Compartment Syndrome In Meningiococcal Septicemia
A Hussein Rahoma, A Sher Malek

Citation

Abstract

INTRODUCTION
In meningococcal septicemia the cutaneous lesions become prominent as subcutaneous hemorrhages. In the case presented, these hemorrhages coalesced to form a circumferential lesion around the right leg and foot leading to ischemia and impending gangrene. The skin lesions changed into eschars acting as a tourniquet around the leg of the patient and created compartment syndrome.  

BACKGROUND
The gram-negative diplococcus Neisseria meningitides is a major cause for childhood death in developing countries. The mortality rate remains around 10%, although in some centers, it decreased to less than 5%. Only meningitis is present in 30-50% of cases of invasive meningococcal disease, whereas 10% of cases have only features of septicemia and 40% have meningitis with septicemia.  

Only meningitis is present in 30-50% of cases of invasive meningococcal disease, whereas 7-10% of cases have only features of septicemia and 40% have meningitis with septicemia. The clinical difference between septicemia and meningitis is important because patients presenting with shock are treated differently than patients primarily presenting with increased intracranial pressure (ICP).

PATHOPHYSIOLOGY
In 30% of teenagers and 10% of adults carry meningococci in the upper respiratory tract. Pathogenic strains are found in only 1% of carriers. Immunity to N meningitidis is acquired through intermittent nasal carriage of meningococci and antigenically cross-reacting enteric flora during the first 2 decades of life.

Disease usually occurs sooner than 10 days after a pathogenic strain penetrates the nasopharyngeal mucosa in a susceptible individual and can survive in the blood stream. Disease may involve septicemia and/or meningitis.

MENINGIOCOCCAL SEPTICEMIA
The clinical syndrome results from the activation and continued stimulation of the immune system by pro-inflammatory cytokines. This process is caused directly by bacterial components, such as endotoxins released from the bacterial cell wall, and indirectly by the activation of inflammatory cells. Combined, the processes produce multiorgan failure that usually causes cardiorespiratory depression and, possibly, renal, neurological, and gastrointestinal failure.

CAPILLARY LEAK
Within two to four days after onset of the illness, vascular permeability massively increases. Albumin and other plasma proteins leak into the extravascular space and urine to cause severe hypovolemia and decrease in the venous return. Hypovolemia is resistant to volume replacement and is associated with increased mortality due to meningococcal sepsis. Children with severe disease often require fluid resuscitation involving volumes several times their blood volume in the first 24 hours of the illness, mostly in the first few hours.

The underlying pathophysiology of capillary leak is unclear. Some evidence suggests that meningococci and neutrophils cause the loss of negatively charged glycosaminoglycans that normally are present on the endothelium. Also, the repulsive effect of albumin may be reduced in meningococcal infection; this change allows the protein leak. Albumin normally is confined to the vasculature because of its large size and negative charge, which repels the endothelial negative charge.

COAGULOPATHY
In meningococcemia, a severe bleeding tendency exists with
severe thrombosis in the microvasculature of the skin, often in a glove-and-stocking distribution that can result in amputation of digits or limbs. Clinicians face a dilemma because supplying platelets, coagulation factors, and fibrinogen may worsen the process. Meningococcal infection affects the following three main pathways of coagulation:

1-Endothelial injury results in platelet-release reactions. Along with stagnant circulation due to local vasoconstriction, platelet plugs form to start the process of intravascular thrombosis. Soluble coagulation factors are consumed, and the natural inhibitors of coagulation are down-regulated; this process further encourages thrombosis.

2-The protein C pathway is thought to be crucial in the development of purpura fulminans. A similar rash is seen in neonates with congenital protein C deficiency and in older children who develop antibodies to protein S after varicella infection. Protein C and S levels are low in children with meningococcal disease, but similar levels exist in patients with septic shock, whether or not a severe rash develops.

3-The fibrinolytic system also is down-regulated in meningococcal disease, reducing plasmin generation and removing an aspect of endogenous negative feedback to clot formation.

Meningococcal septicemia is characterized by fever, rash, vomiting, headache, myalgia, abdominal pain, tachycardia, hypotension, cool extremities, and initially normal level of consciousness.

