Spina bifida aperta in southern Iran, 15 years experience

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Abstract

Background: Spina bifida aperta is a lesion that communicates with the external environment; and includes meningomyelocele and meningocele. We investigated all clinical presentations in meningomyelocele and meningocele patients in Shiraz, southern Iran.

Methods: Patients' files of spina bifida aperta who were admitted from 1989 through 2005 to Nemazee Hospital were reviewed for meningomyelocele and meningocele in Shiraz, southern Iran. CSF shunting by ventriculoperitoneal (VP) shunt was also performed.

Results: Out of 580000 patients, 100 cases of spina bifida aperta were registered. In 67 patients with meningomyelocele and 33 with meningocele with male and lumbar and lumbosacral areas predominance, cystic mass was more common. 35% of patients showed congenital anomalies including talipes equinovarus and congenital hip dislocation that were more in meningomyelocele patients. 10% of the patients were operated in the first 72 hours and the others 1 month to 1 year after birth. CSF shunt insertion was more in meningomyelocele patients. Among meningomyelocele patients, mortality occurred in 3 patients (meningitis in 2 and post-operative pneumonia in one case).

Conclusion: It seems that water-thigh dural closure and use of dural patch could decrease the rate of postoperative CSF leakage. Our results resembled the pattern in other series, except the low prevalence of hydrocephalus and a male predominance. Earlier referral of spina bifida aperta patients should be encouraged, because it could lower the complications and lead to better outcomes.

Keywords: Spina bifida aperta; southern Iran

Introduction

Spina bifida is a group of neural tube defects involving spinal structures. Spina bifida ranges from a severe, obvious, open defect to a less likely recognizable defect. Because of its highly variable characteristics, a uniform definition of the defect is difficult to make. Spina bifida was recognized in ancient times. Peter Van Forest first recorded the describtion of a child with spina bifida in 1587,¹ and in 1610 he performed the first reported surgical procedure to ligate meningomyelocele sac.² Spina bifida aperta is a midline spinal lesion that communicates with the external environment; and includes meningomyelocele and meningocele. The survival for children with spina bifida has dramatically increased during the last 30 years.³ Doctors need reliable data on outcome in order to help parents faced with difficult decisions about termination of an affected pregnancy or treatment after birth.⁴ In children with a meningomyelocele, the spinal cord fails to fuse dorsally during primary neuralation, leaving a flat plate of neural tissues called the neural placode. In children with a meningocele, the spinal cord forms normally but the dura fails to fuse normally, creating a cystic lesion that does not directly involve neural tissue. The prevalence of meningomyelocele has declined in the past several years due to both prevention with folate supplements and pregnancy termination after prenatal diagnosis. It seems that the decline began before the

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widespread availability of prenatal diagnosis.⁵ Despite persistent efforts directed at prevention of spina bifida, the birth of children with this complex malformation is continued, and skilled and compassionate physicians are needed to cope with their unique problems.

In this study, we investigated the clinical presentations, pattern and locations of involvement, therapeutic modalities, associated anomalies, and postoperative complications in meningomyelocele and meningocele patients who were admitted to Nemazee Hospital in Shiraz, southern Iran from 1981 to 2005.

Materials and Methods

This work was a retrospective single institution study of meningomyelocele and meningocele patients who were admitted, from 1989 through 2005, to Nemazee Hospital affiliated to Shiraz University of Medical Sciences in Shiraz, southern Iran. The patients' data were collected by reviewing their files. These included clinical presentations, patterns and locations of involvement, therapeutic modalities, associated anomalies, and postoperative complications in spina bifida aperta. The basis for differentiation of meningomyelocele and meningocele was the presence or absence of neural tissue during reparative surgery or pathologic report, which indicated meningomyelocele and meningocele. The university Ethics Committee approved this study. Statistical analysis was done by SPSS software using Chi Square or Fisher Exact test. P value less than 0.05 was considered significant.

