Anterior Sacral Meningocele:

Report of Five Cases and Review of the Literature*

MATATIAHU OREN, M.D., BENNETT LORBER, M.D., SEUNG H. LEE, M.D., RAYMOND C. TRUEX, JR., M.D., ANTHONY R. GENNARO, M.D.

Section of Colon and Rectal Surgery, Department of Surgery and Departments of Medicine, Radiology and Neurosurgery, Temple University Health Sciences Center, Philadelphia, Pennsylvania

Beware, oh surgeons, lest you open anything improperly which may easily kill a person.— Nicolas Tulp, 1652[14]

ANTERIOR SACRAL MENINGOCELE (ASM) is a herniation of a dural sac through a defect in the anterior surface of the sacrum. The sac is composed of an outer dural membrane, and an inner arachnoid membrane, and contains cerebrospinal fluid (CSF). Rarely, it may contain neural elements, and it is then called a myelomeningocele. Herniation of the meninges through bony defects most often occurs posteriorly in the lumbosacral area. Less frequently, it may occur along the thoracic or cervical spine, the anterior surface of the sacrum, or through defects in the facial bones or cranial vault.

In 1837, a "distinguished surgeon," who preferred to remain anonymous,[9] reported the first case of ASM, which led to obstructed labor and maternal death. Coller and Jackson[14] collected 23 cases reported earlier than 1943, and added another. Eder[19] reviewed 45 cases in 1949, Silvis *et al.*,[73] 51 cases in 1956, and Haddad[30] wrote an extensive review of 53 cases in 1958, including two of his own. The problem of ASM was also reviewed in the French literature by Thierry *et al.*[78] who discussed 67 cases in 1969, but ASM has not been reviewed in English-language medical literature in 18 years.

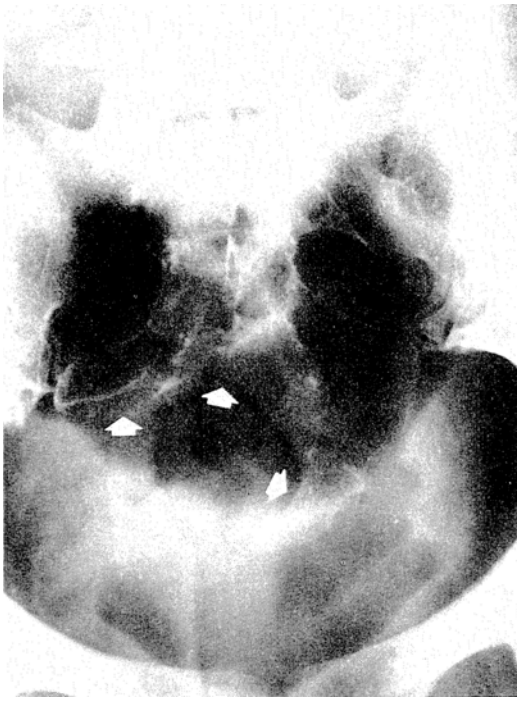

FIG. 1 (Patient 1). Roentgenogram of the sacrum showing a large smooth-bordered defect of the right side. This so-called "scimitar sign" is characteristic of congenital anterior sacral meningocele.

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Address reprint requests to Dr. Lorber: Department of Medicine; Temple University, Health Sciences Center, Philadelphia, Pennsylvania 19140.

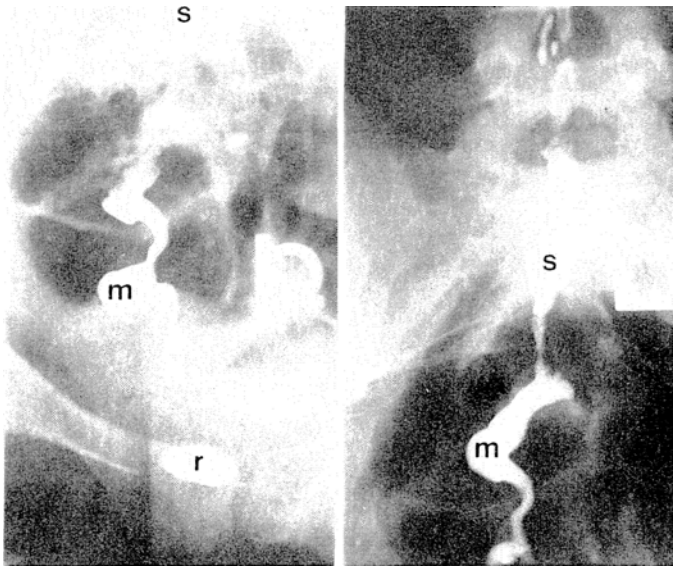

Fig. 2. Two views of lumbar myelogram (Patient 1), showing contrast material in the anterior meningocele (m) with connections superiorly to the normal sacral subarachnoid space (s) and inferiorly to the rectum (r).