COMPARTMENT SYNDROME

Compartment syndrome is a painful condition that results when pressure within the muscles builds to dangerous levels, preventing blood to reach nerve and muscle cells. In the arms, legs, feet and buttocks, muscle groups—with the nerves and blood vessels that flow beside and through them—are covered by a tough membrane (fascia) that does not readily expand. The resulting unit is called a compartment. If pressure within the compartment is excessive, it can cause damage to blood vessels and nerve and muscle cells.

A difference of less than 30 mmHg between tissue pressure and the diastolic pressure indicates need for fasciotomy.

Mean arterial pressure -- 30 mmHg for children and Diastolic pressure – 20 mmHg for adults. Absolute values are of no use. Maintain the limb at level of the heart as elevation reduces the arterio-venous pressure gradient on which perfusion depends. (perfusion pressure is the difference between pressure at the arterial end (30-35mmHg) and that at the venous end (10-15mmHg) of the capillary.

- Compartmental pressure falls by 30% when cast is split on one side, by 65% when the cast is spread after splitting. Splitting the padding reduces it by a further 10% and complete removal of cast by another 15%. (Total of 85-90% reduction occurs by just taking off the plaster.)

Compartment syndrome can be either acute or chronic. Acute compartment syndrome can have disastrous consequences, including paralysis, loss of limb or loss of life. Chronic compartment syndrome is not a medical emergency, but can be a significant problem for an athlete.

Muscles tolerate 4 hours of ischemia well, but by 6 hours results are uncertain and after 8 hours, the damage is irreversible.

Clinical features, include pain, especially that out of proportion to the injury (child becoming more and more restless /needing more analgesia), pallor, pulselessness, paralysis, paraesthesia etc appear very late and we should not wait for these things.

PRESSURE MEASUREMENT

Normal compartment pressure is zero. There is inadequate perfusion and relative ischemia when this rises to within 10 – 30 mm Hg of diastolic pressure. There is no effective perfusion when it is equal to the diastolic pressure.

WHITESIDE TECHNIQUE OF MEASURING THE TISSUE PRESSURE

Aspirate sufficient sterile saline into one arm of the extension tube to half fill it. Insert the 18 gauge needle into the compartment to be measured and turn the stopcock so that all arms are open. Increase the pressure in the system gradually by slowly depressing the plunger of the syringe while watching the column of saline. The manometer will rise, reflecting the pressure in the system. When this pressure has just surpassed the tissue pressure surrounding the needle a small amount of saline will be injected into the tissue and the saline column will move. The manometer reading at this point reflects the tissue pressure.

CASE REPORT

PATIENT

A six months Malaysian male child had developed fever, cough and shivering. Cough was dry at the beginning and
later became productive. Patient was seen by the pediatrician and was admitted to University Malaya Medical Centre (UMMC). As condition became worse, with signs of meningeal irritation, and respiratory failure, the patient was admitted to an isolation room in the pediatric intensive care unit as a suspected case of meningitis. He was intubated and artificially ventilated.

Within few days patient developed a skin rash all over the body and concentrated over the both legs mainly the right limb. Around the legs, the rash changed into skin discoloration denoting patches of skin ischemia and started to coalesce to form a black circumferential eschar.

The right foot distal to the eschar started to become dark blue, then black.

METHODS
With the patient in the pediatric intensive care unit, all routine investigations were done as full blood counts, full blood biochemistry, blood gases, blood culture and sensitivity testing, and culture of tracheo-bronchial aspirate. Blood culture showed meningococcal septicemia. Lumbar puncture was avoided because of hemorrhagic tendency.

Anti meningococcal drugs were started in the form of Aminoglycoside and Penicillin. Later, Ciprofloxacin, and Ceftazidime, were started according to the culture sensitivity test results.

Later, the treating pediatrician consulted the orthopedic surgeon who put forward some options including the decision to amputate the right foot.

The first author was consulted to see the patient from the plastic surgery point of view. The diagnosis of compartment syndrome was established, and the patient was treated accordingly by immediate escharotomy (Fig 4). Later, debridement was done which let to good healing and epithelialisation of most of the raw areas. Patches of skin grafting were applied to remaining areas.

