Heinz Lippmann Disease

To the Editor:

Chronic venous insufficiency has been defined as a pathologic condition involving both the skin and subcutaneous tissues of the lower extremities because of prolonged stasis of venous blood in the superficial veins of the leg. Although some controversy exists as to the pathophysiology of chronic venous insufficiency, most authors agree that ambulatory venous hypertension is the final common pathway leading to ulceration of the skin. Pitting edema of the lower extremities that develops during the latter part of the day, which is relieved by elevation of the affected limb, is often one of the first manifestations of chronic venous insufficiency. Prolonged edema eventually leads to stasis dermatitis of the leg and ankle and is characterized by purpura and hyperpigmentation. Induration, caused by fibrosis of the subcutaneous and dermal layers of the skin, and ossification within the subcutaneous layer, constitute postphlebitic syndrome and may represent a significant disability to the patient. Ulceration of the skin is usually the end result of long-standing venous insufficiency and occurs most commonly in the region of the ankle just proximal to the medial malleoli in the so-called gaiter area. These ulcerations can become secondarily infected and often involve extensive areas of skin.

In addition to the skin changes associated with chronic venous insufficiency, certain radiologic features were also described by Lippman in 1957, which includes periosteal new bone formation and subcutaneous ossification in association with recurrent ulcerations and chronic venous insufficiency of the lower extremities. Other authors have described similar findings. The following case report is an example of Heinz Lippmann disease: a syndrome of subcutaneous ossification, periosteal new bone formation, and recurrent ulcerations associated with chronic venous insufficiency.

Case Report

A 53-year-old male presented to the Pennsylvania College of Podiatric Medicine’s Foot and Ankle Institute with a chief concern of venous stasis ulcers involving both legs. The patient’s medical history was significant for a 30-year history of bilateral leg ulcers. Autogenous skin grafting was tried on two separate occasions several years ago without success. Despite the fact that the patient had last seen a medical professional for local wound care 10 months ago, all ulcers appeared superficially clean without any clinical signs of infection.

Skin Changes

The physical examination revealed two large, irregularly shaped, nonpainful ulcers involving the medial aspect of both ankles and distal one third of both legs. A smaller, nonpainful ulcer was located over the lateral malleolus of the left leg. All ulcers had a punched-out appearance with well defined borders, predominantly granular base, and moderate amount of serous drainage. The skin surrounding the ulcers revealed significant stasis dermatitis, edema, erythema, and areas of hypopigmentation and hyperpigmentation (Figs. 1 and 2). Palpation of the surrounding skin revealed an extremely hard bone-like texture and raised the suspicion of soft tissue calcification. There were no clinical signs of infection noted at the time of initial presentation. Because pedal pulses could not be appreciated, noninvasive vascular assessment was conducted to rule out any arterial component to the ulcerations.

Vascular Assessment

The results of noninvasive Doppler arterial studies of the lower extremities revealed patent popliteal, posterior tibial, and dorsalis pedis arteries bilaterally with triphasic signals. The ankle to arm ischemic indices were found to be 1.59. Cutaneous perfusion to the digits was normal despite spasticity of the microcirculation of the digits at the time of examination. Valvular incompetency revealed by noninvasive Doppler venous studies was noted in the posterior tibial, long saphenous, and superficial communicating veins of both lower extremities indicating significant vascular involvement.

Radiographic Findings

Radiographic findings at initial presentation included bilateral cutaneous calcifications and irregular periosteal bone production involving the dis-
increased and decreased bone density could be seen throughout.

Treatment

Prior treatment before seeking care at this clinic included two failed attempts at split-thickness skin grafting, nylon compression stockings, and daily local wound care. For the past year, weekly appli-
cations of boot dressings combined with periodic mechanical debridement and cleansing with a soft sponge and wound cleanser have also failed to resolve the ulcerations completely, although some improvement was noted in the ulceration involving the left lateral malleolar region. Given the extremely hard texture of the surrounding skin and presence of subcutaneous bone, it is unlikely that another attempt at skin grafting would prove beneficial. Other modalities, such as intermittent pneumatic compression pumps that help extract fluid from the tissues of the lower extremities, are also unlikely to result in significant improvement. Local wound care and avoidance of lower extremity dependency possibly appear to be the most realistic treatment for the treatment of Heinz Lippmann disease.

