Case Report

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Meningocele in a Two Year Old Child: A Case Report

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Abstract

Meningocele is the cystic dilation of the meninges containing cerebrospinal fluid through a defect in the vertebral column without neural tissue. Meningocele has a prevalence of about 2.2 per 1000 live birth in North-West Nigeria. This is a two year old male child presenting with a sacral mass since birth, with fecal and urinary incontinence and failure to achieve vital milestones. The patient had a plain spinal radiograph that showed a soft tissue density mass located at the posterior aspect of the sacral spine with associated scoliosis of the lumbosacral spine. He also had a spinal magnetic resonance imaging that confirmed the diagnosis of a meningocele in the posterior aspect of the sacral spine. The patient had a surgical repair of the meningocele and subsequently discharged to continue physiotherapy and regular hospital follow-up visits. We report the radiologic findings of meningocele in this patient due to its mode of presentation.

Keywords: Spinal dysraphism, Cerebrospinal fluid, Vertebrae, Dura

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Introduction

Meningocele is a form of open neural tube defect characterized by cystic dilatation of meninges containing cerebrospinal fluid without any neural tissue.

Meningocele is an out-pouching of the leptomeninges through a developmental defect in the dura, the arches of the vertebrae at one or more levels are involved with protruded meningeal sac covered with only a layer of skin.[1,2]

Meningocele has an incidence of about 3.4 per 10,000 live births in USA and a prevalence of 2.2 per 1000 live births in North-West Nigeria, despite folic acid fortification of diet and health education.[3,4]

Some genetic and environmental factors are responsible for the causation of neural tube defects that occur within the third to fourth week of intrauterine life.[3,5,6]

Neural tube defects development is often associated with many risk factors, among which are maternal diabetes, maternal febrile illness, antiepileptic medications and family history of neural tube defects.[3,7,8]

Meningoceles are frequently located posteriorly over the thoracic and sacral areas at birth and constitute about 10% of all patients with spina bifida.[9,10] Rarely meningoceles may be seen protruding through anterior, lateral or anterolateral defects with more than three-quarter of cases found in females. Uterine anomalies are often associated with anterior meningoceles.[10]

A rare form of meningocele is the non-dysraphic type, which is characterized by the absence of a congenital defect of the vertebrae and often seen in the anterior thoracic region and frequently associated with neurofibromatosis or less often Marfan's syndrome.[9,10]

Meningocele is usually associated with a congenitally dysraphic vertebrae with the spinal cord remaining entirely confined to the vertebral canal.[9–11]

Spinal meningoceles are also seen to be associated with conditions like Marfan syndrome, neurofibromatosis, autosomal dominant polycystic kidney disease and generalized connective tissue disorders of uncertain type.[3,12,13]

Spinal meningoceles often cause neurological deficits, skeletal deformities, anorectal and urinary bladder malfunctions, paraparesis, paraplegia and sensory loss below the cord level of involvement.[14–16]

Meningoceles are most often diagnosed following ultrasonography during infancy, but post natally, plain radiography, conventional myelography and post contrast computed myelography and magnetic resonance imaging of the spine are also helpful in establishing the diagnosis of meningoceles.[17,18]

The treatment management of spinal meningoceles entails a multi-disciplinary approach, but surgical repair (laminectomy) is the mainstay of management.[3,9]

Case Report

This is a two year old male child, the 8^{th} child of his mother who was aged 40 years at the time of his birth. He was referred for plain radiography and magnetic resonance imaging of the spine on account of a sacral mass since birth, scoliosis, fecal and urinary incontinence and failure to achieve most developmental milestones for his age.

The mother never had any antenatal care, never took folic acid but admitted to taking lots of local concussions during the index pregnancy. She also denied history of similar case in the remaining siblings or close blood relations.

On examination the child is conscious and alert and appeared well oriented, he is not in any form of distress, not pale or dehydrated. Had normal pulse and respiratory rate. The limbs appeared weak with a power of about grade I-II. He could not stand on his limbs or make attempts to move. There is a mass in the posterior aspect of the sacral region well covered with an overlying healthy skin and felt soft upon touching and measured about 16cm x 7.5cm in cranio-caudal and medio-lateral dimensions respectively.

The patient had a plain radiograph of the spine in anterior-posterior and lateral views, that showed a soft tissue density mass in the sacral region posteriorly on lateral view and right lateral aspect of the spine on AP view (figure 1). No calcification or lucency and air-fluid levels were demonstrated within this sacral mass.

Spinal magnetic resonance imaging (MRI) showed a cyst like out-pouching containing cerebrospinal fluid (CSF; hypo-intense on T1 and hyper-intense on T2) with coverings of the meninges and seems continues with the spinal canal through a defect in the posterior aspect of the first sacral vertebra (figure 2), compression of the exit nerve roots were also demonstrated.

A complementary abdominal ultrasound scan showed normal abdominal situs, lesional ultrasound showed a fluid containing echo-reflective sac posteriorly in the sacral spine.

The patient had a successful repair of the meningocele and later referred for physiotherapy and advised on regular follow-up visits.

