Clinics in Surgery

Case Report
Published: 07 Mar, 2017

Long Term Follow-Up of Prenatal Diagnosed Isolated Giant Anterior Sacral Meningocele (ASM)

Enaam Raboei, Alaa Ghallab, Ameen Al Saggaf, Ali Zein Elabdeen and Yazeed Owiwi*

Department of Pediatric Surgery, King Fahad Armed Forces Hospital, Saudi Arabia

Abstract

Background: ASM is a rare congenital disorder. Usually it presents sporadically. A review of literatures indicates that no neurological consequence in comparison to posterior meningomyelocele (PMM). We present a case of ASM diagnosed prenatally and followed up for 14 years. To our knowledge this is the first published case of ASM which developed ESRD from neurogenic bladder. Management course and review of the available literatures on the disease will be presented.

Method: Fetal pre sacral cyst was diagnosed in a 46 years old mother. A normal 3 kg baby girl with isolated giant ASM was followed up post natally, laparotomy and surgical removal of the sac through anterior abdominal approach was done. MRI lumbo-sacral spine was carried out for 16 available family members.

Results: The family screening was negative. Generally good surgical outcome. Our patient ended with ESRD on hemodialysis with long term follow up for 14years.

Conclusions: Surgical management of ASM through anterior abdominal approach is safe in immediate post natal period. The isolated ASM is sporadic and the family screening is not needed. Neurogenic bladder should be addressed with long term follow up of such cases. Treating physicians should take care of bladder dysfunction especially if the patient started to have urinary incontinence.

Keywords: ASM; Giant; Anterior; Neurogenic bladder; Sacral; Meningocele; Inheritance

OPEN ACCESS

Correspondence:
Enaam Raboei, Department of Pediatric Surgery, King Fahad Armed Forces Hospital, PO Box 9862, Jeddah 21159, Saudi Arabia, Tel: +966-2-665-3000 Ext 3038, +966505607930;
E-mail: enaamraboei@yahoo.fr

Received Date: *05 Jan 2017*
Accepted Date: *27 Feb 2017*
Published Date: *07 Mar 2017*

Citation:
Raboei E, Ghallab A, Al Saggaf A, Elabdeen AZ, Owiwi Y. Long Term Follow-Up of Prenatal Diagnosed Isolated Giant Anterior Sacral Meningocele (ASM). Clin Surg. 2017; 2: 1355.

Introduction

Anterior sacral meningocele (ASM) is a rare congenital malformation. Though the incidence of posterior meningocele/myelomeningocele is about 1-2/1000 live births [1-6], only 227 cases of complete or incomplete Currarino syndrome [7] with ASM were reported up to 2000 in English literature. However, the real incidence of the malformation is unknown, as it may remain asymptomatic in many instances. Only 14 cases of anterior sacral meningocele in pregnant mother were reported in the literature [8]. Cesarean section is recommended as the only mode of delivery due to three deaths (22%) during labor as a result of meningeal sac rupture [6]. ASM usually occurs sporadically. Nevertheless, familial cases have been described and generally thought to be autosomal dominant inheritance. Some authors, however, have suggested a sex-linked dominant trait. In comparison to PMM, neurogenic bladder is not a recognized association.

Case Presentation

A 46-year-old woman, G17 P14+3, 14 full term SVD, one still birth and 2 abortions. She has 8 boys and 6 living girls. She was referred for a routine ultrasonic evaluation at 29 gestational weeks, a breach fetus with a pelvic cystic mass measuring 3.3X3.5 cm observed positioned between the sacral spine and the bladder. The mass displaced the bladder and seemed to be a part of it as it was unclear to the obstetrician at this stage. Ureterocele, intra vesical septum or persistent cloaca were suspected at this point of the examination (Figure 1). At 32 weeks gestation, after a complete emptying of the bladder, the cystic mass was identified as a separate mass localized at the anterior aspect of the sacral spine. The cyst measured 5.5x6.3 cm (Figure 2). It has a round shape at its anterior part and doesn't show a digit like projection as it approached the ossification center of the vertebra. This hypoechogenic tubular prolongation was seen in continuity with the spinal canal in the longitudinal section (Figure 3) of post natal MRI. The bladder manifested good function with regular filling and emptying. No other structural abnormality was noted excluding bilateral hydronephrosis more on the left side measuring 12 mm in the antero- posterior diameter. The karyotype was normal. She was counseled in the earlier pregnancy about the various diagnostic possibilities with special