Discussion

It should be stressed that although there are some unique radiographic findings in this case, the diagnosis of Heinz Lippmann insufficiency was made clinically by palpating the rock-hard indurated areas of skin surrounding the ulcerations. Lippmann confirmed the presence of cancellous bone within the subcutaneous tissues by histopathologic examination in four cases. Given the high incidence of venous pathology involving the lower extremities in the general population, Heinz Lippmann insufficiency may be an underrecognized and often misdiagnosed clinical entity.

Any history of a chronic, nonhealing wound with radiographic evidence of new bone formation should be investigated and entities such as myositis ossificans, arterial calcifications and phleboliths, hypertrophic osteoarthropathy, neurogenic osteoarthropathy, Gaucher’s disease, hyperparathyroidism, Paget’s disease, and neoplasms of bone and skin should be ruled out. Woodward and Andreini also described a syndrome of pain, swelling, skin changes, and periosteal new bone formation involving the lower extremities in seven patients with polyarteritis nodosa in the lower extremities. Polyarteritis nodosa could easily be ruled out in this case because of the nonpainful nature of the ulcers. No evidence of a systemic disturbance of calcium metabolism was found in this patient and was excluded in all cases presented by Lippmann.

All 83 cases described by Lippmann were females past the age of menopause. All of the males who were treated by Lippmann and suspected of having subcutaneous ossification were found by biopsy and conventional radiography to exhibit subcutaneous fibrosis only. This information raises the question whether changes in hormone levels and activity may play a role in the formation of extraskeletal bone; however, the case presented here and one other case encountered by the authors both involved males. To the authors’ knowledge, this is the only reported case of Heinz Lippmann disease in males.
Malignancy and Pedal Gangrene

To the Editor:

Pedal gangrene and malignancy can occur in rare situations where vasculitis, hyperviscosity, and hypercoagulability lead to arterial occlusion. Gangrene may present as the only manifestation of the malignancy.

Ischemia of the digits as a manifestation of a neoplasm has been reported in the literature as a rare occurrence usually involving the fingers in a bilateral symmetrical distribution. The onset is usually abrupt in middle-aged women. The symptoms begin as intermittent ischemia, progressing to chronic ischemia, ulceration, and gangrene of the digit.¹

Early changes of ischemia mimic Raynaud’s syndrome; however, Raynaud’s does not end with gangrenous changes to the digits. Raynaud’s syndrome refers to the episodic digital ischemia in response to cold or emotional stimuli. Many patients with gangrene and malignancy have a prior history of ischemia preceding the gangrene by several months.²

Some of the earliest reports of malignancy and thrombosis involve venous thrombosis, which was recorded by Trousseau in 1865.³ Sack et al³ confirmed the aberration in coagulation in patients with a neoplasm in their review of 182 cases. Analysis of blood chemistry showed hypofibrinogenemia, thrombocytopenia, prolonged prothrombin time, increased fibrinogen-fibrin degradation products, decreased levels of factors V and VIII, cryofibrinogemia, and microangiopathic hemolytic anemia.³

Clinically, patients may show thrombosis prior to the malignancy. Often, the thrombosis of the venous system may proceed other symptomatology of the malignancy by weeks or months.⁴

Case Report

A 64-year-old female admitted for unstable angina and acute cholecystitis presented with ischemic changes to the toes of both feet symmetrical, which gradually turned into digital gangrene (Figs. 1 and 2). The patient complained of severe, unremitting pain at the distal aspect of her feet. The patient also complained of pain in the lower back and abdomen. The results of the chest x-ray revealed a paramediastinal lung mass that looked suspicious during a fine needle biopsy for malignancy. Laboratory abnormalities included the following: fibrinogen (500), FSP > (40), white blood count (27), pt (14.1-16.9), and platelet count (53).

On the 16th day of hospitalization, the patient suffered increased mental status changes along with gangrene of the fingertips. The patient died as a result of renal failure on the 20th day of hospitalization. The results of the autopsy were as follows: 1) acute myocardial infarct 7 to 10 days old, left ventricle wall with thrombotic occlusion of the left coronary artery branch supplying the lateral wall
and an older 14-day infarction in the ventricular septum; 2) adenocarcinoma of the lung with lymphangitic spread and metastatic lesion in the lumbar spine, parietopleural and bilateral hilar lymph nodes; 4) multiple 0.5 to 1.0-cm splenic infarcts; 5) bilateral dry gangrene of toes and proximal portion of feet; and 6) acute renal failure with renal parenchymal scar. The cause of death was an acute myocardial infarction with adenocarcinoma of the lung and disseminated intravascular coagulation as precipitating factors.

