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CASE REPORT

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Aplasia cutis congenita in a Nigerian child: A case report

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Abstract*:* Aplasia cutis congenita (ACC) is a rare skin disorder, the cause is not known but intrauterine infections, drugs, chromosomal and genetic disorders, vascular compromise and trauma have been implicated. Clinically the diagnosis is made based on physical findings indicative of intrauterine disruption of skin development. We present an eighteen hours old neonate with Aplasia cutis congenita, this is aimed at creating awareness in view of the rarity of this condition. Conservative treatment of the ulcers has yielded excellent result but not without complication of acquired syndactyly, the child is being followed up for possible surgery to release the digits.

Key words: Aplasia cutis congenita, Epidermolysis bullosa, Neonate, Honey dressing, Frieden classification, Nigeria

Introduction

Aplasia cutis congenita is a rare congenital developmental skin defects, characterized by well demarcated, oval or circular ulcers or scars.[1] It could be in a localized area or widespread at birth. The condition is most commonly seen on the scalp in 90% of cases,[2] but ACC can affect any part of the body.[2, 3] Aplasia cutis congenita is primarily a clinical diagnosis with no specific histological alterations. At birth most cases of ACC have ulcerated lesions which may show total absence of skin, sometimes extending to the bone or dura.[3] Aplasia cutis congenita can be associated with other physical abnormalities or malformation syndromes, chromosomal or other disorders such as ectodermal dysplasia and epidermolysis bullosa.[4]

Frieden created a classification system consisting of nine groups based on the number and location of the lesions and the presence of associated malformations.[4] In Frieden's classification group 1: This was a scalp ACC without multiple anomalies, a collar of hair was often seen around the defect. This can be autosomal or sporadic in occurrence. Group 2; was a scalp involvement with limb anomalies usually lower limbs with asymmetric lesions. Group 3; was a scalp ACC with epidermal and sebaceous nevi. Some patients reported in this group had ophthalmic and neurologic findings typical of epidermal nevus syndrome. Group 4; ACC had hair collar overlying deeper embryologic malformation. Examples were meningomyelocele, leptomeningeal angiomatosis, porencephaly, gastroschesis etc. In group 5: This was ACC associated with fetus papyraceous, while in group 6 ACC was associated with siplex, junctional or dystrophic types of epidermolysis bullosa. Group 7: This was ACC localized to the extremities without epidermolysis bullosa. Group 8; this was ACC specifically due to teratogens. Lastly in group 9: This was ACC associated with malformation syndromes such as Down syndrome, Patau syndrome etc.

The exact pathophysiological mechanism of this disorder is not clear but propositions included; intrauterine trauma, vascular compromise, maternal infections and medications. Aplasia cutis congenita is typically sporadic but autosomal dominant and less commonly autosomal recessive cases have been reported. Globally the estimated incidence of ACC is 3 in 10,000 live births.[2,4] If the defects is small recovery is usually uneventful, with gradual epithelization and formation of hairless, atrophic scar over several weeks. Sometimes surgical intervention may be necessary. The sex distribution is equal except if it is associated with an X-linked malformation syndrome.

The treatments of ACC depend primarily on size, dept and location of the defect and therapy of associated disorders.[5] Variety of dressings both adhesive and non adhesive materials have been used.[6, 7] Surgical repairs of large defects can be done.

This paper present Aplasia cutis congenita; with sole aim of creating awareness of this rare but treatable condition with good outcome depending on the size of the lesion.

Case Report

MBM, 18 hours old product of term female neonate was admitted into the Neonatology Unit of the Bingham University Teaching Hospital Jos on 13th Jan 2015. The mother was a 23 year old lady whose husband was a 30 year local security guard. The pregnancy was uneventful, supervised and delivered in a Plateau State Specialist Hospital Jos. The baby was referred to us when the lesions were noticed at birth. There was no family history of congenital malformation.