Anterior sacral meningocele is rare, and its varied manifestations may bring patients to the attention of a wide range of specialists, including urologists, proctologists, gynecologists, pediatric surgeons, neurosurgeons, orthopedic surgeons, and general surgeons.[10] When it is properly diagnosed and managed, the cure rate is very high; otherwise, central nervous system complications may result in high mortality and morbidity rates.

In the recent past we have seen five cases of ASM, suggesting that the anomaly may be more common than was previously suspected.

Report of Five Cases

Patient 1: A 19-year-old white woman was admitted to the hospital on April 11, 1975, with acute purulent meningitis. For a few weeks prior to admission she had experienced headaches upon arising in the morning and more mild headaches upon standing after classes. Headaches also occurred following defecation and coitus. Relief from headaches was obtained by lying supine. One day prior to admission she had chills and a fever (temperature 101 F). The next evening, following coitus, she had an extremely severe occipital headache and stiff neck, and was brought to the hospital.

Temperature was 103.6 F, the neck was stiff, mild papilledema was evident in the right eye, and the rest of the physical examination was unremarkable. Lumbar puncture yielded grossly turbid fluid under a pressure of 30 cm of H_2O which contained 3,700 leukocytes/mm^3 with 100 per cent neutrophils, protein 1,047 mg/100 ml, and sugar 5 mg/100 ml. Spinal fluid culture grew *Bacteroides fragilis, Peptostreptococcus asacharrolyticus,* and alpha-hemolytic streptococcus. Blood cultures grew *Bacteroides fragilis, Gaffkya anaerobia, Peptococcus prevotti, Propionibacterium acnes,* and *Peptostreptococcus anaerobius.*

Past medical history was significant in that the patient had been hospitalized at 11 days of age for diarrhea and dehydration. Rectal examination at that time had shown a membrane, 1–2 cm above the anal sphincter, through the center of which was a pin-head-sized opening. This stricture was dilated daily for eight days. The patient's parents reported that throughout her childhood she had had bowel movements every five to six days, and passed stools of unusually large volume and diameter.

The patient was treated with large intravenous doses of chloramphenicol. She remained awake and alert, but continued to complain of severe headache and showed signs of meningeal irritation. Urinary retention became a problem, necessitating bladder catheterization. On the fifth hospital day a posterior, bulging, non-tender mass adherent to the sacrum was felt on rectal examination. This mass, thought to be an abscess, was drained transrectally, yielding 300 ml of pus, which grew *Escherichia coli, Bacteroides fragilis, Bacteroides melaninogenicus, Peptostreptococcus anaerobius,* and diphtheroids. Following drainage, the patient's headache and fever lessened.

Repeat lumbar punctures continued to show polymicrobial flora reflecting organisms normally present in the rectum and consisting of mixed

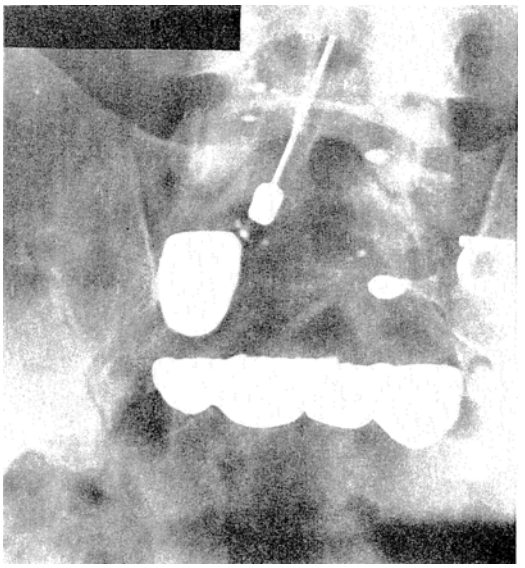

Fig. 3. Lumbar myelogram (Patient 3), showing several collections of contrast material with fluid levels within a large anterior sacral meningocele.

anaerobic and facultative organisms. A roentgenogram of the lumbosacral spine showed a "scimitar" sacrum (Fig. 1), and a myelogram showed an anterior sacral meningocele (Fig. 2). During myelography, contrast material was seen to leave the sacral spinal canal, pass through the meningocele, and enter the rectum.