Results

Out of 580000 patients admitted to Nemazee Hospital, 100 cases of spina bifida aperta were registered. Of these, 67 (40 males, 27 females) and 33 (22 males, 11 females) cases were documented as meningomyelocele and meningocele respectively. Our data demonstrated a male predominance (62% vs 38%), a difference that was statistically significant (p=0.013). The most frequent sites of involvement were lumbar (47 cases), lumbosacral areas (25 patients) and the least frequently affected sites were cervical (3 cases) and cervicothoracic region (one patient). Spina bifida aperta found in sacral, thoracolumbar and thoracic regions were 13, 7 and 4 cases respectively.

The clinical presentations of meningomyelocele and meningocele were shown in Table 1. Cystic mass in the involved segment was the most common presentation occurring in all patients. Neurologic deficit, hydrocephalus and tethered cord syndrome were also other more common presentations. Neurologic deficit and hydrocephalus tend to occur more frequently in patients with myelomeningocele (P=0.001, P=0.014 respectively). A wide variety of congenital anomalies associated with spina bifida aperta were presented in Table 2. As a whole, 35% of spina bifida patients showed some kinds of congenital anomalies. Associated anomalies were higher in meningomyelocele patients compared with meningocele (45% vs 12%, P=0.003). The most frequent anomalies were congenital talipes equinovarus (clubfoot) and congenital hip dislocation (CHD), which occurred in 28% and 6% respectively. Interestingly, clubfoot and CHD in most records were associated with normal neurologic examination without associated sensory or motor deficit. Based on the timing of definitive operation and for better evaluation, children were divided into five groups of birth-1 day, 1-3days, 4 days-1 month, 1 monthlyear, and older than 1 year (Table 3). As demonstrated, only 10% of the patients were operated in the first 72 hours of life and in most patients, operations were performed 1 month to 1 year after birth. A variety of surgical procedures including laminectomy, water-thigh dural closure, release of tethered spinal cord, dural patch, and shunting were carried out to treat patients. Orthopedic and urologic procedures were conducted in 19% and 21% of the patients respectively. CSF shunting, mostly by ventriculoperitoneal (VP) shunt was performed in 25 patients (Table 4). In most cases (73%), CSF shunting was done prior to operation as a result of CSF leak. In these cases, decreased CSF leak after shunting, subsequently allowed a definitive operation. However, there was no case of simultaneous shunting and definitive operation. Obviously because most cases of hydrocephalus occurred in meningomyelocele patients, CSF shunt insertion was also more frequent in these patients, compared to those with meningocele (P=0.013). CSF leakage was observed in 30% and 13% of myelomeningocele and eningocele patients respectively while these figures for decreased neurologic function were 20% and 3%, for meningitis were 8% and 3% and for postoperative mortality were 5% and 0% respectively. All complications were more frequent in meningomyelocele patients (P=0.05), who sustained the highest mortalities. It seemed that water-thigh dural closure and use of dural patch could decrease the rate of postoperative CSF leakage (P=0.05). CSF leakage was also observed in all 6 patients who had meningitis. Postoperative mortality in our study was 3 (3%), all occurring in meningomyelocele patients with 2 cases due to meningitis and one as a result of post-operative pneumonia.

Tabl	e 1:	Clinical	presentations	of	spina	bifida	ap-
erta	patie	ents					

Disease	Myelo ingo- cele(6	omen- 67)	Meningo- cele(33)	
Clinical presentation Neurologic deficit	No.	%	No.	%
Bilateral	41	61	1	3
Unilateral	10	15	0	0
Meningitis	12	18	6	18
CSF leakage	14	21	5	15
Hydrocephalus	23	34	3	9
Tethered cord syndrome	20	30	4	12
Arnold-Chiari typell malformation	5	7	0	0
Trichosis	0	0	1	3
Cystic mass	67	100	33	100