Examination revealed symmetrically distributed ulcers involving both upper and lower limbs. There were no

bullae, discharges, or bleeding. The baby was afebrile temperature of 36.7°C. The baby weighed 2.4kg; the length and the occipito-frontal circumference were within normal limits. The ulcers covering 5% of the total body surface area on each hand, while it covered 10% and 9% on both the right and left leg respectively (see figure bellow). There was no evidence of inflammation. Other systems examination was essentially normal. A diagnosis of aplasia cutis congenita was made with a differencial diagnosis of epidermolysis bullosa.

The patient's packed cell volume was 46%, WBC was 10×10^9/L with 68% neutrophils and 32% lymphocytes, the VDRL was non reactive and the HIV serology was negative. The blood culture yielded no bacterial growth and the wound swab culture was also negative. The serum electrolytes were within normal limits. Patient was commenced on 1/5 normal saline in 8.4% dextrose, intravenous cefuroxime 120mg 12 hourly, intramuscular gentamycin 6mg 12 hourly and a daily wound dressing with honey, she responded well to these treatments and within six weeks the wound had healed except for a complication of acquired syndactyly. Surgical intervention to release the digits is being planned.

Fig 1: Ulcers at presentation

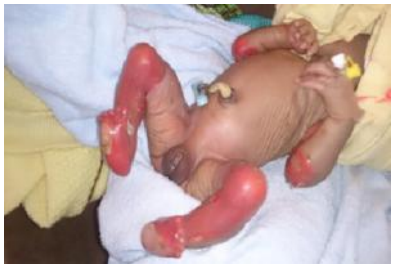
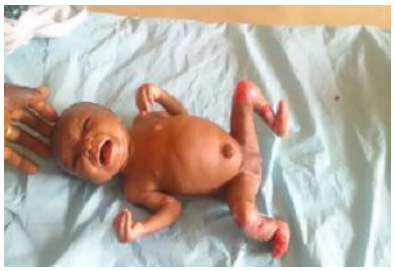


Fig 2: After 6 weeks of treatment



Discussion

Aplasia cutis congenital first reported by Campell in 1826,[8] is a rare disorder. Following this report there have been few cases reported in literature most of which were outside Africa. This condition is most often seen on the scalp in 90% cases.[2, 3] The index child's lesions affected the extremities making it one of the rare group of ACC. At birth most cases of ACC have ulcerated lesions which may show total absence of skin, sometimes extending to the bone or dura.[3] The later did not apply to the index infant, because lesion did not affect the bones; neither was there any lesion on the scalp.

The index child has no other associated malformation defects; this therefore put her in group 7 of the Frieden's classification.[4] In this group, neonates have ACC confined to the extremities without obvious malformation or epidermolysis bullosa. This particular type is said to be very rare, however there was a report of two families with multiple members having ACC on the pretibial lower extremities and the dorsal aspects of the hand and the feet,[9]these reports described features similar to the index child. (See figure).

The index infant has no history suggestive of fetus papyraceus or placental infarcts as the placenta was reported to be normal. In group 8 of Frieden classification of ACC teratogens are known causes. These had also been linked in few cases to intrauterine infections with herpes virus, varicella zoster virus or exposure to methimazole.[10] The mother of this child had no history suggestive of viral infections in pregnancy neither was she exposed to medications apart from the routine antenatal drugs. Ercan et al[1] reported 3 cases of ACC, the first one was linked to intrauterine infection (rubella), the second case was linked to trisomy 13 syndromes and the third case was linked to fetal valproate syndrome. None of these factors appeared to be relevant in our infant.

Pathologically the lesions in ACC are non inflammatory and well demarcated as was the case in the index infant. Laboratory investigations in the index infant were all within normal. This is not surprising; as it has been well documented in the literature that there were no specific laboratory abnormalities that were consistently found in this condition.[2, 3, 4] Elevated alpha feto protein in maternal serum and amniotic fluid as well as elevated acetylcholinestrases in amniotic fluid had been tried in the past, as biochemical markers but they have been abandoned because they lack sensitivity and specificity.[11]

The index child responded very well to medical treatment, within six weeks the wound were virtually completely healed. The decision to use medical, surgical or both modes of therapy depends primarily on size, dept, location of the defects and therapy of associated abnormalities.[5] This child had occlusive honey dressing, but in large defects ACC surgical repair including excision with primary closure, use of tissue expanders and rotation of flap to fill defects, skin and bone grafting may be required.