Following myelography, the patient was taken to the operating room. A midline abdominal incision was made and the retrorectal space developed. In the midline, 10 cm below the sacral promontory, a meningocele stalk, 3 mm in diameter, was found bulging from the sacrum and communicating with the retrorectal cyst previously drained. The stalk was transfixed and severed and a complete diverting sigmoid colostomy and mucous fistula were brought out.

The postoperative course was complicated by urinary retention and development of a secondary *Candida albicans* meningitis, which was successfully treated by administration of 5-flucytosine for nine weeks. As a consequence of the *Candida* meningitis, the patient had severe arachnoiditis resulting in left hemiparesis. Computed tomography of the head demonstrated a communicating hydrocephalus with enlargement of the lateral and third ventricles and a normal fourth ventricle.

The patient was discharged on August 31, 1975, but returned six days later with evidence of markedly increased intracranial pressure due to obstructive hydrocephalus. A ventriculo-jugular shunt procedure was performed. Placement of the shunt was followed by alleviation of symptoms, and the patient showed a marked lessening of left-sided weakness, which has continued to the present. She underwent successful colostomy closure in May 1976.

Patient 2: A 49-year-old white woman was admitted to the hospital on February 2, 1975, for evaluation of a "lump" in her "tail." This mass had been recognized since her childhood, and had remained unchanged and asymptomatic until 17 years prior to admission, when the patient had a cesarean section. Subsequently, it had been noticed to be slowly increasing in size.

Several months prior to admission the patient noticed tenderness in the perineum and thought the mass had further enlarged. During this period she became constipated and started to use laxatives. During periods of constipation she had headaches for a short time before evacuation of the stools. She also noted impairment of rectal continence and occasionally, upon arising in the morning, lost some stool while experiencing an urge to defecate. During this period she also experienced occasional stress incontinence of urine.

Physical examination revealed a soft, cystic, 5 × 5-cm subcutaneous mass. It was visible as a bulge under the skin posterior to the anal orifice, and was palpable on rectal examination, partly between the posterior rectal wall and the skin, and partly anterior to the sacrum. It was tender and, when manipulated, there was pain radiating to the left leg. Rectal sphincter tone was diminished. The sacrum was felt to be shorter than usual, and the coccyx could not be palpated. Sigmoidoscopy disclosed no abdormality. Results of further neurologic examination were normal. Roentgenograms of the pelvis showed a typical "scimitar" deformity of the sacrum. It was decided to follow the mass clinically and to defer elective surgical treatment.

Patient 3: A 22-year-old white man was admitted to the hospital on May 21, 1975, for evaluation of low-back pain. Two years before this admission he had been in an automobile accident in which he sustained trauma to the left chest. Several weeks later he had noticed the onset of low-back pain, mainly on the left side. The pain was present while he was standing or sitting, and was relieved when he was lying down. There was no change in his bladder or bowel habits.

Past medical history revealed surgical treatment of pectus excavatum in childhood. Physical examination revealed marked left dorsolumbar convexity of the spine, and neurologic examination disclosed no abnormality. A roentgenogram of the pelvis showed a large midline sacral defect. A myelogram (Fig. 3) showed a large anterior sacral meningocele. The patient's physician advised against elective surgical treatment.

Patient 4: A 22-year-old white woman with known neurofibromatosis was admitted to the hospital in

April 1965 for evaluation of severe low-back pain. There was no constipation or urinary symptom.

Results of pelvic examination were normal. Rectal digital examination and sigmoidoscopy to 25 cm revealed no abnormality. The roentgenogram of the thoracolumbar spine was normal, but a roentgenogram of the sacrum demonstrated a large midline defect of the upper sacrum with enlarged sacral foramina on the right. A myelogram demonstrated a moderate-sized smooth collection of contrast medium anterior to the sacrum, representing an anterior sacral meningocele (Fig. 4).

Patient 5: A 4-month-old baby girl was admitted to the hospital on December 28, 1975, with symptoms of regurgitation of food and green watery stools. An aunt had previously been operated upon for a sacrococcygeal teratoma.

Physical examination revealed a normocephalic child with dimpling and erythema of the left buttock. The bony prominences of the sacrum were evident only on the right. Rectal examination revealed an extremely narrow anal orifice which would not admit a finger tip. No rectal mass was palpable. There were no sensory, motor or reflex deficits in the perineum or lower extremities.

