CASE REPORT

Cerebrospinal fluid Pseudocyst as an indicator for secondary hydrocephalus following myelomeningocele repair

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Abstract

We report a case of lumbosacral myelomeningocele which has been diagnosed since intrauterine. His Initial CT brain showed no associated hydrocephalus. The patient underwent myelomeningocele repair when he was one month old following that he was quite well initially. one month later the patient started to have soft swelling at the site of old repair which started to progress in size over a week and then became constant in size. When another CT brain was done secondary hydrocephalus was discovered. Shunt operation was performed. Although the shunt was functioning but the lumbar cyst remained constant in size. After six weeks of observation the cyst remained constant in size. The patient was re-operated by removal of the cyst repairing the defect successfully.

Keywords: Myelomeningocele, Hydrocephalus, CSF pseudocyst.

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Introduction:

The incidence of hydrocephalus requiring shunts in children with myelomeningocele (MM) is reported to be very high.[1] Many authors discussed how, when and in whoma shunt is needed. Frequently patients with myelomeningocele without associated hydrocephalus who develop secondary hydrocephalus after myelomeningocele repair can be observed.[2] Features in those patients that make the suspicion of secondary hydrocephalus post-repair should be found out.[3]

Although the incidence of CSF pseudocyst at the site of old repair is extremely rare, but still it can be used as one of the features to suspect secondary hydrocephalus post-repair. In the literatureonly one similar case reported by Takahashi, Y. in which the pseudocyst occurred following shunt dysfunction, the revision of the shunt system failed to reduce the size of the pseudocyst, then the cyst was successfully directly repaired.[4]

Case Report:

Six months old male diagnosed as having myelomeningocele since intrauterine life. The mother delivered the child by elective cesarean section, term, cried and breastfed immediately. The patient has been transferred to us for the management of his myelomeningocele. CT brain requested which showed no associated hydrocephalus. The patient then underwent myelomeningocele repair in 5.12.2011. The postoperative course passed smooth and uneventful.

One month later the patient started to have soft tissue swelling at the site of the repair (figure-1) which increased in size initially over one month then became constant. A new CT brain was requested (figure-2) which showed secondary developed hydrocephalus for which a medium pressure ventriculoperitoneal shunt BMI device system was insertedin 18.3.2012. The patient showed signs of clinical improvement postoperatively (figure-3).

After six weeks the cystic swelling was not changing in size. A lumbosacral spine MRI was then requested.A well circumscribed cyst (figure-4a)

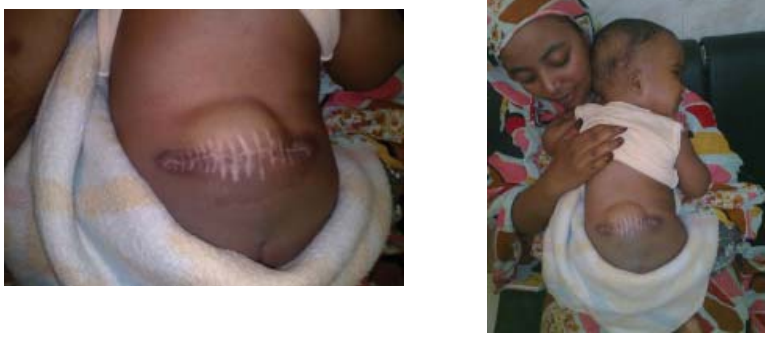

Figure. 1: Pre-operative photographs show the cystic swelling at the site of old repair

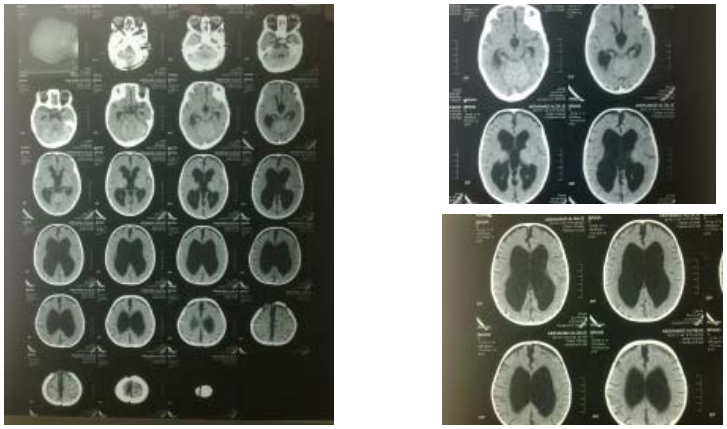

Figure. 2: The CT brain showing the dilated ventricles together with ballooning of the third ventricle

Figure. 3: The child in the first postoperative day of ventriculoperotineal shunt insertion