The prognosis in this child was excellent. This was in conformity with the usual excellent prognosis occurring in groups 7 of the Frieden classification to which this child belongs. But we are mindful of anticipated complications, such as large scars, contracture and acquired syndactyly. The index child had developed acquired syndactyly (See fig 1B). In other types of ACC the prognosis is dependent on the severity of the lesion and other congenital abnormalities.

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CASE REPORT

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Alagille syndrome in an eleven year old Nigerian child – A case report

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Abstract*: Background* Alagille syndrome (AGS) is a rare cause of prolonged jaundice. It has an autosomal dominant inheritance pattern expressed variably, with a reported incidence of 1:100,000 live births in the United States. The objective is to highlight the clinical features and diagnostic challenges to this rare cause of cholestatic jaundice.
Case Report: T.O, Female 11 years, had a history of recurrent episodes of yellowness of the eyes, first noticed soon after birth, associated with pale stools, dark urine, and body itching. From age three she was noticed to have progressive loss of sight, recurrent body swellings and irrational talks. She was diagnosed to have a cardiac murmur at age six, when diagnosis of ALS was made due to features of, recurrent jaundice and perculiar facies of broad forehead, flat nasal bridge, prognathia and genetic report of no mutation in JAG 1 gene, karyotype 46XX. At the time of current hospital admission she was in addition small for age, had hepatosplenomegy, ascities and talked irrationally. Chest radiograph showed multiple butterfly vertebrae; Echocardiograph aortic and pulmonary stenosis; Liver aminotransferase were marked elevated. Brain MRI showed multiple chronic infarcts. She was diagnosed with Allagille syndrome presenting in hepatic failure with encephalopathy.
Conclusion: Allagille syndrome should be considered early in older children with persistent cholestatic jaundice.

Key words: Allagille, syndrome, cholestatic, jaundice, hepatic encephalopathy and congenital heart disease.

Introduction

Alagille syndrome is a complex hereditary disorder that is associated with cardiac, hepatic, skeletal, ocular, renal and facial abnormalities first reported by Alagille et al. in 1969.[1] Peculiar facie, chronic cholestasis, posterior embryotoxon, butterfly like vertebral arch and peripheral pulmonary artery stenosis are the main features that characterize syndromic paucity of interlobular bile duct.[2,3,4] Common facial features include broadened forehead, pointed chin, and elongated nose with bulbous tip, which may not be obvious during infancy but may become more apparent as the child ages. The ocular abnormalities [5] of posterior embryotoxon, an opaque ring present in the cornea, were seen in about 75percent of patients reported in one large series conducted by Emerick et al.[6] Other ophthalmologic findings reported include retinitis pigmentosa, pupillary abnormalities, and anomalies of the optic disc.

Alagille syndrome is an autosomal dominant disorder with variable expression. Mild-to-moderate mental retardation may also be present. Most cases of Alagille syndrome are caused by a mutation in the *JAGGED1 (JAG1)* gene; with less than one percent of cases resulting in a mutation in the notch-2 (*NOTCH2)* gene as the cause.[7,8,9] The JAG1 gene provides instructions for making a protein called Jagged-1, which is involved in an important pathway by which cells can signal to each other. The Jagged-1 protein is inserted into the membranes of certain cells, connected with other proteins called Notch receptors, which are bound to the membranes of adjacent cells. These proteins fit together like a lock and it's key. When a connection is made between the Jagged-1 and Notch proteins, it launches a series of signaling reactions (Notch signaling) that affects cell functions.[8,9] Notch signaling controls how certain types of cells develop in a growing embryo, especially cells destined to be part of the heart, liver, eyes, ears, and spinal column. The Jagged-1 protein continues to play a role throughout life in the development of new blood cells. The NOTCH2 gene provides instructions for making a protein called Notch2, a member of the Notch family of receptors. The lack of *Notch* signaling causes errors in development that result in missing or narrowed bile ducts in the liver, heart defects, distinctive facial features, and changes in other parts of the body. People with JAG1 gene mutations may have one or more of