Barium-enema examination demonstrated a mass between the rectum and sacrum. The sacrum was abnormal, with a "scimitar" configuration.

On December 29, 1975, a sacrococcygeal teratoma was excised. During the dissection, the coccyx was excised to facilitate exposure. When this was done, a flow into the wound of clear, colorless fluid, felt to be cerebrospinal fluid, was noticed and neurosurgical consultation obtained. The anterior portion of the bony sacrum was absent, with protrusion of meninges anteriorly into the pelvis. The dural sac extended caudally into the sacrum and contained an enlarged filum terminale. An extensive dural laceration was repaired with paraspinous fascia to afford watertight closure.

A postoperative myelogram failed to show the anterior meningocele found at operation. Tethering of the conus due to a thickened filum terminale was documented radiologically.

Postoperative urogram and cystourethrogram revealed a flaccid neurogenic bladder with left hydroureter and hydronephrosis. No postoperative neurologic deficit was found in the lower extremities.

Pathogenesis

Most anterior sacral meningoceles are congenital, as evidenced by the age distribution, associated anomalies, and familial incidence. Unlike the more common posterior lumbosacral meningocele, the ASM is concealed in the pelvis, and its diagnosis may be delayed until symptoms appear in later life as the result of enlargement of the cyst.[30]

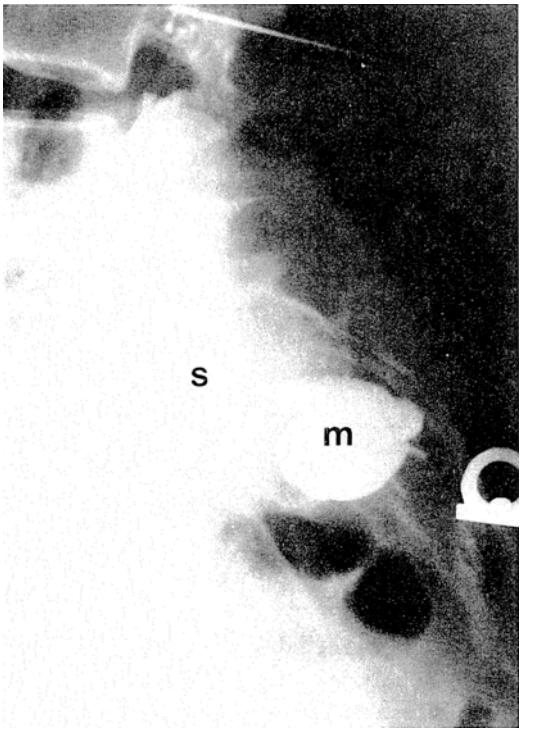

FIG. 4. Lateral view of lumbar myelogram (Patient 4), showing contrast material extending from the distal end of the sacral subarachnoid space (s) to the anterior sacral meningocele (m).

Cohn and Bay-Nielsen,[13] based on data of Cramer[15] and others, thought the pathogenesis was related to reduced permeability of the roof of the fourth ventricle at a certain stage of embryonic life. This would cause the pressure of the CSF to rise, resulting in defective closure of the neural tube, forming meningoceles anteriorly and posteriorly. This theory correlates well with the frequency of hydrocephalus seen in association with posterior meningoceles, but not with ASM where hydrocephalus has not been found, and not with the common anomalies of duplication of the uterus and vagina or anorectal anomalies seen in these cases.

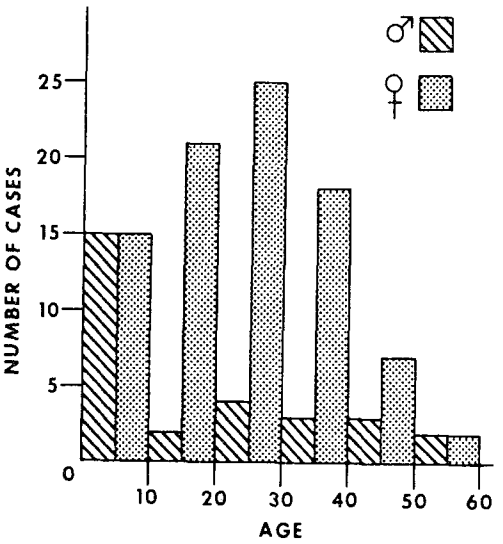

FIG. 5. Anterior sacral meningocele: age by decade and sex distribution at the time of diagnosis.

