrome can be the reason or the effect of thrombosis in arteries. Cold may be felt below the extremity, and warmth on the hyperstomy area, according to newly formed veins.

Sex: In Iran, where contraception is official, hyperstomy syndrome has been observed to be more frequent in young females using contraceptive pills. Our experience shows that males are more frequently affected, at a 2:1 rate.

There seems to be no association between the intensity of subjective symptoms and objective findings; recidivation of venous ulcers is easily cured after hyperstomy treatment.

Oscillometric readings: normally increased in the absence of edema.

Oxymetry: shows arterialization of venous blood near the location of the arteriolo-venular dysfunction, not as intense as in arterio-venous (AV) fistulas.

Distal pulses: can be expanded with decreased intensity according to the number of hyperstomies.

Local signs and symptoms of venous insufficiency are more evident depending on the size of the hyperstomy.

There is also an increase in venous pressure, although not as intense as in AV fistulas.

Tissue O₂ consumption is increased in hyperstomy. Fontaine et al.¹⁷, using Xenon 133, have observed increased depuration of blood flow on the calf volume.

Arteriography: Should be performed if possible, on fast serial and with a chronometer over the radiograph plates, to register exactly how many seconds were required for the image to appear on the plate after the beginning of the dye injection. This should be done instead of using the automatic radiography changer, which fails to register the time lost when there is the need for a bigger interval to see the late backflow, which is well-registered on the plates by the chronometer. In addition, this allows a change in position to see from the aorta down to the foot; for that, an additional 6 seconds are lost.

The most typical findings in the inguinal femoral artery, commonly by handmade pressure injection of 60 ml of contrast are:

- a) Simultaneous filling of the superficial and deep femoral arteries and veins in the thigh within 3 to 6 seconds. This is important in patients with large-sized artery occlusions, when more than 15 seconds of interval may be necessary for the leg to be seen. This time is not registered, for example, on the automatic changer of Schonänder, but the hand chronometer shows that exactly on the plate.

- b) A blurry or smudgy appearance of the muscle mass around the arteriolar branches during the second (or arteriolar-capillar) phase, in the places corresponding to 7 to 9 seconds, is an unusual symptom of hyperstomy, as well as high temperature and oscillometric increase.
- c) Abnormal artery size, number, extension and direction, often going up instead of making an acute angle with the exit of artery branches.
- d) Incomplete or delayed filling by the contrast dye of the distal branches of the arterioles of the leg due to the derivation of the blood flow through the short circuits, commonly in the thigh and calf, in the posterior tibial branches and in the superficial femoral artery. For an accurate mapping of these abnormal branchings, it is advisable to place the X-ray plates so as to allow internal and external rotation of the foot to clearly show the position of the tibia and fibula bones, and to avoid superposition with the tibial and fibular arteries. It is important to remember that the normal X-ray plate shows only vessels larger than 0.10 mm. Therefore, a normal X-ray will never accurately show the newly formed hyperstomic arteriolar-venular shunts, whose terminal size is 30 to 60 micra.

Differential diagnosis

Congenital arterio-venous fistulas

Arruda¹⁸ wrote that these fistulas are commonly present since birth, or they may appear around puberty. They present strawberry marks, angioma and abnormal direction of the large varicose veins, with increased size of the leg before puberty, as is very frequent in Klippel-Trénaunay Syndrome,^{19,20} often with dysgenesis of deep veins. There is also an immediate, fast and precocious venous-varicose backflow. Some patients present more atypical arteriolo-venular congenital angiodysplasias with the following features:

- The fading of the arterial contrast phase is more intense distally in relation to the fistula. If it is a case of hyperstomy, the distal filling is more delayed;
- The spotty appearance in the fistula area is much more visible and intense than the blurry appearance around hyperstomy branches;
- The big dilatation of surrounding veins is absent around hyperstomy branches. There is only a precocious venous backflow, but the veins show a

normal shape or only small phlebectasy in more advanced cases;

- AV fistulas are commonly diffuse around the limbs. In hyperstomy, branches are more localized. Bone lesions are common in AV fistulas. If AV fistulas are hypoactive, the radiologic signs are very similar to those of hyperstomy, but in the active and bigger AV fistulas, the arteries are more convoluted and open directly into the veins of great size. The communication with the veins is very visible, showing that the problem is not in the microcirculation. Because of this, bigger veins and heart insufficiency are especially common in AV fistulas, where the Nicoladoni-Branham sign of bradycardia after local digital compression is always present.

