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Hyperstomy Syndrome – A New Approach for the Treatment of Lymphedema of the Lower Legs

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Summary

To improve the results after surgical treatment of the lower legs' lymphedema, a new approach was developed by surgical ligation of the abnormal arteriolovenular shuntings. (Hyperstomy Syndrome). These findings are very common in congenital and post-phlebitic lymphedemas. The abnormal branchings are easily localized by serial arteriography. The excision techniques like the Charles' operation, presents after this new method much better esthetic appearance with out formation of hyperkeratosis or verrucous warts commonly found after these operations.

The treatment of most cases of lower leg lymphedema continues to be a challenge for the lymphologist.

Since 1960 we have been interested in the hyperstomy syndrome described by *Pratesi* and *Malan* in Italy, and have considered the possibility to the routine methods of therapy.

Initially the Hyperstomy Syndrome has been presented as an arteriolovascular abnormality that simulates an ischemic arterial disease, without showing obvious signs of occlusive disease. Following these observations, similar radiographic findings were noted in some severe cases of Post-Phlebitic Syndrome with ulcerations and lymphedema. Due to the successful results we obtained with both medical and surgical treatment of patients with marked post-phlebitic syndrome complicated by lymphedema, we became interested in routinely studying all cases of lower leg lymphedema while carefully reviewing arteriographic studies of the lower leg in light of this new approach. Today we are convinced that we must pay more attention to the influence of the hyperstomy syndrome as an etiopathogenetic factor in lymphedema, because of the resulting venous capillary and lymphatic stasis, increased by the presence of abnormal shunting between the arteriolo-venular circulation, and producing an untimely diversion of the arterial blood through the arteriolo-venular shunts. It is important to remember that the standard X-ray tube is only able to differentiate the small vessels larger than 0.1 mm. Therefore our serial angiographic studies cannot display the small shunting of the capillary bed whose size ranges between 30 and 60 micra. Only the functional disturbances resulting from an abnormal capillary circulation could be well demonstrated.

Two clinical patterns are well known. The first, as in most cases of congenital idiopathic lymphedemas, is of unknown etiology. The second appears in conjunction with some organic diseases or functional syndromes. These include post-phlebitic syndrome, trauma to the legs, and prolonged use of contraceptives. Also included are blockages of lymphatic flow, such as in chronic obstructive lymphangitis with blocking occurring at the level of the lymph nodes following repeated outbreaks of erisypela and other types of infection in the extremity. The hormonal syndrome has been well demonstrated by *Amir-Jahed* to result from abnormalities of the regulatory mechanisms of the circulation that increase the blood volume and the venous pressure in the extremity.

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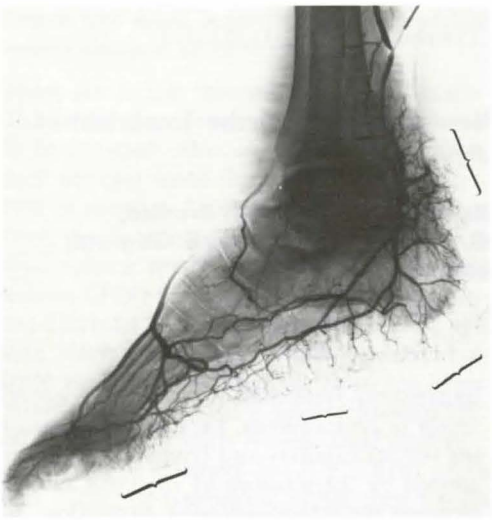

Fig. 1a Lymphedema of the foot and ankle. Arteriography through the posterior tibial artery demonstrating abnormal and excessive hyperstomy branching in the ankle and foot sole at first second

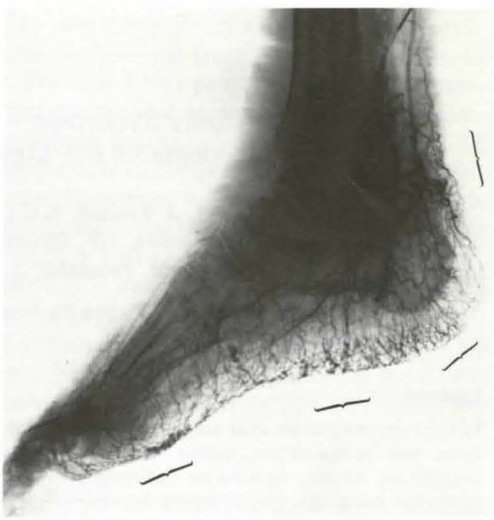

Fig. 1b At third second there is already intense and simultaneous filling of arteries and veins with a diffuse smudgy appearance

