

A review of diagnosis and modes of presentation of tropical idiopathic lower limb gangrene

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Abstract

Background: Tropical idiopathic lower limb gangrene (TILLG) is also known as Symmetrical gangrene in the African, Idiopathic gangrene in the African and Idiopathic peripheral gangrene of the tropics. The aetiopathogenesis of this clinical entity is a mystery.

Objective: To review methods of diagnosing tropical idiopathic lower limb gangrene (TILLG) and highlight its clinical variants.

Method: All Literature on idiopathic gangrene of the extremities was searched from libraries, colleagues and internet but only literature on TILLG (in Africans) from 1947 to date was scrutinised. Each case was studied to find out the basis of diagnosis.

Result: TILLG is not fully understood and not easy to recognise. Two sets of criteria are known to be helpful in establishing diagnosis. These criteria can be classified as major and minor criteria. Major criteria are those clinical data that can establish the diagnosis of TILLG. No devices are required to identify them. Minor criteria are pathological changes that are consistent with TILLG. Devices are required to identify them. Three pathomorphological types of TILLG were described in literature and are classified as types A, B and C.

Conclusions: This review is supposed to sensitise the clinician and make diagnosis easier. This will also encourage more researches. As more information becomes available, aetiopathogenesis of TILLG will be clearer and more clinical variants of the disease may be reported. This additional information will help in the prevention of gangrene, reducing the socioeconomic problems arising from amputation.

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Introduction

The first report of tropical idiopathic lower limb gangrene (TILLG) is credited to M. Gelfand¹. The features of this first case were; gangrene of unknown aetiology, which was bilateral and simultaneous. The gangrene was sudden and mostly in males who were in the second and fourth decade of life. The first sign was oedema of both feet accompanied by pain. Febrile condition was associated with TILLG. Many cases have been reported in East and Central Africa but recently two variants of the disease have been seen in Nigeria^{2,3}. Two colleagues in Nigeria discussed cases of TILLG they could not report with the author (personal communications). This suggests the incidence of TILLG could be higher than previously thought. Diagnosis of the disease has always been difficult. Below is a review of the methods of diagnosing TILLG and a highlight of its clinical variants.

Diagnosis of TILLG

TILLG has always been diagnosed on the bases of the similarity of history of the case in question and the first case reported¹. Amongst the features of the cases reviewed, the sign that was always present was dry gangrene of any of the lower limbs of sudden onset. The

index of suspicion of the physician was very instrumental in establishing the diagnosis^{1,2,3,4}.

Lower limb gangrene can result from necrotising fasciitis⁵, but there is always history of trauma and most often the gangrene was wet. Atherosclerosis can cause dry gangrene. The patient is usually elderly, above fifty years of age and histology will reveal typical arteriosclerotic changes in the form of plaques obliterating vascular lumen. In the neonate, gangrene can result from congenital syphilis. VDRL test (Venereal Diseases Research Laboratory Test) in any of the parents will be positive. This is the differential diagnosis of TILLG.

This method of diagnosis is based only on clinical data and can be called the major criteria for establishing the diagnosis of this disease. Recognition of the signs and symptoms of TILLG is within the competence of most clinicians.

There are pathological changes that have been found to be consistent with TILLG. In vivo there is equilibrium between coagulation and fibrinolysis. In TILLG there is slight increase in the level of fibrinolytic activity^{6,7}. Thus analysis of blood sample from a patient diagnosed as a case of TILLG, reveals raised levels of plasma fibrinogen, factors V and VII, shortening of clotting and thrombin times.

TILLG has a particular arteriographic pattern⁸. The lumen of the arteries of the involved lower limb is not uniformly narrowed and collateral circulation develops whenever metatarsal or proximal digital arteries were occluded. TILLG results in distinct microscopic

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vascular changes⁹. Part or the whole of the circumference of the arteries were thickened as a result of proliferation of the intima by loose tissue composed of basophilic, mucoid, cystic material whose contents are fibroblast and smooth muscle cells. These mucoid cysts are also present in the media and adventitia. In some of the cases proliferation of the intima is accompanied by elastic degeneration of the lamina. In cases that present late there are foci of calcification in all the layers of the arterial wall. There is increase of collagen and hyaline tissue in this layer. Intimal proliferation and mucoid swelling of the media are either cushion like or diffuse. Frank arterial thrombosis is rare and when present may undergo mucoid transformation. These pathohistological changes are not limited to the arteries alone. The same type of intimal proliferation and mucoid swelling of the media are also seen in bigger veins. These pathological changes can be called the minor criteria for establishing the diagnosis of TILLG. These investigations are not possible in the rural hospitals where these patients will first present.

Modes of presentation of TILLG

Different clinical variants of TILLG^{1,2,3,8} have been described in literature and aetiopathogenesis of the disease remains a mystery. Therefore the only possible classification is based on the different pathomorphological changes associated with the different clinical variants which can be grouped into three:

Type A

This is the type¹ whose first description is credited to M. Gelfand. It may be seen in younger age. This type always results to bilateral below knee amputation.

Type B

This is the gangrene of one lower extremity in a neonate³ (and below the age of one year). Blisters on the upper extremities usually precede this gangrene. Two cases have been seen. One has been reported in literature and the second was diagnosed recently in this hospital. This type requires below knee amputation of the affected limb only.

Type C

The pathomorphology varies but gangrene of the whole foot is never seen. This type presents as gangrene of the toes. There are ulcerations of the lower extremities. Blisters on the upper extremities may precede the pathology in the lower extremities. This type^{2,9} has been seen in adulthood and adolescence only. The typical arteriographic changes in TILLG are seen in these cases. Only gangrenous toes are amputated.

Conclusion

This review is supposed to sensitise and make diagnosis easier for the clinician. This will also encourage more researches. As more information becomes available, aetiopathogenesis of TILLG will be clearer and more clinical variants of the disease may be reported. This additional information will help in the prevention of gangrene, reducing the socioeconomic problems arising from amputation.

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