The "dysraphic" theory of partial or complete agenesis of sacral segments is another possibility presented by Amacher *et al.*[4] as a factor leading to the formation of an ASM. However, these theories are mainly speculative.

Acquired ASM has been reported in very few instances. Strand presented one case of ASM in association with Marfan's syndrome,[76] where it was considered the result of dural ectasia and transient elevation of spinal fluid pressure. Our Patient 4 had enlarged sacral foramina as a result of von Recklinghausen's disease, through which the ASM emerged.

Epidemiology

Since 1837, 120 cases for which sufficient data are available for inclusion in this report have been reported. A few cases included in previous reviews have been omitted because of insufficient data.[7, 54] Five additional cases of ASM seen by us are described above. All patients have been Caucasian. Of 125 patients, 94 were female, 30 male, and one was an infant of unstated sex.

Distribution by age and sex is shown in Figure 5. The female preponderance is obvious, and is most evident during the childbearing ages. This can only partly be explained by symptoms of dysmenorrhea, dyspareunia, or dystocia, leading to pelvic examination revealing a retrorectal mass. Only 19 female patients were diagnosed as having ASM primarily because of gynecologic or obstetric problems.[9, 13, 16, 18, 19, 29, 30, 34, 36–39, 47, 52, 69, 79, 85, 87, 88] In the younger and older age groups the incidences of ASM in the two sexes are almost equal. So, it is not clear whether females are more affected. We share the view of Silvis *et al.*[73] that innumerable cases in the male go undetected.

Aaronson[1] described a family with three siblings who were born with covered anuses; one had a duplication cyst at the anal canal, and one had as ASM. Klenerman and Merrick[43] described a family in which one girl had an ASM. Sacral deformities were found in both her father and her uncle, who were asymptomatic; no further investigation was made to confirm meningoceles. Cohn and Bay-Nielsen[13] described six familial cases of partial absence of the sacrum; four of the affected individuals had ASM and three had congenital anal stenosis. Kenefick[41] described four generations of a family of which nine members had sacral deformities. Of these, four females had presacral masses – one ASM, one dermoid cyst, and two of unknown etiology. He suggested sex-linked dominant transmission. The brother of another patient[73] with ASM had a presacral dermoid.

Clinical Manifestations

Many of the histories were not fully detailed, but the major symptoms and signs are summarized in Table 1. As reported by Brown and Powell,[8] most clinical manifestations are caused by pressure of the ASM on adjacent structures such as the rectum, urinary bladder, female genital organs, and

TABLE 1. *Clinical Manifestations of Anterior Sacral Meningocele*

Symptom or Sign	Number of Cases	References
Asymptomatic	8	4, 29, 38, 46, 65, 72, 77, 80
Constipation	54	2, 4, 6, 9, 10, 11, 13, 14, 15, 16, 19, 21, 22, 26, 33, 35, 36, 37, 38, 39, 40, 42, 43, 44, 46, 49, 51, 56, 58, 60, 61, 64, 65, 67, 70, 71, 72, 73, 76, 81, 85, 89, present case 1
Pelvic mass	Few exceptions	
Abdominal mass	19	3, 4, 11, 15, 20, 27, 32, 38, 44, 45, 57, 63, 68, 74, 78, 80, 81, 83, 90
Abdominal pain	9	10, 15, 24, 32, 44, 50, 69, 78, 85
Urinary symptoms		
Retention	11	4, 6, 11, 12, 15, 22, 41, 44, 49, 65, 68
Incontinence	11	4, 8, 13, 21, 23, 26, 40, 49, 64, 72, 84
Frequency	7	26, 28, 45, 46, 64, 72
Infection	9	8, 10, 13, 40, 46, 85
Dysuria	15	4, 12, 24, 26, 30, 35, 36, 37, 38, 39, 46, 73, 85
Low-back pain	12	4, 21, 23, 24, 27, 44, 46, 59, 79, 85
Low-back pain radiating to legs	15	2, 8, 15, 16, 22, 30, 36, 53, 58, 71, 72, 76, 89, present cases 2, 3
Hypalgesia of perineum	14	4, 8, 11, 26, 36, 37, 41, 43, 53, 67, 72, 89
Muscular weakness of legs	7	4, 13, 21, 37, 67, 84, 90
Reduced anal sphincter tone	8	4, 8, 21, 43, 72, present cases 1, 2
Fecal incontinence	6	4, 8, 13, 40, 53
Dystocia	15	8, 9, 12, 13, 16, 23, 30, 34, 38, 52, 65, 71, 88, present case 2
Dysmenorrhea	12	8, 19, 30, 39, 43, 49, 56, 69, 72, 79, 85, 87
Dyspareunia	2	36, 37
Headache	15	8, 10, 36, 38, 43, 44, 53, 59, 71, 73, 76, 85, 89, present cases 1 and 2

sacral nerve roots. Pressure on the meningocele, on the other hand, will increase the intracranial pressure and cause headache.