these problems. The syndrome has been mapped to the chromosome 20 (20p12-jagged-1 locus) *JAG1*.[8, 9] A minority (6-7percent) of patients have complete deletion of *JAG1*, and approximately 15-50percent of mutations are spontaneous.[10,11] In the United State, reported incidence rate is approximately one case in every 100,000 live births.[12] Alagille syndrome is the fourth leading indication for liver transplant.[13] The first reported case in Nigeria was by Akinyinka et al in 1998.[14]
The objective of this case report is therefore to highlight the clinical features and diagnostic challenges of this rare cause of cholestatic jaundice.

Case Report

T.O, a female presented at age of 11 years with a history of recurrent yellowness of the eyes since birth, progressive loss of sight and recurrent body swelling of over 5years, reversal of sleep and irrational talk of a month duration.
Jaundice was noticed on the first day of life and this was present most days with only few days jaundice free in between episodes. It progressively deepened with associated passage of pale stools and dark urine. There was associated body itching and generalized whitish body rash in early childhood. At about age 3, she was noticed to have been bumping into objects from gradual loss of sight. This led to her withdrawal from formal school to a school for the blind. She had a fall about this time and sustained a fracture of her left femur and was subsequently home schooled. The body swelling involved the lower limbs and abdomen mainly. She was seen at various hospitals and treated for the yellow eyes and body swellings with several medications that included urosordil 750mg twice daily to reduce itching. She was reviewed by an ophthalmologist for her poor vision and treated for the pathological fracture by the orthopedic surgeon. Because of heart murmurs she was diagnosed to have ventricular septal defect (VSD) at 8months and at 6years with aortic and pulmonary stenosis following echocardiography evaluation and had surgery in India. With the combinations of clinical features of cholestatic jaundice, visual impairment, cardiac lesions and genetic testing, she was diagnosed to have Allagille syndrome at the age of 6years old. Her karyotype was 46XX, and genetic analysis showed no mutation in JAG 1 gene.

Her pregnancy and delivery was supervised and mother well except for a history of polyhydramnios in the 3rd trimester. Birth weight was 2.4kg and immunization was completed according to the National programme of immunization guideline. She had delay in walking until 2years old and eruption of teeth. She is the second of four children in a monogamous non-consanguineous marriage; other children alive and well. Father is a 49years old lawyer; mother is a 39years old caterer with tertiary level of education with no family history of a similar illness.

She presented to our health centre at age 11years with deepening jaundice, increasing body swelling, reversal of sleep and irrational talk of one month duration. Prolonged bleeding was noticed from puncture site, with epistaxis while on admission. She had a history of poor growth when compared to her other sibs. She was found to be small for age, ill looking and deeply icteric (greenish tinge), with sparse fluffy hair, broad forehead, flat nasal bridge and prognathia (fig 1 and 2). She had poor oral hygiene with a cross bite dentition. She was, pale, acyanosed, afebrile, with digital clubbing of grade 3 and bilateral pitting edema up to the knees. She had neck retraction, thin long limbs, broad wrist and elbow joint with rachitic rosary, genu valgum. She was drowsy with irrational talks had neck stiffness and increased muscle tone. Eye examination showed bilateral optic atrophy. Her precordium was hyperactive with a palpable thrill; normal first and second heart sounds with a grade 4 pansystolic murmur loudest at the left upper sternal border radiating to the axilla and root of the neck. Her abdomen was distended with prominent abdominal veins, tense generalized vague tenderness with moderate ascites. The diagnosis was Alagille syndrome with hepatic failure and encephalopathy.

