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TROPICAL IDIOPATHIC LOWER LIMB GANGRENE: CASE REPORT

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### SUMMARY

**Tropical idiopathic lower limb gangrene is a rare disease. It was first described by Gelfand amongst the indigenous inhabitants of present day Zimbabwe. It is a bilateral and simultaneous gangrene of both lower extremities due to no obvious cause and usually seen in men during the second and fourth decade of life. The onset is always sudden and the first sign is oedema of both feet accompanied by pain. The patients are usually people who have been previously healthy. This is a report of a clinical variant of the disease.**

### INTRODUCTION

Gelfand(1) was the first to describe this rare disease, idiopathic lower limb gangrene in the tropics. This is a bilateral and simultaneous gangrene of both lower extremities in mostly males in their second and fourth decade of life. The aetiology is usually not known. The onset is always sudden and the first sign is oedema of both feet accompanied by pain. It may be associated with an infectious condition. Below is a report of a clinical variant of the disease.

### CASE REPORT

A 15-year old boy was referred to the department with a history of sudden onset of fever, headache and vomiting three weeks earlier. The headache was severe, generalised, throbbing and persistent. Two days later after being treated for malaria, with only chloroquine injections, these symptoms subsided but the upper limbs became painful and swollen. A week later after the pains in the upper limbs had subsided the lower limbs got painful. These pains were also accompanied by swelling and formation of blisters. The boy looked ill and emaciated. His conjunctivae were pale. There was no jaundice. Examination of the central nervous system did not reveal any pathology. The lungs were clear. The heart was not enlarged. The first and second heart sounds were heard. There were no murmurs. The blood pressure was 120/70mmHg. His abdomen was not distended. There was no organomegaly. There were skin lesions on the limbs which were indicative of resolving blisters. These lesions were on the distal 2/3 of the anterior aspects of both forearms and the distal 1/2 of the anterior aspects of both legs. There were also lesions on the posterior aspects of both thighs extending slightly to the buttocks. In the lower limbs there was a uniform change of skin colour in the parts that had no lesions. From about 6 cm below both tibial tuberosities, apart from the medial aspect of both legs and areas about 2 cm in diameter on the dorsum of both feet, the skin was bluish black in colour. All the toes of the right foot except the middle toe were gangrenous. All the toes of the left foot looked healthy. The pulsation of both femoral and popliteal arteries were felt but that of both dorsalis pedis arteries was not felt. The father inquired of the prognosis and was informed his son may lose both legs. Hoping to get help from unorthodox medicine he signed against medical advice and removed his son from the hospital.

Figure 1



Figure 2



Three months later, unable to get help from traditional medicine he returned with the child to the hospital. The child did not look different. Nothing significant was found on examination of the central nervous system. The chest was still clinically clear. The blood pressure was 125/70mmHg. There was no pathology in the abdomen. The legs were oedematous. This oedema extended to just below the knees. There were deep subcutaneous ulcers in the bluish black parts of the legs. All the toes were still healthy

except the gangrenous toes of the right foot which had fallen off. The pulsation of both dorsalis pedis arteries was still not felt.

Blood smear for plasmodia was negative. Packed cell volume, white blood cell, platelet and erythrocyte sedimentation rate were 16%,  $8.4 \times 10^9/l$ ,  $300 \times 10^9/l$  and 22mm fall for the first one hour respectively. The haemoglobin electrophoretic pattern was AS. There was neither glucose nor albumin in his urine. The patient had bilateral above knee amputation. The stumps healed well and ten days later the patient was discharged.

## DISCUSSION

Ever since Gelfand(1) first described this disease amongst the inhabitants of Zimbabwe, more cases have been reported mostly in East Africa.

The cases Gelfand reported were due to vascular occlusion. His explanation was that malnutrition with the resulting hypoproteinaemia and vitamin deficiencies increased capillary wall permeability. This caused oedema which might have interfered with the circulation. In one of his cases, after administration of thiamine the oedema subsided but there was already gangrene. Vitamin deficiencies can also cause vascular wall thickening with narrowing and obliteration of the tissues of the small arteries of the feet and legs. He also associated the disease with malaria.

Anne Lloyd *et al*(2) reported a series of peripheral gangrene in infancy and childhood. Because of response to treatment with azothioprine and hyperbaric oxygen, it was presumed that the gangrene was due to polyarteritis nodosa. Histological analyses confirmed the diagnosis in two of their four cases. Koten reported the disease in four children(3). In each of the cases there was an infectious condition (pneumonia or gastroenteritis) before presentation. Histological analyses showed vascular changes in two and proliferative endovasculitis with secondary thrombosis in the rest. Lowenthal and Okojie(4) reported paediatric cases of the disease from Zambia and Nigeria but no cause was incriminated.

The equilibrium between coagulation and fibrinolysis *in vivo* maintains a patent and intact vascular wall. Turpie *et al*(5) were able to show impaired fibrinolysis in three paediatric cases of the disease. There were increased levels of fibrinolytic inhibitors despite normal coagulation. This depressed significantly spontaneous plasma fibrinolytic activity. The same imbalance between coagulation and fibrinolysis was seen in adult cases of the disease by Barr *et al*(6). In these patients blood investigations revealed: raised levels of plasma fibrinogen, factors V and VIII, clotting and thrombin times were shortened. The underlying cause of this unbalance is still unknown.

Three cases of idiopathic gangrene were reported by Bhana and Baddley(7). Two of the cases were women. Arteriography of two of the cases showed that the involved vessels were not uniformly narrowed and collateral circulation developed when metatarsal or proximal digital

arteries were occluded. The third case was a pregnant woman who had taken a herbal preparation throughout the gestational period to facilitate labour. It was a case of classical ergotism.

Steiner and Hutt properly documented microscopic vascular changes in this disease(8). In 14 cases, the vascular changes were similar. In the arteries, there was proliferation of the intima by loose tissue composed of basophilic mucoid cystic material which contains fibroblast and smooth muscle cells. This thickening may be diffuse or involve only a segment of the circumference. In some of the cases, the mucoid cysts are also present in the media and adventitia. There was also internal elastic lamina degeneration associated with the intimal proliferation. In cases of longer duration, there was increase in collagen and hyaline tissue in all the layers of the arterial wall with occasional foci of calcification. Bigger veins also showed similar diffuse or cushion-like intimal proliferation and mucoid swelling of the media. There were deposition of haemosiderin with the intimal mucoid cystic material in two of the cases. This shows that an arterial thrombus may undergo mucoid transformation. In one of the cases there was thrombosis in an artery.

In this case the upper limbs were swollen, painful and had blisters first. Although all these resolved the same pathological processes occurred in the lower limbs and progressed into deep, extensive, subcutaneous ulcers and oedema of both legs with eventual spontaneous amputation of four toes from the right foot. There were patches of healthy skin in between the ulcers. The pattern of the healthy skin was identical. This clinical manifestation was peculiar. Necrotizing fasciitis can also manifest in a leg the same way but never with associated gangrene of any toe and not bilateral(10). Despite lack of arteriography, haematological and histological analyses, this case was diagnosed as a clinical variant of tropical idiopathic lower limb gangrene on the basis of clinical data.

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