Similar aberrations of thrombosis and malignancy have been reported in the literature; however, these must be differentiated from other possible causes of thrombosis through history and physical and appropriate laboratory work-up.

Discussion

The patient showed disseminated intravascular coagulation but also had evidence of vasospasm preceding the gangrene. The autopsy results showed metastasis of the adenocarcinoma of the lung to the lumbar spine. While no mention of involvement of the sympathetic ganglion is noted, several articles have mentioned a link between vasospasm and neoplasm of the sympathetic ganglion.\textsuperscript{5}

Vasculitis has been implicated in digital gangrene among many autoimmune complexes with the exact mechanism unknown. A probable theory suggests antigen antibody complexes that bind to artery walls and cause the coagulation cascade to be triggered.\textsuperscript{6}

Many patients with digital gangrene also have hypercoagulability. Patients have laboratory values with short bleeding time, decreased partial prothrombin time, and an increased intolerance to heparin. In a study by Miller et al\textsuperscript{7}, of 50 patients with malignancy, 16% had thrombotic disorder.

Increased viscosity secondary to protein production from the tumor has been noted with IgG serum level increased with intensity at ischemia.\textsuperscript{8} Patients with chronic granulocytic leukemia often present with white blood cell counts greater than 150,000/cu mm and anemia, which often leads to ischemia.\textsuperscript{9}

Treatment

The success of treatment is dependent on the improvement of circulation of the affected limb before the ischemia results in gangrene. Patients will show improvement with resection of the neoplasm or chemotherapy, which has been shown to have reversal.\textsuperscript{1, 6}

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Lisfranc Fracture-dislocation in a Windsurfer

To the Editor:

Lisfranc, a pupil of Dupuytren, described and performed amputations through the tarsometatarsal joints of soldiers in Napoleon’s army with gangrenous injuries.1 Lisfranc fracture-dislocations are rare, accounting for only 0.2% of all fracture-dislocations reported.2 However, as much as 20% of Lisfranc fracture-dislocations are initially undiagnosed or misdiagnosed, especially in the patient possessing other more life-threatening and distracting injuries.3

Because of the unique architecture of Lisfranc’s joint and because the variety and combination of forces that can be applied to the forefoot are numerous, many types and patterns of fracture-dislocations can occur. Additionally, the severity of injury can range anywhere from gross subluxation with multiple comminuted fractures to subtle malalignment.2

The classification system of Myerson et al3 is the most widely accepted and is as follows: 1) type A or total incongruity of Lisfranc’s joint, displacement of the entire forefoot, either medially or laterally; 2) type B1 or partial medial incongruity, the first metatarsal is displaced medially; 3) type B2 or partial lateral incongruity, one or more of the lesser metatarsals are displaced laterally; 4) type C1 or divergent pattern, the first metatarsal is displaced medially and any combination of the lesser metatarsals is displaced laterally with partial incongruity; and 5) type C2, a divergent pattern of displacement with total incongruity.

The medical literature divides Lisfranc fracture-dislocations into those caused by direct and indirect trauma.4, 5 Direct trauma to Lisfranc’s joint results in no specific, discernible pattern of dislocation and is frequently associated with complex comminution and open fractures.5

Indirect trauma is most commonly caused by axial loading of the plantarflexed foot or rotation of the leg and rearfoot as a unit on a stabilized forefoot.3, 4 For example, prior to a motor vehicle accident, the occupant’s feet are commonly plantarflexed and braced against the automobile’s floorboard. During a collision, the occupant is thrust forward, causing the metatarsal heads to bear the total body weight. The force placed on the metatarsal heads is transmitted proximally, hyperplantarflexing the foot at the site of least resistance, the dorsal aspect of Lisfranc’s joint.5 Under such tension, the weaker dorsal ligamentous and capsular structures will rupture and subsequent dorsal dislocation of the metatarsals will occur.5 If there is an inversion or eversion component to the force, the metatarsals will dislocate dorsolaterally or dorsomedially.