Her chest radiograph revealed multiple butterfly vertebrae (fig 3) and brain magnetic resonance imaging (MRI) multiple chronic infarcts involving the right parietal lobe and the basal ganglia (fig 4). Laboratory investigations revealed deranged liver function test with an 8-fold increase aspartate aminotransferase (AST), a 2 -fold rise alanine aminotransferase (ALT) and Gamma glutamyltranspeptidase (GGT), a 9-fold rise Alkaline phosphatase (AP); prolonged prothrombin time (PT) and partial thromboplastin time (PTT), and abnormal international normalization ration (INR); decreased serum proteins and albumin. Viral serology markers; namely hepatitis B surface antigen (HBsAg) or anti-HBV core (anti-HBC) for hepatitis B infection and Hepatitis C virus (HCV), anti- HCV for hepatitis C infection and HIV screening were negative. Others were hypokalemia, hypocalcemia, hypophosphatemia, and thrombocytopenia. She had intravenous unasyn, oral neomycin and lactulose and other supportive care. Parents were counseled on need for liver transplant. By the second week of admission patient deteriorated and became comatose and was bleeding from the nose and puncture sites and died subsequently.

Fig 1: Showing fluffy hair, deep jaundice, flat nasal bridge, broad nose

Fig 2: Showing broad forehead and nose, with flat nasal bridge, widely spaced eyes

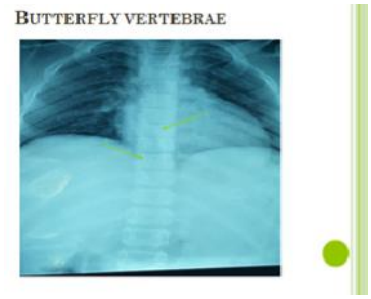

Fig 3: Showing multiple butterfly vertebrae

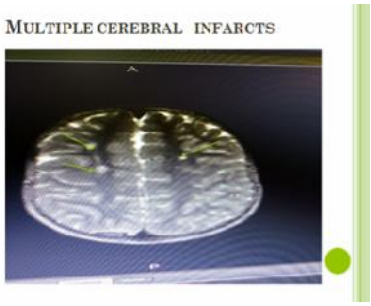

Fig 4: Showing multiple cerebral infarcts

Discussion

The case report was diagnosed Alagille syndrome (ALGS) with hepatic failure and encephalopathy, an autosomal dominant multi-system disorder affecting several body systems based on clinical presentation. The case report did not have JAG1 mutation, hence the possibility of Notch 2 mutation. However, the typical clinically consistent features of the syndrome has mutationin JAG1 in up to 94percent of cases, with a small 2percent having a mutation in NOTCH2.[8,9] Some reports have shown thatover half of individuals with mutations in the gene did not inherit it from either parent, and thus have a de novo mutation.[8,9,15]

This case report had the constellation of five main symptoms consistent with ALGS,[2,3,4] namely chronic cholestasis characterized by the paucity of intrahepatic bile ducts, congenital cardiac anomaly involving pulmonary stenosis, butterfly-like vertebral abnormalities, ocular changes of posterior embryotoxon, and peculiar facial abnormalities (broad forehead, deep-set and widely spaced eyes, small pointed chin, and saddle or straight nose). The case report was diagnosed at about 6years of age after several hospital visitations. The diversity of symptoms and lack of routine genetic analysis may have contributed to the delay diagnosis. In the case reported by Akinyinka et al,[14] the characteristics facial features suggestive of Alagille syndrome and the clinical and echocardiological evidence of pulmonary stenosis were first observed when the child was 58months. Because of the diverse clinical features, diagnosis may be delayed until established by age 4-5, which may explain why our case report was diagnosed at age 6 after several hospital consultations. Another report showed that affected children were evaluated when younger than 6months for either neonatal jaundice (70percent), or cardiac murmurs and symptoms (17percent).[12]