Occlusive arterial diseases

Commonly they are the cause of the appearance of hyperstomy. This is important for surgeons to remember – it is the reason why a lumbar sympathectomy can worsen the ischemia in extremities, as frequently observed, producing paradoxical gangrenes. A ganglioplegic spinal block can be made before surgery in sympathetic spinal ganglions using chlorpromazin 25 mg, intra muscular injection, or a spinal block with 20 ml of procaine 1% in saline. Low body temperature measured by skin thermometry is a contra-indication for this operation.

Tumors of soft tissues

The early symptoms of hemangiosarcomas on the thigh or calf are painful. Arteriography shows a big anarchy, similar to what is observed in more severe cases of hyperstomy, as a good mapping for biopsies. In Marjolin ulcers, due to epidermoid carcinomas, arteriography always shows the abnormal findings of intense hyperstomy, whose radical correction has helped long term cure after a skeletization of all hyperstomies to kill the tumor by preventing arterial feeding.

Treatments

Conservative

Since in young people the problem may be functional, it is advisable to wait for some weeks or use a placebo treatment. According to Dramez et al.,⁸ intra-

arterial procaine injections have given good results in the long term.

In the majority of cases secondary to ischemia we recommend treatment by intra-arterial injections of hydrogenated alkaloid derivative ergotoxin, together with injections of procaine and oral intake of ergotoxin derivatives²¹ (100 g capsules three times a day). In patients with hyperstomy secondary to venous stasis and with lymphedema, mainly with venous problems, we always recommend the correction of venous insufficiency by elastic compression of highest pressure possible, day and night.

With larger ulcerations, in addition to the etiological treatment, the main symptoms must be treated as well, mainly pain, with strong analgesics administered intravenously, if necessary. When the main problem is to reduce the excess of exsudation, drug treatment is recommended, with derivatives of bovine peptides of coagulation factor VIII, in pills of 0.125 mg, three times a day, in addition to phlebotonics. Other possible drugs are micronized pills of diosmin 450 mg with flavonoid titulated hesperidin 50 mg, two pills each day in the morning, and pills of benzopyrone 15 mg (coumarin) troxerutin 90 mg, six times a day, followed by hand massages on the trunk, inguinal region and legs, with a 40 ml cream of benzopyrone 200 mg; 300 or 500 mg pills of heparin 2000 UI, a day of B-hydroxyethyl-rutosides, six pills a day. To avoid infection, exsudation is reduced mainly by local bandages, closed with a transparent biological cellophane sheet. This significantly decreases the number of bandages required and the speed of scar formation. As the wound becomes anaerobic and aseptic, it is not necessary to use antibiotics. Before resuming walking, the patient must wear elastic stockings for a long period.

Surgical treatment

If the conservative treatment is not enough for the correction of hyperstomy, we recommend skeletization of the main arteries whose branches are responsible for the typical symptoms in the legs or arms. This is possible thanks to the accuracy of diagnosis by selective and fast serial arteriography. Diagnosis may also be based on phlebography, lymphography, scintigraphy or other imaging methods, such as Duplex scanning echo Doppler, helicoid tomography or magnetic resonance (Figure 1).



Figure 1 - Magnetic nuclear resonance (August 8, 2001). Atherosclerotic renal artery. Left iliac artery, femoral artery and early vein filling on the leg.

Below we present some cases of patients presenting hyperstomy who were successfully treated.

Case 1

This is a 46-year-old woman. Following surgery for Brown dermatome for lymphedema of the leg, she developed a great lymphedema over the entire thigh after erysipelas and lymphangitis with marked incapacity during 15 years due to excessive weight of the mass (Figure 2).