Symptomatology

During the clinical examination it is important to note the disparity between subjective and objective symptoms or signs. It is more common to find subjective complaints than objective findings. The symptoms of pain are similar to the arterial claudication, but they never are completely relieved when at rest, except after elevation of the leg and light massage of the calf. Most of these patients have an increased sensation of heaviness while walking or standing. Increased skin temperature around the superficial veins is a very common finding in post-phlebitic lymphedema, but it is difficult to evaluate because of the common incidence of cellulitis or chronic erysipela in these patients. In such cases it is important to study the oxymetry which demonstrates different degrees of arterialization of the superficial venous blood, close to the areas of the hyperstomy shunting.

Peripheral pulsations – when there is not too much edema, one may observe a difference in the amplitude of peripheral pulsations in the presence of a steal syndrome of the celf. This involves excess flow to the regional veins which slows the rate of flow to the ankle and foot. This phenomenon is very well displayed under fluoroscopy during arteriography when monitored by an image intesifier, which shows a delay in the filling of the arterial trunk in the presence of increased hyperstomy abnormal branching.

The local signs of chronic venous insufficiency and lymphostasis are not different in cases of hyperstomy syndrome, but are aggravated by its presence. The arteriolo-venular shunting becomes a very important factor in explaining why local symptoms recur so easily and how difficult they are to cure unless surgical interruption of the hyperstomy is undertaken.

The superficial venous pressure of the leg shows moderate changes. To better demonstrate this finding, the use of a sphygmomanometer cuff around the thigh is recommended to induce venous stasis using the diastolic blood pressure of the patient. If there is hyperstomy in only one leg, one can observe that the elevation of the venous pressure appears much more rapidly on the abnormal side.

An oscillometric reading, using small cuffs to isolate the anterior tibial artery region from the posterior tibial and peroneal artery region, is very useful to detect the abnormal branching of the hyperstomy syndrome in the calf.

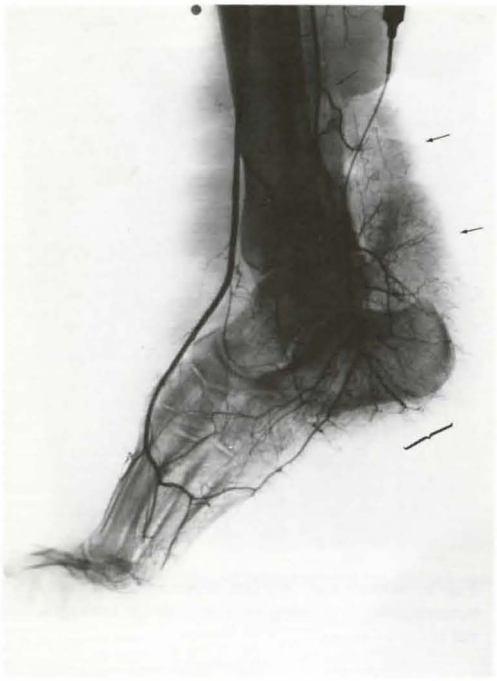

Fig. 2 Lymphedema of the lower leg. Arteriography through a teflon catheter in the posterior tibial artery showing abnormal hyperstomy branching around the ankle and heel of abnormal size, number, extension and direction

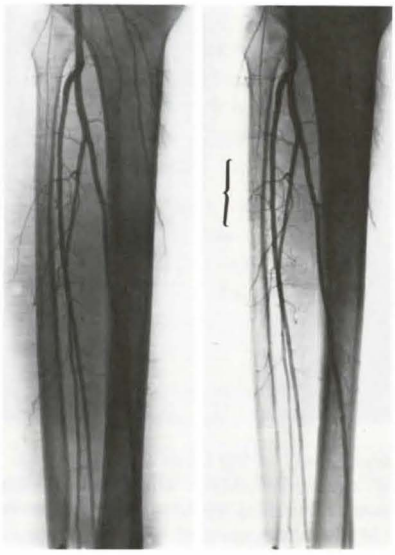

Fig. 3 Arteriography of the arteries of the calf with internal rotation of the leg, to avoid superposition of the three main trunks. There is hyperstomy abnormal branching in size and number from the fibular artery

This finding is more common in lymphedematous patients.