The case report had sub optimal growth with smallness for age, fluffy hair changes and body swelling with low proteins and albumin levels as features of chronic liver disease. Sub optimal growth has been documented [12] as a consequence of Alagille syndrome. The history of fall and fractures could have been associated with presence of rickets with other skeletal abnormalities. This case report had rickety rosary chest and ribs cage, hypoplasticenemal teeth, butterfly hemivertebrae and low serum calcium and phosphate. Vitamin D levels could not be determined. Emerick et al [6] in a large series of patients with Alagille syndrome reported the presence of butterfly hemivertebrae in half of the patients analyzed. Visual loss from optic atrophy seen in this case has been reported as part of a posterior embryotoxon, which was observed in more than 75percent of patients in one large series conducted by Emerick et al.[6]

Treatment is usually supportive and some would require liver transplant. As at 2010 ALGS has been reported as fourth most common cause of liver transplantation in children.[7] The case report died as a young adolescent after 15days on admission. The presence of significant co- morbidities, that include cardiac malformation, cholestatic liver disease and vascular brain lesions seen in this case have been reported as some of the leading causes of morbidity in patients with ALGS.[10,12]

A 20years predicted life expectancy in 75percent for all ALGS patients, 80percent for those not requiring liver transplantation, and 60percent for those who required liver transplantation was reported by Emerick, *et al*.[6] The case report lived up to young adolescent age before death with the several morbidities. She did not have the benefit of transplantation. Of the 41 cases in the Korean report,eight patients died after a median period of 2.67 years (range, 0.33-15 years) and those with combined severe liver and heart disease had the poorest survival (P<0.001).[16]

ALGS could be differentiated from other rare conditions that present as cholestatic liver disease from its syndrome clinical features, laboratory tests and imaging studies.

Conclusion

Alagille syndrome is a rare syndrome, presenting as cholestatic liver disease, with peculiar features. Multidisciplinary approach in management is required and survival depends on severity of associated morbidities and available transplant facilities care.

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CASE REPORT

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Usefulness of peripheral nerve block as an aneasthetic technique in a critically ill Child– A case report

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Abstract*:* Regional anaesthesia in children is a growing field of interest in current anaesthesia practice. We report a case of brachial plexus block for a child with severe forearm necrotizing fasciitis and septicaemia. The need to avoid the multiple shortcomings of general anaesthesia in a critically ill child prompted the use of regional anaesthesia. This case is reported to highlight the prospect of regional anaesthesia for critically ill children who require surgical interventions in resource-poor settings.

Keywords: brachial plexus block, children, critical illness, septicaemia, regional anaesthesia

Introduction

The growing use of regional anaesthesia in infants, children and adolescents has increased the popularity of peripheral nerve blocks (PNB) in children. PNB is a form of regional anaesthesia which is accomplished by injecting a local anaesthetic near a nerve/nerves that controls sensation and movement to a specific part of the body.[1]This causes temporary numbness in the area. The growing use of the technique is a result of the increased confidence of anaesthetists in performing peripheral nerve blocks. PNB are typically used for surgeries of the upper and lower extremities, also for some procedures around the neck and groin.
Peripheral regional anaesthesia is of great utility in children undergoing surgeries of the upper extremities. In contrast to general anaesthesia, it avoids airway instrumentation and the use of many drugs.[1,] The peculiarities of this technique include the meticulous attention to dosing as a result of the poor development of connective tissues and the likelihood of extensive spread of locally administered drugs. Other important considerations include the risk of rapid absorption with attendant systemic toxicity, reduced duration of action and age-related anatomic variations.

The use of ultrasound guidance during axillary approach to brachial plexus blockade allows for real-time visualizaion of anatomical structures; however some anaesthetists prefer nerve stimulation to guide peripheral blockade, while some use both methods for greater accuracy and safety. However, the use of ultrasound scan and / or peripheral nerve stimulator is subject to availability. Our centre does not have the capacity for either. Complications of axillary approach include infection at the puncture site, axillary tenderness, heamatoma, intravascular injection and nerve damage[1,2]. This case report is to enumerate the prospects of blind axillary approach to peripheral brachial nerve block, in a low-resource setting, in a sick child who is not suitable for general anaesthesia.