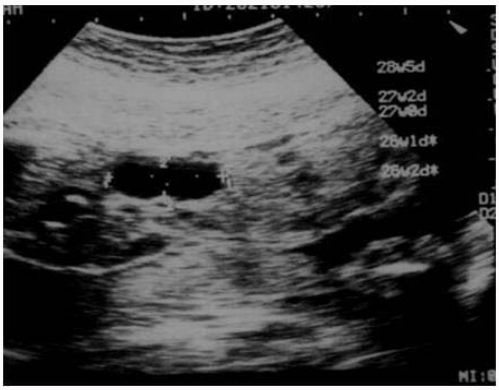

Figure 1: US at 29 weeks gestation.

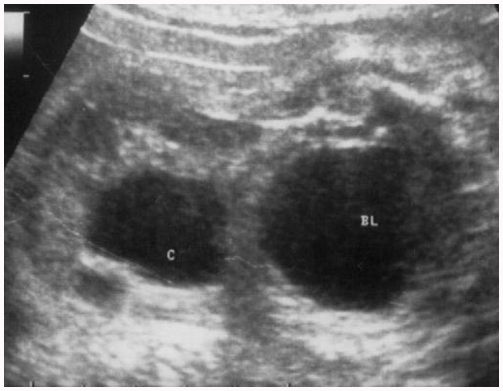

Figure 2: US at 32 weeks gestation, after a complete emptying of the bladder.

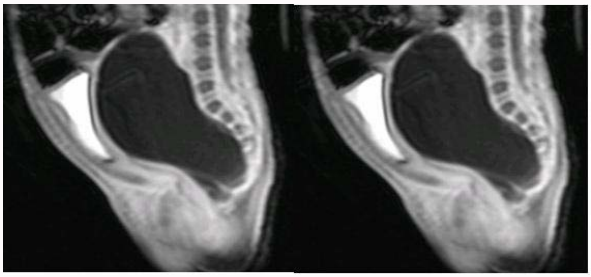

Figure 3: MRI post natal, hypoechogenic tubular prolongation was seen in contiguity with the spinal canal in the longitudinal section.

attention to the unpredictable prognosis concerning the possible of Currarino syndrome. She decided not to terminate pregnancy. An elective caesarean section gave a normal baby girl weighed 3.3 kg with no morphological anomalies. The Ultrasound identified a large cystic mass in the pelvis extending cranially from the lower poles of the both kidneys down to the pelvic floor measuring about 5.5x6.3 cm with no calcification and no obvious significant contrast enhancement with dilatation of the left ureter at the proximal and middle aspect due to mass compression effect on the distal part associated with moderate hydronephrosis of the left kidney on MCUG. Ultrasound was limited in visualizing the stalk. As CT with contrast enhancement has limited diagnostic capacity in visualization of the stalk, MRI was the diagnostic method of choice [9]. The baby was operated at 7 days of age through abdominal approach where the sac was completely excised. The baby went home at 5th day post-operative. Histology of the sac proved the diagnosis. The baby was followed regularly in the clinic and the hydronephrosis disappeared post operative. Three

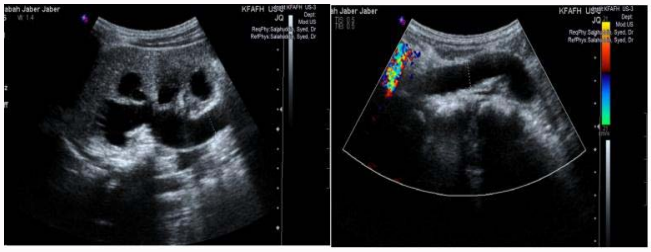

Figure 4: US showed hydroureteronephrosis.

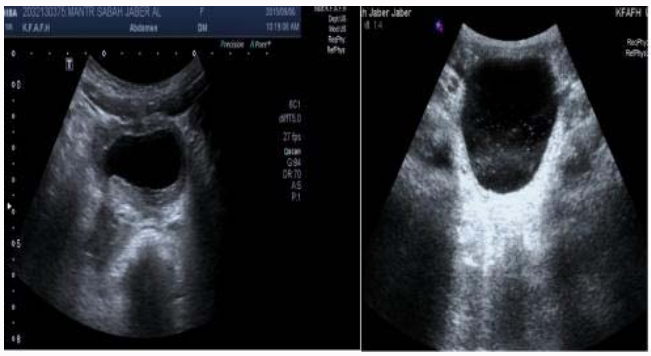

Figure 5: US showed thick wall bladder.