A phlebography of the leg revealed marked compression by excessive weight over the femoral vein in the thigh. This was corrected by elevation of the entire mass (Figure 3).



Figure 2 - Great lymphedema of the thigh, on the right and after 15 days post-op.

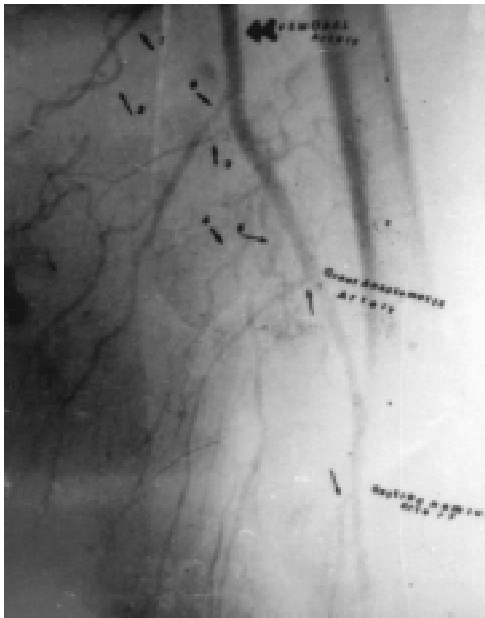


Figure 3 - Phlebography of the left thigh. The arrow shows compression of the vein.

A femoral arteriography revealed six new abnormal hyperstomy branches in Hunter's canal (Figure 4).



Figure 4 - Arteriography of the left thigh, six abnormal hyperstomy branches.

After ligation of the six abnormal branches, the mass shrank significantly. It was then easier to resect (lumping operation) the excessive tissue in the main part of the thigh. The thigh was markedly improved.

In 1993, 19 years after the surgery, and in 1999, without any kind of treatment, there was a small relapse of the lymphedema of the leg, but the patient is ambulant and fully active (Figure 5).



Figure 5 - 19-year follow-up.

Case 2

This is a 43-year-old man who had a small intractable lymphedema with a big ulceration recidivate after a traumatic skin injury to the foot and ankle when he was 2 years old. Lymphography depicted mild lymphedema; phlebography showed tortuous phlebectasis (Figure 6). Arteriography showed intense hyperstomy branching of the anterior tibial artery, just below the ulceration (Figure 7). A biopsy showed a squamous cells (epidermoid) carcinoma (Marjolin ulcer).

After seven days of chemotherapy (methotrexate + cyclophosphamidid + fluoruracil + prednisone), all the soft tissue of the ulcer was excised with all the six branches of the anterior tibial artery through a selective infusion on the popliteal distal artery (Figure 8).

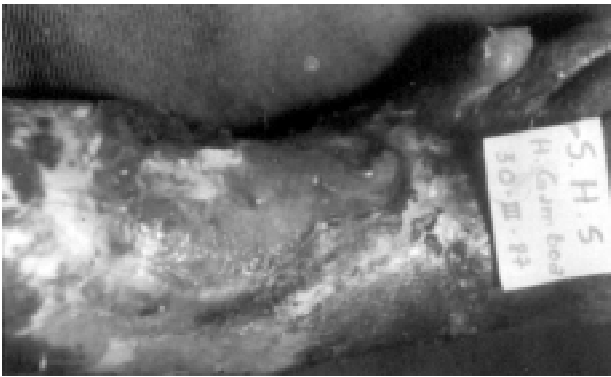


Figure 6 - Small intractable lymphedema. Large ulceration on March 30, 1987, present since 4 years of age.



Figure 8 - Ulcer cured on May 14, 1987 with Thiersch graft.



Figure 7 - Arteriography: large hyperstomy. Black arrow shows six abnormal branches.

(Figure 9). Conventional lymphography by hydrosoluble dye, not oil, showed only two lymphatic branches on the internal border of the skin flap.