Table 2: Associated malformations in spina bifida

 aperta patients

Disease		Myel ingo (67)	lomen- cele	Meningo- cele(33)		
Malformation		No.	%	No.	%	
Clubfoot	Bilateral	14	21	0	0	
Clubiool	Unilateral	12	18	2	6	
CHD	Bilateral	4	6	0	0	
CHD	Unilateral	2	3	0	0	
Microceph	aly	2	3	1	3	
Scoliosis		4	6	0	0	
Lordosis		1	2	0	0	
Situs inve	rsus	3	5	0	0	
Cleft palat	e	0	0	1	3	
Congenita	l heart	3	5	0	0	
disease						
Imperforat	te anus	1	2	0	0	
Femoral h	ernia	1	2	0	0	
Inguinal h	ernia	1	2	0	0	
Hypospad	iasis	1	2	0	0	
Craniofaci	al	1	2	0	0	
dysostosis						
Diastemat	omyelia	2	3	0	0	

Table 3: Age-distributio	n of	spina	bifida	aperta	pa-
tients who had been ope	rate	d			

Disease	Myel goce	omenin- le	Meningocele		
Operation time	No.	percent	No.	Percent	
Birth-1day	2	3	0	0	
1-3days	4	6	4	12	
4days-1month	8	12	2	6	
1month-1year	36	53	19	57	
>1year	9	13	6	18	
No operation	8	12	2	6	

*Parents of ten children did no consent for operation.

 Table 4: CSF shunting in patients with spina bifida aperta

Disease	Myelo gocele	menin- Ə	Meningocele	
CSF shunt	No.	%	No.	%
Total cases of	23	100	3	100
hydrocephalus				
requiring shunt				
Total cases of	22	96	3	100
CSF shunting				
VP shunting	20	87	1	33
VA shunting	2	9	2	66
Shunt insertion	16	70	3	100
prior to definitive				
surgery				
Shunt insertion	3	13	0	0
following surgery				
Shunt insertion	3	13	0	0
without surgery				

VP:Ventriculoperitoneal VA:Ventriculoatrial

Discussion

Over the past 30 years, continuous progress has been made in our knowledge about spina bifida patients. Nevertheless, little information is available about different aspects of this disease in Iran. We have encountered two unusual results about our patients. Firstly, percentage of meningocele more than 30%, in contrast to most cases of meningomyelocele in spina bifida reported previously.^{6,7} This relatively high percentage of meningocele patients may be due to their better prognosis urging the physicians to admit and operate them more than meningomyelocele patients. Secondly, male preponderance in spina bifida aperta patients as opposed to a slight female predominance reported in most series.^{8,9} The present study was not

population based and therefore could not provide any estimates of the prevalence of spina bifida aperta in our region. Further studies are needed to have a better insight into this disease in Iran. Regarding the site of involvement, our patients were similar to other series. The majority of meningomyeloceles were located in the caudal thoracolumbar spine or more distally. Cervical myelomeningoceles are often in reality very similar to meningoceles without associated Chiari II malformation and hydrocephalus.¹⁰ In a similar study in India, lumbosacral region followed by lumbar region were the most common sites of spina bifida.⁶ The presence of cystic mass in the involved segment of the spine was a predictable finding. In fact, meningomyelocele and meningocele are also called spina bifida cystica. The incidence of hydrocephalus in spinal dysraphism is low in Iran in comparison to western hemisphere.^{11,12} but studies from Spain, northern India and Malawi also reported a relatively low inci-dence of hydrocephalus.^{6,13,14} Spina bifida can occur as part of malformation syndromes resulting from known chromosoml abnormalities and single gene disorders.¹⁵ In addition, rare families, segregating spina bifida in patterns consistent with X-linked and autosomal recessive inheritance has also been reported.¹⁶⁻¹⁸ However, the vast majority of cases cannot be attributed to either chromosomal aberrations or the effects of a single genetic locus. Multiplicity and diversity of malforamations associated with spina bifida has advanced to a hypothesis that many congenital anomalies of non-neural organs may be produced by damage to their mesodermal or endodermal anlagen caused by overdistention of the embryonic neural tube.19,20 Orthopedic problems presenting as primary malformations or occurring secondary to neurologic deficits are very common in spina bifida aperta patients. Congenital talipes equinovarus and congenital hip dislocation were the most common anomalies in our series. Presence of these anomalies in our patients, despite a normal neurologic finding, overestimates their primary occurrence in spina bifida aperta. Significant scoliosis and congenital talipes equinovarus were the most frequent neuroorthopedic deformities in another study.⁶ Spina bifida is generally accompanied by a high incidence of foot deformities and the pattern of feet abnormalities are related to the level of spine involvement in some series.²¹ In patients with thoracic lesions, the most frequent deformity was an equinus lesion (55%), a club foot with mid-lumbar lesion (87%) and a calcaneal foot with sacral lesions (34%). We did not find any