RESULTS
The result as shown in the pictures was salvage of about 80% of the right foot (Figure: 6 & 7) after immediate escharotomy of the leg (Figure 4). Only the distal toes dropped off and spontaneous healing occurred except in some areas which needed skin grafting. Patient stayed in the hospital for around three months, and was followed up in the plastic surgery and orthopedic clinics as well as the rehabilitation center in the University Malaya Medical Center.

Figure 1
Figure 1: Early cutaneous lesions, note the dark blue coloration of the right foot. Seen 4-5 days after the onset

Figure 2
Figure 2: Right foot shows severe ischemia
Compartment Syndrome In Meningococcal Septicemia

Figure 3
Figure 3: Left leg and foot showing the actual skin gangrene over the left knee and leg.

Figure 4
Figure 4: Right leg after fasciotomy. Gangrene was established over the foot.

Figure 5
Figure 5: Left leg and foot after debridement of eschars

Figure 6
Figure 6 & 7: Both legs after debridement. It shows the gangrene in right foot was limited to the toes only and rest of the foot is viable

DISCUSSION

COMPARTMENTAL ANATOMY OF THE LEG

The anterior compartment is located anterolaterally between the tibia and fibula, bounded deeply by the interosseous membrane and superficially by the deep fascia of the leg. It contains the tibialis anterior, extensor digitorum longus, extensor hallucis longus muscles, as well as the anterior tibial artery and the deep peroneal nerve which innervates all the muscles.  

The lateral compartment is innervated by the superficial peroneal nerve, and contains the peroneus longus and brevis muscles. The compartment is bounded medially by the fibula, and by anterior and posterior crural intramuscular septa.
The posterior compartment contains the gastrocnemius, soleus and plantaris muscles which combine tendiously to form the Achilles tendon. The compartment is bound by fascia and intermuscular septa.

The deep posterior (or medial) compartment is enclosed between the deep fascia of the medial aspect of the leg, the interosseous membrane and a deep transverse intermuscular septum which separates it from the posterior compartment. It contains the tibialis posterior, flexor hallucis longus and flexor digitorum longus muscles as well as the posterior tibial artery and nerve.

Cutaneous lesions as a complication of Meningococcal septicemia can be explained on the basis of understanding of the basic sequences occurring in the cutaneous vessels.

The ischemia which develops in deep tissues especially muscles can be explained on the following bases:

1. Arterial spasm due to increased compartment pressure. Subcutaneous bleeding also can lead to arteriolar spasm.

2. Theory of critical closing pressure- Because of the small diameter and high mural tension in the arterioles, a significant transmural pressure difference (arteriolar pressure minus tissue pressure) is required to maintain patency. If tissue pressure increases or arteriolar pressure decreases so that this difference does not exist, ie, critical closing pressure is reached, they will close.

3. Because of their thin walls, veins will collapse if tissue pressure exceeds venous pressure. If blood continues to flow from capillaries, the venous pressure continues to increase until it exceeds the tissue pressure and venous drainage is re-established. However, this reduces the arterio-venous gradient and reduces arterial inflow.

Histamine like substance is released that dilate the capillary bed and increases endothelial permeability. This leads to intramuscular transudation of plasma with red cell sludging and decreased microcirculation. The muscle gains weight up to 50%.

If this ischemia affects the whole circumference of the extremity as in our patient, it will act as a tourniquet, compressing the vascular tree and compartment syndrome develops.

Fasciotomy can be done by fibulectomy, perifibular fasciotomy or double incision fasciotomy. Compartment syndrome of the foot is not common but can occur in crush injuries. Decompression is done through dorsal incisions and a medial foot incision to decompress the sole of the foot. In our case the syndrome arises from an external pressure. So, just relieving this pressure and then open the deep fascia can be satisfactory.

CONCLUSION

Circumferential skin lesions around any extremity can carry the danger of causing compartment syndrome. We can conclude that it is better to do fasciotomy if there is any circumferential skin lesion around extremities rather than leave it for conservative follow up.

References

17. Parkes AR: Traumatic ischaemia of peripheral nerves with some observations on Volkmann's ischemic contracture. Br J Surg 32:403-414, 1945
Author Information

Ahmed Hussein Rahoma, MS, MD, FRCSI
Associate Professor, and Consultant Plastic Surgeon, Surgery Department, Faculty of Medicines, University Technology Mara

Alam Sher Malek
Professor of Paediatrics, Faculty of Medicine, University Technology Mara