In most of the asymptomatic cases the patients were female, and the ASM was detected on routine pelvic examination.

Symptoms

Constipation was found to be a constant symptom when bowel habits were questioned. Many of the patients used laxatives and even daily enemas. Because of the retrorectal mass the rectum cannot fill with feces and initiate physiologic evacuation. Constipation usually begins in childhood, and there is a tendency for it to become more severe as the ASM expands.[30] Our Patient 1 and Haddad's patient[30] had had bowel movements only every five to six days since childhood, and the stools were of such enormous caliber that passing each one became a traumatic episode.

Urinary difficulties were variable. Some of the urinary symptoms may be explained on the basis of displacement of the bladder anteriorly and cephalad by the ASM; others, mainly by incontinence and retention, as a result of pressure on the sacral nerve roots, or secondary to congenital defects in the nerve supply to the bladder. Our Patient 1 had urinary retention preoperatively, which continued as a problem for several weeks.

Her cystogram showed a neurogenic bladder pattern. This problem was documented in five other cases.[2, 16, 25, 46, 78] Three other patients had enuresis until late childhood.[13, 27, 74]

Low-back pain was felt in the midline at the level of the sacrum and coccyx, and in some patients radiated to the inner aspects of the thighs. Pain was sometimes acute and disabling, sometimes chronic. The pain may be differentiated from more common causes of low-back pain by its lower location and by a history of pain following long periods of standing, sitting, or constipation, relieved by a bowel movement or a period of bed rest (our Patients 1 and 3). Pressure on sacral nerve roots may cause perineal anesthesia or hypalgesia, depending on the sacral dermatome affected. In some cases fecal incontinence occurred and in others weakness of the sphincter tone was found.

Headaches are produced when pressure applied to the meningocele displaces the CSF through the stalk and increases the intracranial pressure suddenly, as was shown by Rowlands.[65] The typical history begins in childhood, with sudden headaches during periods of straining, *e.g.*, defecation or squatting. In later life headache may accompany coitus, as seen in our Patient1 and others.[65] During examination of infants, when digital pressure is applied over the ASM, bulging of the fontanelles may be seen[68]; in others, digital examination produced headache.[72]

Meningitis as a presenting symptom was found only in our Patient 1. We can assume that microperforations in the rectal wall occurred with passage of bacteria to the meningocele itself or that a retrorectal or high intermuscular abscess was formed and communicated with the ASM to cause meningitis. Polymicrobial meningitis due to bacteria normally found in the intestinal tract should raise the possibility of infected meningocele.

Dystocia. Like any pelvic mass, ASM may interfere mechanically in the process of labor. When the meningocele is small and soft, there is a chance for normal delivery, as has been reported in four cases.[39, 71, 72] More often, labor is difficult and dangerous when ASM is present. Two women [8, 30] had stillbirths with high-forceps delivery, and needed cesarean sections to terminate other pregnancies. Cesarean sections were necessary due to obstructed labor in our Patient 2 and six other women,[16, 38, 52, 65, 71] and high-forceps delivery was necessary in three.[6, 13, 23] During the last century, when cesarean section was not a common procedure, one woman[9] died following obstructed labor, and two others[34, 88] died as a consequence of rupture of the meningocele during delivery.

Abdominal pains are usually dull, chronic, and located in the hypogastrium. A few patients had severe, acute pain and underwent emergency laparotomy.[15]

Signs

A retrorectal mass can be palpated in most instances. When the rectal examination is done the finger should carefully delineate the anterior surface of the sacrum and coccyx in order to reveal the ASM or bony anomaly. The meningocele is usually felt as a soft, cystic mass adherent to the sacrum and posterior to the rectum, but it can emerge to either side. In only a few instances was no mass palpable on rectal examination.[4, 58] This was true in our Patient 4, where the ASM emerged through a high sacral foramen. In two others the ASM had a stalk that emerged through the greater sciatic foramen to form a gluteal mass.[17, 28] Only when the ASM achieved quite large dimensions could it be palpated during abdominal examination.