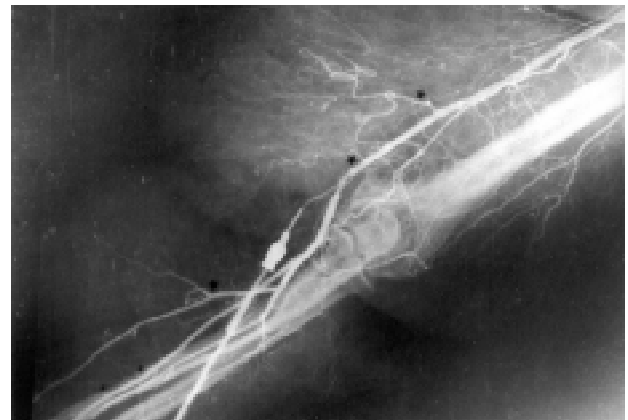


Figure 9 - Arteriography of the left arm, abnormal branches of hyperstomy.

In May 1997, after 10 years, the patient was cured without relapse or metastasis of the tumor and edema on the leg. He required no special treatment except for elastic stockings.

Case 3

This is a 55-year-old woman presenting a big lymphedema of the left arm after radical mastectomy followed by radiotherapy. After conservative therapy, a phlebography revealed severe stenosis of the axillary vein. Contrast injection into the brachial artery showed an abnormal six-branch arteriolo-venular hyperstomy

During a small lumping operation on the arm for resurfacing of the skin all the arterioles with hyperstomy were resected. Complete debridement was performed around the axillary vein. Only one lymphatic-vein anastomosis was done. After surgery, the perimeter of the arm was reduced from 87 to 32 cm (Figure 10).

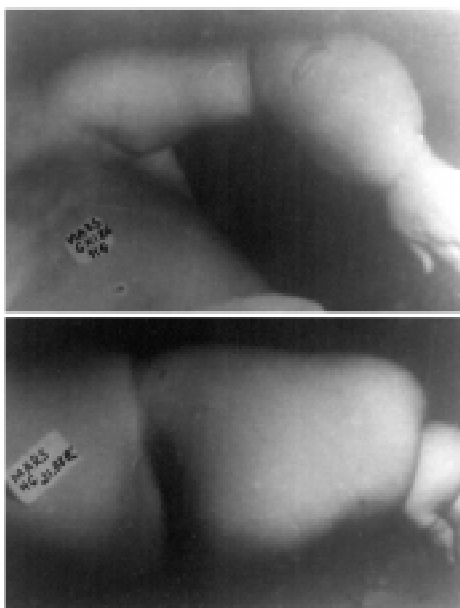


Figure 10 - Big lymphedema of the left arm, 86 cm in perimeter. Afterwards, improved lymphedema of the left arm, 32 cm in perimeter following surgery.

Case 4

This is a 70-year-old woman seen at Hospital da Gamboa on July 19, 1997, presenting serious necrotic ischemic plaques (Figure 11), infected and ulcerated, on the left leg's antero lateral border. She also presented an infected ulcer on the internal ankle of the same leg and a plantar ulcer resulting from type II diabetes. We observed segmental obliterans arteriosclerosis on the left internal iliac artery, occlusion of left kidney artery and a 6-cm atherosclerotic segment in the superficial femoral artery, as well as segmental occlusion of the three arteries of the leg with good collateral circulation filling the foot. Arterial hypertension MX 200 min 110. Initially, the indication was for amputation. We decided to treat the patient using conservative methods first, and began intravenous administration of sodium heparin 5,000 units every four hours, day and night, followed by broad spectrum antibiotics, guided by repeated antibiograms with selective sensitivity.