Case report

Presentation, admission and treatment

A 9 year old presented in the children emergency ward of Olabisi Onabanjo University Teaching Hospital, Sagamu, Nigeria on referral from a private hospital where he had presented with a 6 day history of fever, body pains and general malaise. He was admitted at the private hospital and given intravenous medications through the dorsum of the left arm. After three days the left arm subsequently became swollen, tender with limitations of active movements, with blister formation. These, coupled with the persistence of the symptoms necessitated his referral to the teaching hospital.

On admission he was acutely ill looking, pale and febrile (39ºC). He was in respiratory distress, tachypneic, tachycardic with a tender hepatomegaly. The left arm was swollen, tender with coalescing blisters, necrotic eschar and copious purulent exudates. The packed cell volume was 15% and chest x-ray showed evidence of pulmonary oedema. generalized widespread fluffy exudates. A diagnosis of septicaemia with focus in necrotizing fasciitis of the left upper limb, severe anaemia and congestive cardiac failure was made. He was resuscitated, commenced on parenteral antibiotics, diuretics, digitalis and blood transfusion. Bacteriological culture of pus aspirate yielded Proteus sp. He was planned for extensive wound debridement three days after admission to remove focus of sepsis.

Pre-anaesthesia Review

When he was reviewed on the morning of surgery he was still having temperature spikes despite intravenous antibiotics. Urinary output was adequate. There were no

known allergies. He had been on nil per oris for 12 hours. He was mildly pale and weighed 28kg which was appropriate for age. The respiratory and heart rates, blood pressure and precordial activities were normal. Airway assessment for ease of intubation and mask ventilation showed no abnormality. He had Mallampati 2 class with adequate mouth opening.
Abdominal examination revealed minimal ascites and hepatomegaly of 4cm below costal margin. The left elbow was in gauze and bandaged dressing. The initial laboratory findings included PCV 30%, negative retroviral screening, normal haemoglobin genotype and INR of 1.0.
Assessment was resolving septicaemia, resolving congestive cardiac failure. An American Society of Anaesthesiologist (ASA) physical status class assessment of IIIE was made.
The mother consented to the use of peripheral nerve block with sedation following adequate patient information.

Procedure In theatre

The child was placed supine. Pulse oximeter and precordial stethoscope and Non- Invasive Blood Pressure (NIBP) measurement apparatus were attached. Baseline parameters were; PR 107bpm, Bp 106/67mmHg, SpO2 was 97% at room air.
He was pre-medicated with IV hydrocortisone 100mg, sedated with IV 2.5mg midazolam with IV Paracetamol 50mg for analgesia. The injectate for the nerve block was made by mixing 5mls of 0.5% plain bupivacaine, plus 10mls of lignocaine in adrenaline and 10mls of sterile water.
A 22G hypodermic needle, first made blunt by passing it through the sterile plastic sheath, was used for the axillary block. The patient's arm was abducted to 90 degrees (Fig 1) and the palpation method was used to identify the axillary artery. Skin infiltration was done with plain lidocaine. Test aspirate was done and 20mls of the injectate was used for the axillary block and 5mls for the blockage of the musculocutanoeus nerve (Fig 2 and 3). Efficacy was confirmed with loss of pain to surgical stimulation after 10 minutes. Supplemental 100% oxygen was administered by face mask at 6l/minute flow rate.

Fig 1: Palpation method of locating the axillary artery prior to anaesthestic infiltration

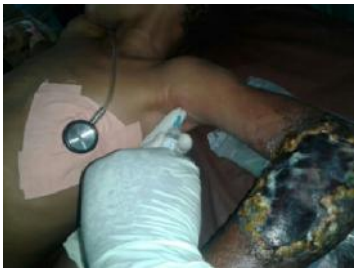

Fig 2: Injection of local anaesthetic into the axillary sheath

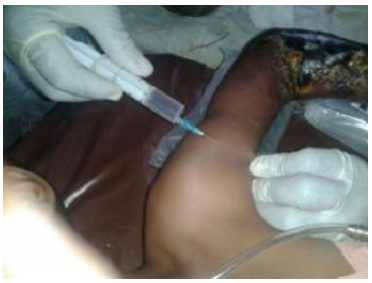

Fig 3: Blockage of the musculo-cutaneous nerve.