A "scimitar" sacrum (Fig. 1) is almost a pathognomonic finding. A roentgenogram of the pelvis shows a defect on one side of the sacrum, as if the sacrum had developed

TABLE 2. *Congenital Anomalies and Tumors Associated with Anterior Sacral Meningocele*

	Number of Cases	References
Anomalies		
Duplication		
Uterus	10	13, 18, 24, 29, 30, 38, 43, 44, 62, 69
Uterus and vagina	5	19, 31, 36, 56, 87
Kidney, ureter	4	8, 46, 51, 74
Anal		
Stenosis	8	13, 40, 60, 70, present case 5
Atresia	2	40
Anal membrane	3	1, 64, present case 1
Rectovaginal fistula	1	42
Musculoskeletal		
Spine	5	20, 40, 50, 63, 90
Club foot	4	20, 44, 63, 72
Hypoplastic leg	1	13
Polydactyly	1	63
Petit's hernia	1	25
Large umbilical hernia	1	40
Tumors		
Tetratoma	6	3, 30, 62, 66, 83, present case 5
Tetratocarcinoma	1	25
Dermoid	6	8, 15, 24, 26, 34, 67
Lipoma	2	31, 58
Origin?	4	29, 41, 72, 86

around the cyst, acquiring the shape of the old Arabic saber. The rounded border is smooth, and there is no destruction of the remaining bone. This deformity can sometimes be palpated, as in our Patient 2. Pelvic roentgenograms of patients with ASM showed this deformity, with few exceptions.[4] ASM may occasionally be associated with a midline sacral defect (our Patient 3). As would be expected, the "scimitar" anomaly was absent in acquired cases such as those in patients who had Marfan's syndrome[76] or neurofibromatosis (our Patient 4). A few familial occurrences in which some members had ASM, and others a typical sacral deformity without documented ASM have been described.[41, 43]

Additional anomalies and congenital tumors are presented in Table 2. In the majority of cases the patients were not fully investigated by excretory urograms or careful pelvic examination to determine the coexistence of common anomalies, and small presacral tumors could go unrecognized in the absence of excisional surgical intervention.

The interrelationship among these various anomalies was not stressed in previous reviews, and is presented in Figure 6. Parts of this scheme have been investigated. Williams and Nixon[91] found that more than half of infants who had anorectal anomalies had bony malformations of the sacrum. Smith[75] reported similar findings and men-

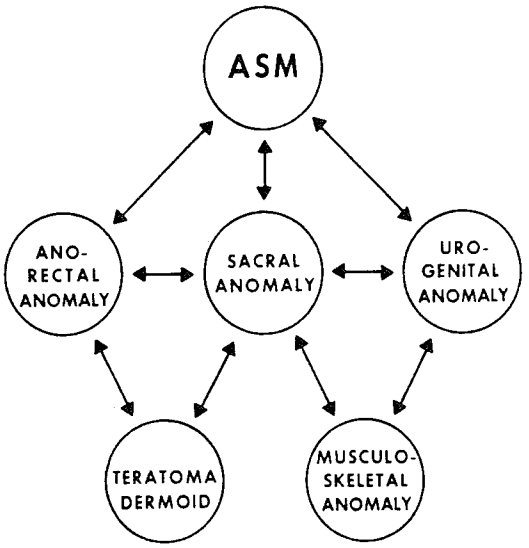

FIG. 6. Relationship of anterior sacral meningocele (ASM) to other congenital anomalies and tumors.

tioned eight urogenital anomalies and other musculoskeletal anomalies among 26 cases of anomalous sacrum. Ashcraft and Holder[5] pointed out the association of teratomas and anal stenosis.

These associations are in favor of the "dysraphic" theory of dysgenesis of the sacrum as the basic anomaly, and should encourage clinicians to evaluate patients with any of the anomalies described above more completely.

Diagnosis

The rarity of ASM has made prompt diagnosis difficult in most cases. However, palpation of a retrorectal mass in association with symptoms related to malfunctioning pelvic organs, the peculiar headaches and backaches, and association with other congenital anomalies should evoke suspicion of ASM. A roentgenogram of the sacrum showing the "scimitar" anomaly is virtually diagnostic.