The patient was kept in absolute rest, in bed, with slight elevation of the side of legs, due to the presence of severe superficial and deep varicose veins, without

thrombosis on phlebography. An excellent arteriography and aortography magnetic nuclear resonance confirmed the arterial lesions and also the great hyperstomy lesions below the left leg. For this reason, we treated the patient with 100 mg of oral ergotoxin derivative, diosmin 450 mg pills, and oral benzopirone derivatives as recommended by Casley Smith.²² When infection was controlled and a good granulation appeared on the ulceration, we made several split skin grafts with material from the thigh (Thiersch method), with excellent results (Figures 12, 13). In December of 2002 she was able to walk for ambulatory treatment. Since the patient presented hypochromic anemia, she received iron sulfate and 500 ml of blood. Only 2 cm of the graft were lost in the foot, and the perforant plantar ulcer relapsed. Because of the intense presence of hyperstomy around the lower part of the left leg's anterior tibia, we decided to submit the woman to surgery at this point to



Figure 11 - Necrotic ischemic ulceration (July 16, 1997).

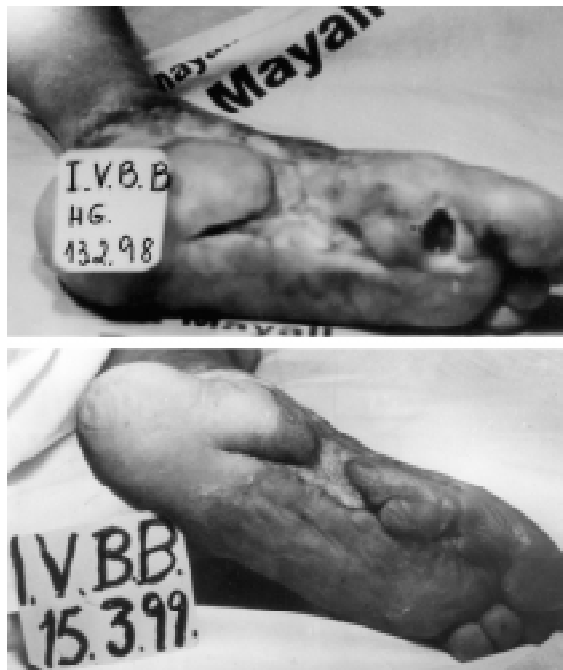


Figure 12 - Necrotic ulceration of malleolar before in February 2, 1998, and in March 1999.



Figure 13 - Necrotic ulceration of malleolar (July 1997). Ulcer cured (March 15, 1999).

avoid venous hypertension around the vein and in the foot area. This was being controlled by 30 mgHg elastic bandages to allow walking. A simultaneous operation of the left kidney artery and leg hyperstomy syndromes was planned to cure arterial hypertension.

Case 5

A 30-year-old man with lymphedema of the right lower leg secondary to erysipela and lymphangitis was admitted to Hospital da Gamboa for conservative treatment with local hygiene, low-salt diet, massage and exercise followed by elastics bandages and antibiotics.

An arteriography of the retrograde retromalleolar posterior tibial artery of the right leg showed an intense hyperstomy on the three ascending branches with indication for surgery (Figure 14). After ligation of the three abnormal branches in the right leg, hyperstomy was easily controlled with elastic stockings. Fifteen days after the surgery, he was considered well enough to work normally. In 1999 (37 years later) he came back for a check up without lymphedema and capable of leading a normal life, always wearing elastic stockings and following a low- salt diet. There was no relapse of the erysipela and lymphangitis.

Case 6

This is a 25-year-old black woman who presented with a grade III lymphedema on the right arm (Figure 15). She was treated during 6 months at Policlínica do Botafogo with conservative methods, without any improvement. One night, she was observed at the clinic to be sleeping with a hard cotton ball under the right axilla, compressing all the axillar vessels.