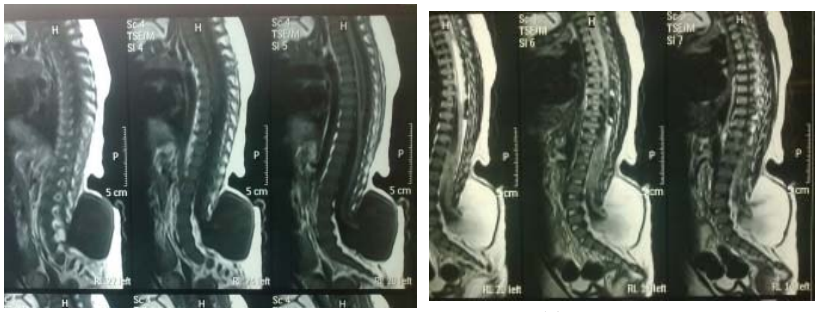

Figure. 4: T1 WI shows the thick walled cyst with tethered cord (a). T2 WI showed different density CSF (b).

which was continuous with the subarachnoid space around the cord but with different intensity of the intracystic fluid (figure-4b) together with features of tethered cord was found

The patient was operated by surgical removal of the cyst, detethering of the hanging cord and nerve roots and then anatomical repair of the defect. The post-operative course passed uneventful and the patient discharged in the third post-operative day in a good condition.

Discussion:

Hydrocephalus might associate myelomeningocele whether before or after surgical closure of the defect that might requires the placement of a ventricular shunt to prevent additional cerebral damage.[5] The adventure of the shunt to be used for the treatment of hydrocephalus in myelomeningocele cases has aided too much in the reduction of the overall mortality and morbidity in myelomeningocele cases.[6]

Timing of shunt insertion in infants with myelomeningocele and hydrocephalus has been debated.[7] However, some prefer repairing the MM then observe for the secondary hydrocephalus[8,9] while others prefer shunt insertion before or simultaneous with MM repair.[10]

In our center, we used to do the shunt operation first for the associated hydrocephalus then schedule the patient for myelomeningocele operation three to six weeks later. This is because that we think hydrocephalus treatment is more important than myelomeningocele repair. That is because preventing further cerebral damage has a priority on repairing an irreversible spinal nerves problem. Also, we have noted that placing the V.P. shunt before repair makes the repair much easier with less tension and complications of the wound like wound dehiscence and CSF leakage or fistula post-repair. However, we still frequently see patients with myelomeningocele without associated hydrocephalus who develop secondary hydrocephalus after myelomeningocele repair.

Although the incidence of CSF pseudocyst at the site of old repair is extremely rare, but still it can be used as one of the features to suspect secondary hydrocephalus post-repair as in this case.

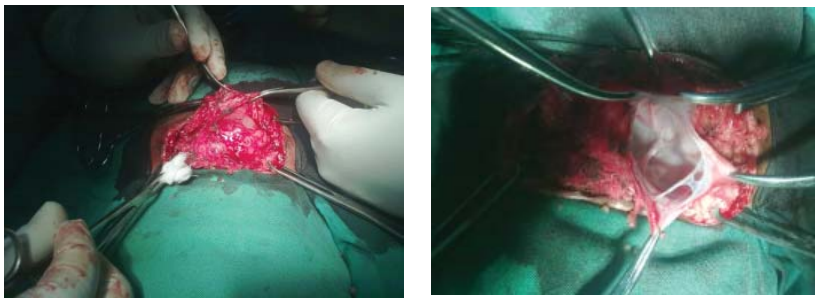

Figure 5: Intra-operative photographs showing the multilayers covering the cyst (a) and the tethered neural tissue after exploring the cyst (b).

We reviewed the literature and we have identified only one similar case report from Japan published by Takahashi, Y. in which the pseudocyst occurred as a result of shunt dysfunction and this support the postulation that this CSF pseudocyst can be used as an early sign to suspect secondary developed hydrocephalus.

The occurrence of pseudocyst in the current case report can be explained by the increased intracranial pressure created by closing of the releasing point at the defect site leading to the secondary developed hydrocephalus which in turn exert tension at repair site causing secondary cyst at that site. Associated chiari malformation could not be excluded.

The repair of myelomeningocele should be done in the classic three layers anatomical repair which are the dura, the facia and finally the skin.[6,11] Secondary developed hydrocephalus after myelomeningocele repair is a raw area that needs to be investigated and further studies need to be done to find out the other early signs of secondary hydrocephalus in myelomeningocele patients post-repair. The placement of the shunt is not enough to treat the pseudocyst and surgical removal of the pseudocyst together with re-repair of the defect is advised.

Conclusion:
we conclude that cerebrospinal fluid pseudocyst as an indicator for secondary hydrocephalus following myelomeningocele repair.

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Role and contribution of authors:
Dr. Mohammed Awad Elzain, collected the data, references and wrote the menuscript

Dr. Abu bakr Darrag Salim, critically review the article and made final changes

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