The debridement lasted 15minutes, with an estimated blood loss of 50mls. He had 100mls of 4.3% dextrose in 5th saline. The pulse rate ranged between 120 and 135 beat per minutes during the procedure. There were no critical incidences or complications intra-operatively
Post operatively the arm was put in a POP back slab for support, and patient was transferred to the recovery room and later back to the ward. He made full recovery and was discharged a month later.

Discussion

Although the clinical diagnosis of septicaemia was bacteriologically confirmed, it was unresolved whether cellulitis of the arm resulted to septicaemia or the reverse occurred. The cellulitis could also have been due to drug -induced tissue injury with subsequent bacterial colonization. However the removal of pus and eschar in this patient was essential for infection control.[3] Anaesthetists are involved in the care of septic patients for resuscitation, intensive care and anaesthesia for infection source control. The latter includes drainage of abscess, debridement of necrotic tissue, removal of infected devices and foreign bodies. The anaesthesia of septicaemic patient for infection source control poses some challenges because of the gross heamodynamic instability and these risks are higher in children.

The available anaesthetic technique options include general anaesthesia or a brachial plexus block. The advantages of the use of regional anaesthesia as opposed to general anaesthesia include avoidance of the airway, reduced postoperative nausea and vomiting, improved tissue perfusion during re-implantations and the ability to provide a continuous technique for repeated procedures, and early hospital discharge.[4] Brachial plexus block was chosen in this case because a full general anaesthesia will lead to administration of anaesthetic agents that may further depress the myocardium thus worsening the heart failure. It is also important to avoid the airway and the chest due to the accompanying pulmonary oedema. Peripheral nerve blocks in septic patients cause less haemodynamic instability and less immunosuppression but also carry the risk of nerve damage, allergic reaction, intravascular injection and risk of toxicity.[3] The index case had none of these complications.

Various approaches can be used for the brachial plexus block; interscalene, supraclavicular, infraclavicular and

axillary approaches. Axillary approach is the commonest, easiest and safest approach in all age groups especially if it is done under the palpation method alone.[1,2,5] The continuous axillary block variant involves inserting a peripheral block catheter in the perineural space for intermittent injections of the local anaesthetic solutions. This improves outcomes after microvascular surgery.[5] Another safe method is the lateral infraclavicular approach which reduces the risk of pneumothorax[2]. The latter method is new in children, but gives a wider degree of blockage, and can be done with the patient's arm at the sides thus minimizing pain. Both can be made safer and easier with the use of peripheral nerve stimulator and / or ultrasound scan. [6,7.] Unfortunately, in low resource centres where peripheral nerve stimulator and an ultrasound are not routinely available, the blind technique by palpation becomes extremely useful and handy.

There are no published studies comparing general anaesthesia to upper limb regional anaestheisa[1]. There are also none comparing general anaesthesia with or without upper limb blockade. Performing a nerve block in children usually requires the use of sedation. Benzodiazepines and short acting opioids are useful in this regard. Midazolam, a short acting benzodiazepine was used in the index case. It provided sedation, hypnosis and anterograde amnesia without compromising the airway or the cardiovascular status of the patient. Paracetamol was used for pre-emptive analgesia after the effect of the nerve block must have worn off. This also avoided the respiratory depressant effect of opioids.

Conclusion

The case highlights the usefulness of peripheral nerve block, in a septic peadiatric patient undergoing infection source control surgery. It is a viable option to consider when the patient is unstable for general anaesthesia. It is safe, cost friendly and effective.

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Reference

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