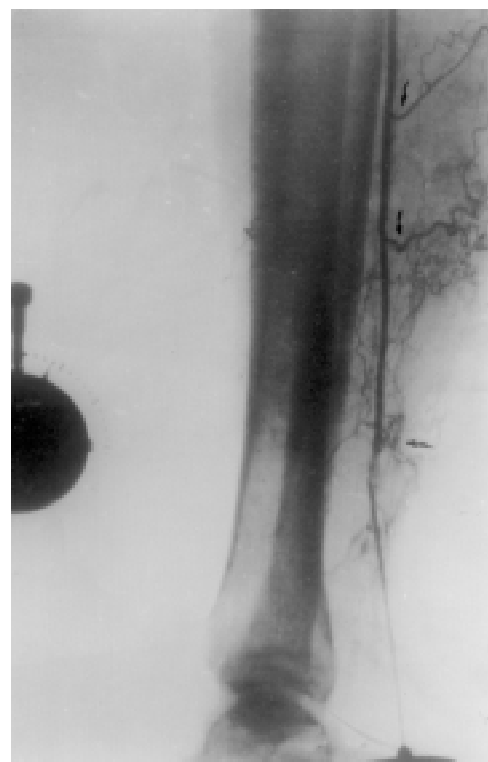


Figure 14 - Arteriography of retrograde posterior tibial artery, three abnormal hyperstomy branches.



Figure 15 - Voluntary lymphedema before and after treatment.

When asked about this, the woman said that she did this voluntarily, because she had problems with her employer, who would not allow her to walk with her boyfriend. She decided to sleep every night on her side, over the right axilla, which was responsible for the voluntary lymphedema. Then, she was immobilized with the right arm raised using a plaster of Paris, with elevation of the arm 45 cm above the level of heart. She was able to stay with the arm in hyper abduction during 45 days, when the visible fingers no longer presented lymphedema. The plaster was removed. To our great surprise the lymphedema was cured. She also received psychological advice. She has not used any drugs, only a low-salt diet and elevation of the right arm.

This case demonstrates that to resolve a severe lymphedema of many months with eight hours of voluntary compression daily, it was only necessary to immobilize the arm in hyperabduction, without any need for skin hygiene, manual drainage and exercise.

This woman was able to cure the lymphedema without bathing during 45 days, without any local hygiene, no drugs and no elastic garments. This is the reason why in our daily routine we always recommend the maximum rest possible, and elevation of the limb, which was fundamental in this case, supported by psychological advice.

Comments

Peripheral hyperstomy was initially described by Pratesi,^{1,2} in Italy, to explain peripheral ischemia.

However, for us it is an important entity in the absence of arterial occlusion, and encompasses some venous and lymphatic disorders, also including cases of cancer.

These documented vascular disorders, described by us as hyperstomy syndrome, must be considered in the treatment of some complicated lymphatic diseases. In these patients, the lymphatic dysfunction was aggravated by an arteriolo-venular communication. These cases were successfully treated only in conjunction with ligation of the hyperstomic abnormal feeding of the arteriolar-venular branches. This technique was emphatically presented by Mayall¹⁵ at the Madrid Congress of Lymphology, where Campisi²³⁻²⁶, from Genova, presented his excellent paper about Angiodysplasies, peripheral lymphedemas and tumorigenous syndromes.

We believe that the consensus on lymphedema must pay more attention to these problems. They found a much less invasive solution, with skeletizing and correction by ligation of the abnormal arterial branches with hyperstomy, so that dysregulated hiperdynamic blood flow is easily treated.

There was the possibility, for some patients, to resume an almost normal life, and they have been active, without any kind of medical control for 10 to 40 years.

Patients with hyperstomy syndrome will be very lucky when physicians remember this disorder in special and difficult secondary cases. The true significance of these vascular disorders or dyskinesias needs further investigation to allow a better consensus on the lymphatic dynamic in primary, and especially in the frequent secondary lymphedema troubles, avoiding the indication for amputation, as has been done already.

Finally, we hope that the surgeons interested in developing microsurgery techniques of lymphatic diseases, like Campisi²³⁻²⁶ and Baumeister,²⁷ will be able to achieve better results for the near future. In 1995, during the 15th International Congress of Lymphology, when speaking about lymphedemas, we emphasized that they must be studied not only as lymphatic disorders, but also as venous-arterial oncologic disorders, stressing the value and need for arteriography to clear some issues concerning angiodysplasia, and for phlebography of the deep venous circulation.

We do not have the right to tell the patient or his family that there is no treatment for the lymphedema. It is better to give the patient a word of hope.

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