To confirm the diagnosis, the most accurate method is to perform serial arteriography. When it is possible to monitor the angiography under fluoroscopy while using an image intensifier, one obtains much better detail of all functional findings of the hyperstomy syndrome. The arterial deficiency and stagnation of the arterial blood flow is demonstrated just below the site of the abnormal branching and shunting to the small arteriolo-venular network.

It is advisable to obtain radiographic exposures of the leg from various angles to produce an acurate mapping of the abnormal shunting within the calf.

Thus one eliminates superimposition of the main trunks of the posterior tibial artery, fibular artery and the bones of the lower leg.

During rapid sequence arteriography, with the needle or the catheter placed as close as possible to the area of interest, the most important pathognomonic findings are:

a) Simultaneous filling of the arteries and veins with the contrast material, showing a premature venous backflow through the superficial and deep veins during the first seconds of the arteriography.

b) A blurry appearance of the muscle mass of the leg around the arteriolar branches in the areas corresponding to the main clinical symptoms and the thermometric and oscillometric findings during the second arteriocapillary phase.

c) Abnormal size, number, extension and direction of the arteriolar branches.

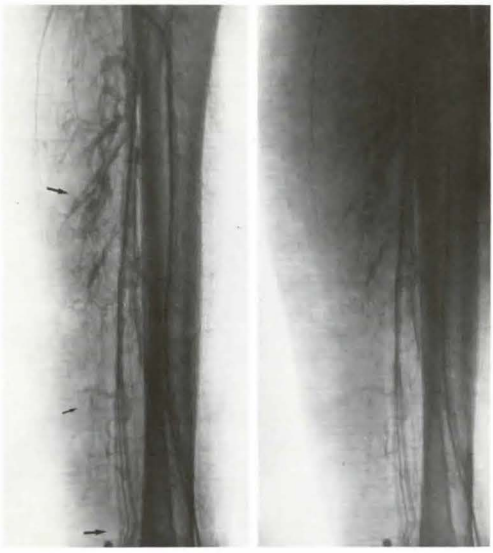

Fig. 4 Lymphedema of the lower leg. Arteriography through the posterior tibial artery in the ankle shows simultaneous filling of the arteries and veins, inclusively in the soleus varicose veins

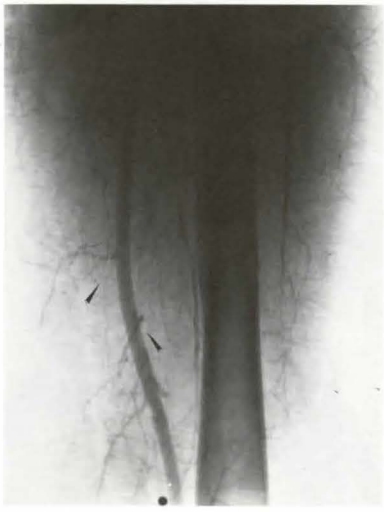

Fig. 5 Superficial femoral arteriography presenting hyperstomy branching at the level of Hunter's channel

d) Incomplete or delayed filling of the distal arteries of the leg due to deviation of the blood through the collaterals, most commonly localized in the ankle, calf and mid-thigh resulting in insufficient bloodflow through the more distal capillary network.

Differential Diagnosis

In some patients with congenital or acquired lymphedema, it is important to establish a differential diagnosis between the hyperstomy signs and symptoms and the following diseases.

a) *Congenital arteriovenous fistulas:*

They are commonly apparent since birth or puberty, and are evidenced by strawberry marks, angiomata on the legs and arms, giant varicose veins with abnormal direction, and also by an increased size and length of the extremity. The arteriography also shows abnormally large branches of the main arteries of the extremity which end in the angiomatous masses of varicose veins with a very rapid and premature backflow. It is important to establish a differential diagnosis by means of arteriography: a) the fading of the contrast material is more intense distal to the congenital fistula in the arterial phase; while in the hyperstomy syndrome the distal filling of the arteries is delayed but normal. b) The spotty appearance in the congenital fistula area is much more intense than the smudged shape of the hyperstomy branching. c) The massive enlargement of the satellite veins around the hyperstomy branches is absent, but the veins are of normal configuration and there may be slight phlebectasis or varicose veins in the more advanced cases. d) arterio-venous fistulas are commonly diffuse in the limbs, whereas hyperstomy branches are more localized. e) bone lesions are rare in hyperstomy, but commonly observed as osteolytic arterio-venous fistulas. f) In A-V shunting, the arteriolar branching is more convoluted and plexiform in appearance having a direct communication with the great venous trunk with early backflow. It is not rare to observe aneurysmal swelling of some of the anastomotic channels. In the presence of a wide lumen, heart failure and local signs like a continuous thrill and a machine-like bruit louder during the systolic phase, are frequently observed. These clinical signs are never observed in hyperstomy syndrome.

