Calcific myonecrosis following snakebite

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Abstract

Calcific myonecrosis is a rare condition and is believed to be a late sequela of untreated compartment syndrome. Patients usually present with a progressively enlarging mass, years after the initial injury, that can be misdiagnosed as a soft tissue sarcoma. Calcific myonecrosis following snakebite is extremely rare. The anterior compartment of the leg is most frequently involved and appears to be especially vulnerable to developing this complication. Conservative management should be considered in asymptomatic patients but spontaneous soft tissue breakdown with sinus formation may develop and prolonged surveillance is advised. We report two similar cases of calcific myonecrosis of the anterior compartment of the lower leg that developed decades following snakebite.

Key words: calcific myonecrosis, compartment syndrome, soft tissue calcification, snakebite

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Introduction

Calcific myonecrosis is a rare condition and less than 50 cases have been reported in the English literature. It was first described by Gallie and Thompson in 1960 as a late sequela of compartment syndrome.[1] Although a hypothesis for the development of this complication was proposed by Janzen *et al.* in 1993 and O'Keefe *et al*. in 1995, the exact physiological mechanism is not fully understood.[2,3]

The condition is characterised by muscle necrosis of an entire osteofacial compartment with central liquefaction and peripheral calcification. The clinical and radiographic findings can be misdiagnosed as a soft tissue sarcoma.[4-6] Differential diagnosis includes synovial sarcoma and soft tissue osteosarcoma as well as benign conditions like myositis ossificans, dermatomyositis, tumoral calcinosis and calcific tenosynovitis (*Table I*).[5]

We report two similar cases of calcific myonecrosis of the anterior compartment of the lower leg following snakebites and review the available literature on this rare condition.

Table I: Differential diagnosis of calcified soft tissue lesions

Malignant conditions	Benign conditions
Synovial sarcoma	Heterotypic ossification/myositisossificans
Epithelioid sarcoma	Dystrophic soft tissue calcification
Soft tissue osteosarcoma	Dermatomyositis/polymyositis
Soft tissue mesenchymal ondrosarcoma	Tumoral calcinosis
	Diabetic myonecrosis
	Calcific tenosynovitis

Case report

Two similar cases were managed at our unit during the same year. These cases involved female patients aged 62 and 81 years respectively, who both reported a painless, progressively enlarging mass in the anterolateral aspect of

the left and right lower leg respectively. In both cases the mass was initially noted approximately one year prior to presentation. Both patients reported a snakebite to the lower leg decades earlier. The first patient sustained a snakebite at the age of 13 years (49 years earlier) while the second patient was 16 years old (65 years earlier) at the time of the injury.

Clinical examination revealed a large, painless, fluctuant mass located in the anterior compartment of the leg of both cases. One patient presented with two sinuses that developed spontaneously and drained white 'toothpaste-like' liquid. Radiographs confirmed a calcified soft tissue mass in the anterior compartment of the leg in both patients. The masses had a fusiform appearance with linear calcifications and well defined borders. (*Figures 1a and 1b*) In both cases the tibia and fibula were uninvolved. Magnetic resonance imaging (MRI) demonstrated T1 isointense and T2 hypointense fusiform lesions along the tibialis anterior muscles with a thin rim of low signal capsule and no contrast enhancement (*Figure 2*).

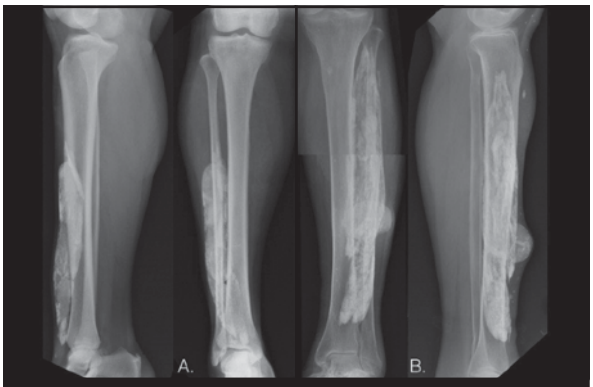

Figure 1. Antero-posterior and lateral X-ray images of both patients. Fusiform calcified lesions can be seen in the anterior compartment.

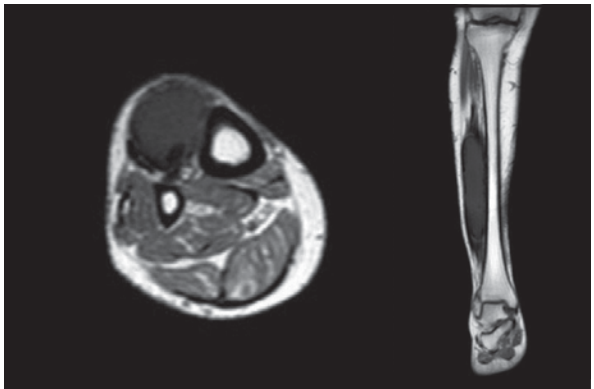

Figure 2. Axial and coronal MRI images from one of the cases. A central hypointense area is surrounded by a pseudocapsule of low signal intensity. No enhancement was noted following administration of intravenous contrast medium.

Management consisted of surgical debridement, prolonged suction drainage and a short course of prophylactic antibiotics in both cases. Macroscopic examination of the specimens revealed elongated shards of debris that corresponded to the linear calcifications identified in the radiographs (*Figure 3*). Histological examination showed lesions composed of calcific debris surrounded by fibro-connective tissue septae, and confirmed the diagnosis of tumoral calcinosis in both cases. Both patients were followed up for six months after surgery. While one patient had complete healing of the surgical incisions with no reaccumulation or septic complications, the patient that presented with draining sinuses had persistent drainage from these sinuses for two months following the surgery.