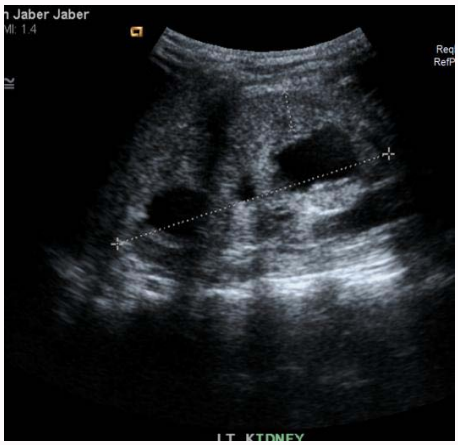

Figure 6: US showed post op improvement.

year later, with no recurrence of the cyst with mild symptoms of bladder irritability. The child was started on oxybutynin, laxative and CIC. The families were not compliant because of social problem and disappeared for 6 years. At age of 9 years, the patient presented with increased hydro-ureteronephrosis and thickened wall bladder (Figure 4 and 5), incontinence, repeated UTI and mild renal failure (creatinine values Table 1) Mitrofanoff was done for her and she was improved clinically, laboratory and radiologically Figure 5. The patient improved clinically and radiologically on follow up Figure 6. The patient's care was transferred to nephrologist at age of 12 years. The patient and family were not compliant for CIC nor for CRF medical management. Her condition deteriorated and progressed to ESRD and started on hemodialysis at age of 14 years.

Discussion

ASM can be described as intramural, pelvic, abdominal-pelvic, pelvic-sciatic and mixed anterior and posterior. ASM is usually classified among the neurulation defects dependent on the failure

Table 1: RF on long term follow up.

RF / date	Value	Clinical Picture	Comments
12/09/2004	56		Pre op
14/09/2004	47	Post Excision ASM	Immediate post op
08/12/204	25	3/12 post op	
12/01/2012	149	Hydronephrosis, incontinence & UTI	Patient was refusing CIC from native urthra
12/04/2012		Mitrofanoff	
23/05/2013	87		
07/01/2014	154	Repeated UTI	Non compliance
28/12/2014	266		
26/04/2015	392		
08/01/2017	698	ERDS	Hemodialysis

of the neural tube to close. The pathology is due to herniation of the arachnoid membrane through a primary dural defect, resulting in pulsatile stresses that erode the bone due to the CSF pressure oscillations. The sac may become progressively larger, even reaching volumes greater than 1000 cc [10]. When symptomatic, ASM are usually discovered before the third decade of life. A female predominance has been reported, but not confirmed, in studies dealing with the pediatric population [11]. In comparison to PMMC, ASM is not usually complicated post operatively by hydrocephalus or neurogenic bladder [12]. Patients with ASM often remain asymptomatic for a long time. The non specific clinical picture may delay the diagnosis, especially in newborns and young children. Conservative management is associated with 30% mortality mostly due to meningitis. With widespread of prenatal Ultrasound screening the diagnosis of ASM might increase. Only 14 cases of anterior sacral meningocele in pregnancy were reported in the literature[14]. Three (22%) of them died due to rupture of the meningeal sac during labor [6]. Cesarean section was recommended to be considered the only mode of delivery. In our case elective caesarean section and tubal ligations were done simultaneously. In our case the post natal investigations with MRI limited the differential diagnosis between ASM and Sacrococceageal Teratoma. Surgery is the treatment of choice because there is no spontaneous regression. The posterior approach, trans-sacral or sagittal, is the preferred one because of the lower complication rate [2]. The anterior laparotomy approach noticed to be associated with high morbidity and mortality (22%) due to infections and fistula formation. For the same reason trans-vaginal and trans-rectal puncture are contraindicated. Recently an endoscopic approach was suggested as an easier and shorter alternative [1,7]. The author preferred the anterior approach because the baby was operated at 7 days old, NPO on TPN and was covered well with antibiotics. We believe this approach will allow for complete excision of the sac and ligation of the stalk to prevent recurrence. We did not counter any complications including fistula reported by others through this approach [11]. This is most probably due to early diagnosis and operating on asymptomatic patient. Our patient has neurogenic bladder manifestation and renal impairment started late on long follow up which was presented with urinary incontinence and recurrent UTI. Diagnosis was done by US, deterioration of renal function Table 1 and uroflometry. The genetic inheritance pattern of ASM has not been ascertained. An X-linked dominant pattern was proposed [8]. In most cases with family recurrence, autosomal dominant transmission is suggested. The cytogenetic defect site was reported to be linked to the HLA [4,5] in a location, 7q36, which also contain a gene for holoprosencephaly. On the other hand, Chatkupt et al. [13] could not confirm the HLA region as the location for the ASM in members of five family generation with this disorder. ASM usually occurs sporadically. Nevertheless, familial cases have been described, generally with autosomal dominant inheritance. Some authors, however, have suggested a sex-linked dominant trait. Currarino triad familial cases were reported before [8]. There are case reports in the literature in which an ASM occurred as a familial, isolated disorder (in the absence of other caudal abnormalities or syndromes). These observations may justify MRI screening of the relatives of affected subjects by some authors [13]. In our case the girl is coming from a large family which permits the screening of the immediate family members with MRI and it was negative.