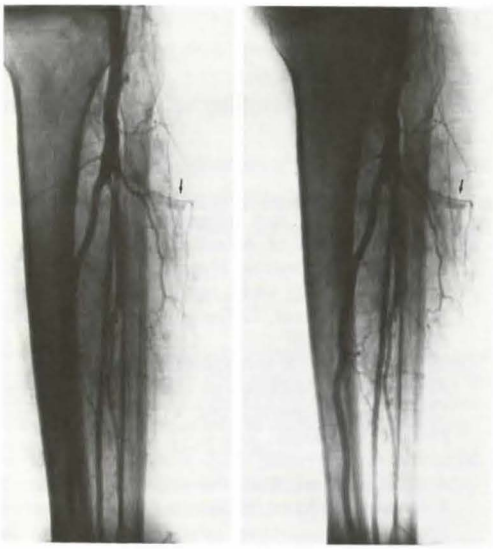

Fig. 6 The arrow shows hyperstomy branching on the fibular artery

b) *Soft tissue tumors*

Some tumors of the soft tissues such as hemangio-sarcomas of the thigh or calf, and *Marjolin's* ulcerations, complicating post-phlebitic syndromes in the early stage, present with a slight increase in the soft tissues with localized pain when exercising, local hyperthermy, and hyperoscillometric readings which constitute an indication for arteriography. Arteriography reveals anarchy of the vascular tree very similar to the most marked cases of hyperstomy. As far as we are concerned, these findings constitute a good indication for a tissue biopsy with the final diagnosis being made by histopathological assessment.

Treatment

The conservative treatment of the hyperstomy syndrome is satisfactory only in the early stage of functional and primary cases, when the functional disturbances are still reversible. For this reason, we begin the treatment with a clinical trial of non-surgical methods, including the use of hydrogenated ergotoxine alkaloids, associated with raubasine and high pressure elastic stockings. Unfortunately, when the hyperstomy syndrome lymphedema is secondary to organic pathology, this conservative treatment alone is insufficient.

At this point, it is important to remember some contra-indications. One must emphasize that, in the sexually active woman, the use of contraceptives must be forbidden because their hormonal contents represent a potential danger of worsening the symptomatology due to an increase in venous stasis, which is also a well-known pathogenic factor of hyperstomy.

Another important contra-indication is sympathectomy, because of the risk of increasing the opening of the arteriolo-venous shunting, which potentiates the borrowing-lending hemodynamic phenomenon, well-known as hemometakinesis.

Using the arteriographic mapping in the area of the lymphedema as a guide, it is very easy for the surgeon to perform the operation, which has completely modified the prognosis for many severe cases of giant secondary lymphedemas. The surgical treatment can be performed as sole procedure or as a complement to the other known and well accepted methods of surgical treatment of the severe cases of lymphedemas. The abnormal arterial branching must be ligated as close as possible to the main arterial trunk which is producing local signs such as hyperoscillometry or increase in O_2 concentration in the regional superficial veins.

This can be easily accomplished during *Charles'* procedure. It is not advisable to ligate the abnormal branching within the muscle masses, because of the risk of disabling sequelae. Presently, we have 18 years of experience with this technique without having observed any major complication. There were only small localized hematomas and transient neuritic pains noted.

Through this additional procedure during *Charles'* operation, the final esthetic appearance was much better with excellent reduction of the hardening of the skin of the extremity, which became smooth and free of acanthosis or hyperkeratosis. Before the use of hyperstomy ligation of abnormal branching in severe cases of lymphedema, it was quite common to observe an intense hypovolemic

shock during the major operation. The last three cases of *Charles*'s operation made simultaneously with the hyperstomy operation never presented hypovolemic shock and there was no need to correct the anemia and hypoproteinemia, because the interruption of the abnormal shunting produces a true plasma and blood reinfusion within the extremity, due to the pronounced shrinking of the elephantiasic tissues.

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