association between the level of spine involvement and feet anomalies. Dislocation of the hip is a wellrecognized entity that frequently occurs in children with spina bifida and require surgery in most cases.²²⁻²⁴ Other orthopedic problems in our patients were scoliosis and lordosis. Occurrence of scoliosis in our series was not high, but up to 30% of spina bifida patients had a congenital scoliosis in some reports and the curve may not manifest until 5 to 10 years of age.²⁵ Inguinal hernias have been reported to increase after VP shunt procedures in pediatric patients.²⁶ Giant inguinal hernia has been reported in a 5-year-old boy with hydrocephalus.²⁷ This patient had multiple neurosurgical procedures performed in the neonatal period for spina bifida and hydrocephalus, including the placement of a VP shunt. Close observation of infants and children who undergo VP shunting is required, to allow early detection and repair of inguinal hernias. We only had one report of inguinl hernia in our patients. The association of imperforate anus and spina bifida has also been well described previously.^{28, 29} This condition was also observed in one of our patients with meningomyelocele.

Martinez-Frias claimed statistically significant association between hypospadiasis and spina bifida. This association was identified through the Spanish Collaborative Study of Congenital Malformations, where the observed number of cases among males was greater than expected.³⁰ We only had one case of hypospadiasis in a meningomyelocele patient. Spinal dysraphism with dextrocardia or situs inversus have been reported in only few cases. Successful management of this sophisticated entity requires appropriate understanding of embryology, anatomy, and imaging and has implications in neurosurgical and periopera-tive anesthetic care.^{31,32} The meningomyelocele repair can be performed safely up to 72 hours after birth, which allows time for completion of the initial evaluation.³³ Any delay after 72 hours markedly increased the chances of meningitis or ventriculitis.³ Delayed closure is associated with a 37% incidence of ventriculitis compared with 7% with early closure.³³ Unfortunately most of our patients underwent operation after the golden time of 72 hours due to delayed referral. As stated before, the hydrocephalus tended to occur less frequently in our series compared to western countries, so only about 25% of patients required CSF shunting. In most cases, CSF shunt was inserted prior to definitive operation due to CSF leakage. The delayed referral of these patients also seemed to play a role in rupture of meningeal cyst and CSF leakage. High frequency of postoperative complications in our series was predictable due to delayed operation. Cerebrospinal fluid leakage from operation site, deteriorating neurologic function, and meningitis were the most common postoperative complications. In a similar study, CSF leakage, pseudomeningocele, wound infection and meningitis were the most common postoperative complications.⁶ Postoperative mortality rate in our series was 5% in meningomyelocele and 3% in general. The mortality rate was acceptable and comparable with other series.⁶ The one-year survival among populations referred to major medical centers has generally been close to 90%.¹² Nowadays, more than 95% of infants born with a meningomyelocele will survive the first 2 years; but unfortunately; despite intensive care, 10% to 15% of children with spina bifida will

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eventually die before the age of 6 years.^{12,33}

Clinical presentations, involvement sites, therapeutic modalities, associated anomalies, and postoperative complications in patients with f spina bifida aperta in southern Iran was comparable to other series except the unexpectedly low incidence of hydrocephalus and a male predominance. Earlier referral of spina bifida aperta patients should be encouraged because it could lower the complications and lead to better outcomes.

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