Conclusion

Surgical management of ASM through anterior abdominal approach is feasible in immediate post natal period and might not carry the risk of fistula formation. The isolated ASM is sporadic and MRI screening is not recommended. Neurogenic bladder should be addressed and followed up especially in giant isolated AMC.

References

1. Clatterbuck RE, Jackman SV, Kavoussi LR, Long DM. Laparoscopic treatment of an anterior sacral meningocele (case illustration). J Neurosurg. 2000; 92: 246.
2. Otagiri N, Matsumoto Y, Yoshida Y. Posterior sagittal approach for Currarino syndrome with anterior sacral meningocele: a case report. J Pediatr Surg. 2000; 35: 1112-1114.
3. Cohn J, Bay-Nielsen E. Heredity defects of the sacrum and coccyx with anterior sacral meningocele. Acta Pediatr Scand. 1969; 58; 268-274.
4. Lynch SA, Bond PM, Copp AJ, Kirwan WO, Nour S, Balling R, et.al. A gene for autosomal dominant sacral agenesis maps to the holoprosencephaly region at 7q36. Nat Genet. 1995; 11: 93-95.
5. Chatkupt S, Speer MC, Ding Y, Thomas M, Stenroos ES, Dermody JJ, et al. Linkage analysis of a candidate locus (HLA) in autosomal domonant sacral defect with anterior meningocele. Am J Med Genet. 1994; 52: 1-4.
6. Andersen C, Tange M, Bjerre P. Anterior sacral meningocele occurring in one family. An autosomal dominantly inherited condition. Br J Neurosurg. 1990; 4: 59-62.
7. Raftopoulos C, Pierard GE, Retif C, Braude P, Brotchi J. Endoscopic cure of a giant sacral meningocele associated with Marfan's syndrome: case report. Neurosurgery. 1992; 30: 765-768.
8. Zvi Kivilevitch. Anterior sacral meningocele. 2000. 10-21-13.

9. Lee KS, Gower DJ, McWhorter JM, Albertson DA. The role of MR imaging in the diagnosis and treatment of anterior sacral meningocele. Report of two cases. J Neurosurg. 1988; 69: 628-631.

10. Galzio RJ. Giant anterior lumbosacral meningocele associated with intracranial meningiomas and multiple congenital malformations. Surg Neurol. 1982; 18: 419-424.

11. Lee SC, Chun YS, Jung SE, Park KW, Kim WK. Currarino triad: anorectal malformation, sacral bony abnormality, and presacral mass-a review of 11 cases. J Pediatr Surg. 1997; 32: 58-61.

12. Singh SJ, Rao P, Stockyon V, Resurrection L 3rd, Cummins G. Familial presacral masses: screening pitfalls. J Pediatr Surg. 2001; 36: 1841-1844.

13. Fitzpatrik MO, Taylor WA. Anterior sacral meningocele associated with a rectal fistula. Case report and review of the literature. J Neurosurg. 1999; 91: 124-127.