Rotation of the leg and rearfoot as a unit on a stabilized forefoot is another common mechanism of injury that can cause Lisfranc fracture-dislocations.3, 4, 6 Classic descriptions of Lisfranc fracture-dislocations were derived from equestrian accidents in which one of the rider’s feet would become entrapped in the stirrup and forcefully plantarflexed and abducted as the rider was thrown from the saddle.3, 4

Currently, however, this mechanism of injury more commonly results from motorcycle accidents than equestrian accidents, although any situation that immobilizes the forefoot and allows rotation of the leg and rearfoot as a unit can potentially traumatize Lisfranc’s joint. For example, while someone is windsurfing, his or her forefoot is immobilized within a foot strap that extends from the tip of the digits to the midfoot. Similar to the sequence of events that occur in equestrian or motorcycle accidents, it is probable that when an individual is thrown from a windsurfing board, the trapped forefoot would become forcefully plantarflexed and abducted at Lisfranc’s joint (Fig. 1).3, 4

The multitude of violent rotational forces placed on Lisfranc’s joint in such accidents could produce many different patterns of fracture-dislocations. Only one previous description of a Lisfranc fracture-dislocation occurring in a windsurfer exists in the medical literature.7
Case Report

On January 31, 1996, a 23-year-old male presented to the Pacific Coast Hospital surgical clinic 24 hr after an injury to his left foot sustained while windsurfing. The patient reported that his left foot had not released during a fall from his windsurfing board. The patient felt intense pain in the forefoot area of his left foot. After returning to shore, the patient was assisted home by friends, where he kept his left foot elevated and iced until presenting to the clinic. The patient has been able to apply only minimal pressure to his left foot while attempting to stand or walk since the injury.

The patient denied any prior history of injury to either the left or right foot. The patient's medical history, social history, and review of systems were unremarkable and noncontributory. Physical examination of the left lower extremity revealed mild swelling and ecchymosis over the midfoot at the level of Lisfranc's joint. Musculoskeletal examination of the left lower extremity revealed maximum tenderness with any attempt of motion of the lesser metatarsals. No significant structural abnormalities were noted on gross appearance, and the remainder of the physical examination was unremarkable. Bilateral dorsoplantar and lateral radiographs were taken that revealed a Lisfranc's dislocation without fracture of the lesser metatarsals, and some dorsal displacement of the second and third metatarsals (Figs. 2 and 3).

Closed reduction was performed under local anesthesia with intravenous sedation and satisfactory anatomical alignment was achieved. Multiple crossed Kirschner wire fixation was used to stabilize the reduction and the patient was placed in a nonweightbearing below the knee cast. Immediate postreduction dorsoplantar and lateral radiographs confirmed satisfactory anatomical alignment of Lisfranc's joint in both the transverse and sagittal planes. The Kirschner wires were removed 3 weeks after the injury.
after reduction, and the patient converted to a below the knee walking cast for an additional 3 weeks. Dorsoplantar and lateral radiographs taken 6 weeks after reduction revealed continued anatomical alignment of Lisfranc’s joint in both the transverse and sagittal planes. The patient has subsequently returned to work and other activity, including windsurfing.

Discussion

A number of treatment options have been advocated ranging from gentle manipulation with immobilization to open reduction with internal fixation to primary arthrodesis. Of utmost importance when treating Lisfranc fracture-dislocations is early recognition of the injury and restoration of an anatomically correct joint. Prompt diagnosis, proper understanding of the extent of injury, early treatment, and anatomically correct reduction will help to lessen the chance of post-traumatic degenerative arthritis, although these actions will not guarantee a properly functioning and painless joint.

Individuals in whom the injury was initially misdiagnosed or the extent of the injury under estimated often have poor results and sequelae such as recalcitrant pain, swelling, stiffness of the joint, and abnormal forefoot to rearfoot deformities.

With the recent surge in recreational sports that require that the forefoot of one limb be strapped down and immobilized, such as windsurfing and snowboarding, injuries to Lisfranc’s joint may become more frequent as these sports increase in popularity. Regardless of the mechanism of injury or classification system used, each potential Lisfranc fracture-dislocation should be given individual consideration. The severity of injury, presence of soft tissue interposition, interarticular fragments, and the ease of reduction all play a role in the initial diagnosis, form of treatment rendered, and long-term prognosis.

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