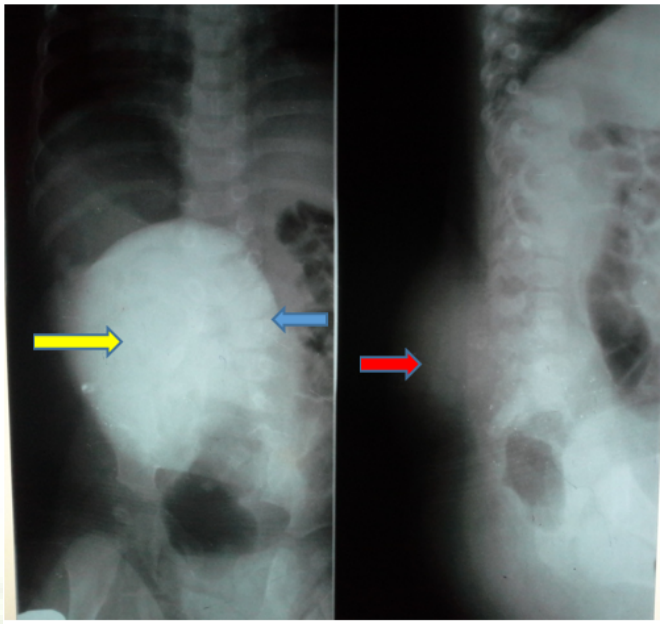

Figure 1: Plain radiograph of the lumbosacral spine anterior-posterior (AP) on the left and lateral view on the right showing scoliosis of the lumbosacral spine (left blue arrow) and a soft tissue density mass in the distal spine; posteriorly located on the lateral view (right red arrow) and right lateral aspect of the spine on the AP view (right yellow arrow).

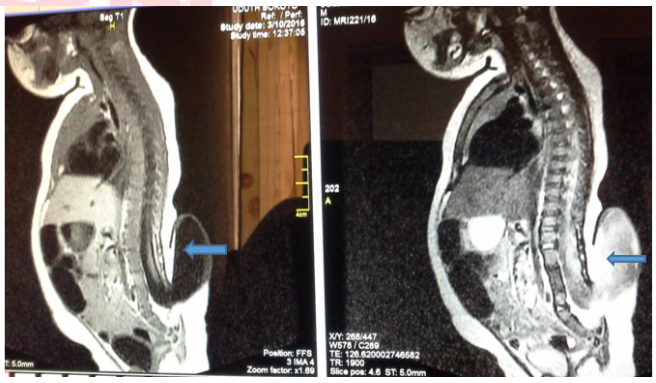

Figure 2: MRI images; sagittal views with T1 on the left and T2 on the right each showing a posteriorly located CSF filled sac like structure (left blue arrows) that seem continues with the spinal canal at the first sacral vertebral level. No spinal cord tissue is seen within this sac. This is a meningocele.

Discussion

Meningocele is a form of open neural tube defect characterized by cystic dilatation of meninges containing cerebrospinal fluid without any neural tissue as documented by most literatures. The index case also had a cyst like mass containing CSF with

meningeal coverings conforming to what is documented in the literature.

Most spinal meningoceles are found in the female gender in about three-quarter of cases,[10] the case presented happens to be a male falling among the remaining one-quarter of cases of meningoceles.

Neural tube defects development is often associated with many risk factors, among which are maternal diabetes, maternal febrile illness, antiepileptic medications and family history of neural tube defects.[3,7,8] The mother of the index case denied history of diabetes and family history of neural tube defects, however she confessed to having episodes of febrile illnesses and intake of local herbs during the pregnancy of the index case.

Some genetic and environmental factors are responsible for the causation of neural tube defects that occur within the third to fourth week of intrauterine life.[3,5,6] No any form of genetic factor was implicated in this case but rather the mother had no history of antenatal care and intake of folic acid which is been postulated to reduce the occurrence of neural tube defects in the 3^{rd} to 4^{th} week of intrauterine gestation.

Meningoceles are frequently located posteriorly over the thoracic and sacral areas at birth and constitute about 10% of all patients with spina bifida.[9,10] The index case also presented with a mass extruding from a defect in the posterior aspect of the first sacral bone since birth, conforming to these literatures.

Meningocele is usually associated with a congenitally dysraphic vertebrae with the spinal cord remaining entirely confined to the vertebral canal, there is association of meningocele with neurofibromatosis, Marfan's syndrome, autosomal dominant polycystic kidney disease and generalized connective tissue disorders,[9–11] the index case had a dysraphic first sacral vertebra with an intact spinal cord within the vertebral canal. No any feature or evidence to suggest the association with polycystic kidney disease, Marfan syndrome, neurofibromatosis or any form of connective tissue disorder at the time of this report.

Meningoceles are most often diagnosed following ultrasonography during infancy, but post natally, plain radiography, conventional myelography and post contrast computed myelography and magnetic resonance imaging of the spine are also helpful in establishing the diagnosis of meningoceles.[17,18] The index case was diagnosed at the age of two years, he had plain radiography and magnetic resonance imaging of the total spine; the diagnosis was finally made following MRI as it is the gold standard in diagnosis of meningocele, thereby conforming to most literatures.

Spinal meningoceles often cause neurological deficits, skeletal deformities, anorectal and urinary bladder malfunctions, paraparesis, paraplegia and sensory loss below the cord level of involvement.[14–16] The index case had paraparesis, poor developmental milestones, urinary and fecal incontinence to mention but a few, thereby conforming to these literatures.

The treatment management of spinal meningoceles entails a multi-disciplinary approach, but surgical repair (laminectomy) is the mainstay of management.[3,9] The patient had a successful surgical repair of the meningocele and was referred for physiotherapy and regular hospital follow-up visits conforming to most literatures.

Conclusion

Adequate health education and good antenatal care with adequate folic acid intake should be encouraged amongst pregnant females, routine intrauterine ultrasonography should also be encouraged to detect cases of meningocele earlier in order to decide and institute the appropriate management thereby reducing the prevalence of this condition in our population.

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