Discussion

Calcific myonecrosis is a rare condition and its pathophysiology is not well understood. The anterior compartment of the leg is most frequently affected, but other anatomical locations including the upper limbs have also been reported.[5-7] It usually follows several years after trauma and is probably related to an untreated compartment syndrome.

Compartment syndrome usually results from trauma but has been reported following snakebites.[7-15] These bites are usually from snakes that produce cytotoxic venom and most reports implicate snakes from the Viperidae family. Snake venom contains at least 26 different types of enzymes; 12 of these enzymes are common in all venoms, and the rest occur separately in certain species.[16] Up to 80–90% of Viperidae venom consists of enzymes that possess very strong proteolytic activity, while that of Elapidae, with the exception of the m'Fezi or Mozambique spitting cobra (*Naja mossambica*), have very weak proteolytic action.[17]

Vipers have long, hinged fangs that permit deep envenomation of proteases, peptid hyaluronidases and phospholipases that cause vascular endothelial damage with resultant oedema and local tissue necrosis.[7-9]

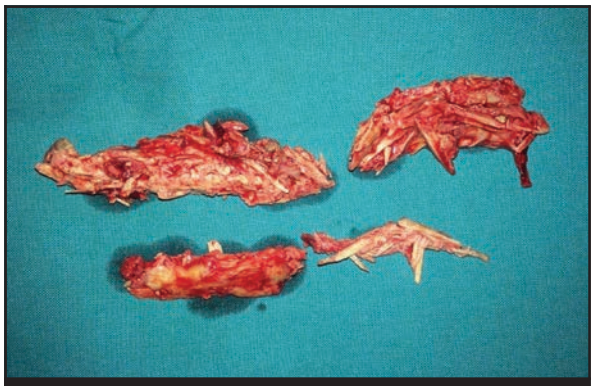

Figure 3. Macroscopic appearance of the calcified lesions excised from the anterior compartment in one of the patients. The lesions were filled with white-grey material which had a toothpaste-like consistency.

Only two reports of calcific myonecrosis following snakebites have been reported in the English literature and these cases are almost identical to the cases presented here. Yuenyongviwat *et al*. reported a case of a 66-year-old Thai woman who presented with calcific myonecrosis 52 years after being bitten by a Malayan pit viper (*Calloselasma rhodostoma*).[18]

The anterior compartment of the leg was affected and after a four-year period of conservative management, soft tissue breakdown necessitated surgical intervention in the form of mass resection and primary closure with vacuum drainage. Chun *et al*. also reported a case of calcific myonecrosis following a snakebite. Their case presented 44 years after being bitten, again involved the anterior compartment of the leg and was treated by mass resection.[19]

Although neither of our patients could recall the species of snake, a member of the Viperidae family was probably involved. Three viper species are frequently encountered in KwaZulu-Natal, including the puff adder (*Bitis arietans*), berg adder (*Bitis atropos*) and night adder (*Causus rhombeatus*).[15]

The puff adder specifically, is one of the most common venomous snakes in South Africa and is responsible for more fatal bites than any other snake. If not fatal, puff adder bites frequently result in extensive necrosis that may require amputation of the affected limb.[20]

A bite from one of these snakes is consistent with previous reports of compartment syndrome[7,13,14,21] and calcific myonecrosis[18,19] following viper bites. Interestingly, venom from the saw-scaled viper (which contains ecarin that activates prothrombin) has also been associated with renal cortical necrosis followed by calcification.[16]

Acute anterior compartment syndrome involves the isolated anterior compartment of the lower leg and usually follows trauma.[22]

Wood *et al*. reported that approximately 9% of patients with rapid progressive swelling (RPS) after serious snakebites develop compartment syndrome.[23]

In the acute setting, management involves release of this compartment through a fasciotomy. Compartment syndrome as a result of snakebite frequently involves the anterior compartment in isolation and may be the result of envenomation of this compartment and the specific anatomical characteristics of the anterior compartment of the lower leg. The sequelae of untreated compartment syndrome include rhabdomyolysis, permanent nerve damage, contracture and loss of function, infection and even amputation. Calcific myonecrosis is believed to be a rare complication of untreated compartment syndrome and follows several years to decades after the trauma.

Conservative management of calcific myonecrosis is advised in asymptomatic patients. Some authors even propose that calcific myonecrosis be considered as 'don't-touch' lesions due to the high risk of infection following surgical intervention.[24-27] A conservative approach does not, however, preclude infective complications. Affected patients may present with soft tissue breakdown as the mass enlarges and *de novo* infection has also been reported.[5] Spontaneous soft tissue breakdown with sinus formation was seen in one of our cases and in the case reported by Yuenyongviwat *et al*.[18]

Patients with calcific myonecrosis should be followed closely so that early intervention can be initiated once soft tissue breakdown occurs.

The theory that calcific myonecrosis develops after compartment syndrome suggests that a treatable cause for this complication exists. Patients who present with snakebites to the lower limb should be carefully evaluated for compartment syndrome and the appropriate treatment instituted.

Conclusion

Calcific myonecrosis is a rare complication following snakebites. The anterior compartment of the leg is most frequently involved and appears especially vulnerable to developing this complication. Conservative management should be considered in asymptomatic patients but spontaneous soft tissue breakdown with sinus formation may develop, and prolonged surveillance is advised.

Compliance with Ethics Guidelines

Written consent was obtained from both patients for publication of this report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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