A myelogram with opaque contrast medium will usually demonstrate the meningocele, and lateral views are valuable in delineating the width of the stalk and the level at which it emerges from the sacrum. However, a negative myelogram does not exclude the diagnosis, since the stalk may be so narrow[6] that the passage of contrast material can be prevented or even delayed for days. Air-myelogram has been recommended for these rare instances.

Lovelady and Dockerty[48] established a classification for retrorectal tumors, which may serve as an aid when the rare problem of a retrorectal or presacral mass is encountered. The so-called "developmental cyst" may manifest as a dimple in the posterior part of the anal canal. Chordomas, although of congenital origin, usually appear in adulthood, with symptomatology similar to that of ASM; but radiologic examination of the pelvis shows destruction of the sacrum and occasional calcifications.[55] The same is true for osseous tumors and teratomas. Neurofibromas are more solid and tender on palpation.

Anterior sacral meningocele was erroneously diagnosed as an ovarian cyst in some instances.[16, 22, 37, 69, 74, 78, 87] Careful rectal examination would eliminate this possibility by demonstrating the anatomic location.

Treatment and Results

Of the 120 patients with ASM, 42 did not have surgical treatment. Of these 42, eight died. A constipated infant had accidental rupture and died of meningitis[33]; a 31-year-old woman developed a spontaneous fistula connecting the ASM to the rectum and died of meningitis four years later[25]; another woman had rupture that led to fistulization of CSF and death following delivery.[88] Two other women[9, 34] died during or after delivery from obstructed labor and rupture of the ASM. Another woman[56] survived aspiration via the vagina but had spontaneous rupture weeks later and died of meningitis. A three-month-old baby had

TABLE 3. *Treatment and Results*

Procedure	Number of Patients	Number of Deaths	Complications and Remarks	References
No procedure or unknown	42	8		3, 4, 9, 10, 13, 15, 21, 25, 26, 27, 32, 33, 34, 38, 40, 46, 52, 65, 66, 70, 71, 72, 74, 80, 82, 90, present cases 2, 3, 4
Aspirations via rectum, vagina or "sterile"	17	6	8 recurrence 1 meningitis, cured 3 had definitive surgery later	4, 22, 23, 41 44, 47, 56, 57, 61, 64, 65, 71, 72, 78, 79, 81, 84
Vaginal or rectal drainage	5	3	1 arachnoiditis 1 meningitis, definitive surgery later	37, 42, 44, 79, present case 1
Abdominal surgery with closure of stalk	17	3	1 meningitis, cured 1 failure to reach the stalk	4, 11, 14, 25, 29, 39, 46, 51, 63, 64, 68, 76, 83, 87, present case 1
Abdominal surgery without closure of stalk	15	6	2 long-standing CSF fistula 1 arachnoiditis, paraplegia 3 meningitis, cured 2 recurrence 3 definitive surgery by laminectomy	12, 15, 19, 20, 23, 25, 26, 35, 36, 50, 69, 77, 78, 81, 87
Kraske approach, closure of stalk	16	0	1 infected hematoma, meningitis cured 2 long-standing CSF fistula, cured 1 wound infection	6, 17, 23, 25, 28, 30, 31, 41, 43, 62, 67, 72, 73, 86, 89, present case 5
Kraske, no closure of stalk	2	2	(one was "perineovaginal" approach)	18, 24
Laminectomy	19	0	3 failure to close wide stalks (two had definitive abdominal surgery later)	1, 2, 4, 8, 15, 16, 30, 36, 45, 49, 53, 58, 59, 76, 78, 85
Unknown procedure	1	0	1 CSF fistula	60

surgery for posterior meningocele and died. His ASM was found at postmortem examination.[66] Death also occurred in the case of an 11-year-old boy whose treatment was not described.[32] Three women, including our Patient 2, needed cesarean sections,[38] probably for obstructed labor. Four other patients were asymptomatic.[4, 38, 65, 80] The remaining patients had symptoms of varying severity.

Observing this group of untreated patients, it appears that leaving ASM without surgical intervention carries a significant risk, especially when there is a chance of pregnancy.[85] We conclude that even asymptomatic patients with ASM should have surgical treatment when the cyst is found to be enlarging on repeated rectal examination or when there is a chance of pregnancy.

For symptomatic patients, surgical treatment is the treatment of choice. The main goal of the operation is to divide the stalk of the ASM and provide watertight closure of the dura at the sacral side. Since the cyst itself does not produce fluid, it will eventually shrink. This treatment was considered sufficient by most investigators.[30]

As mentioned above, teratomas and dermoid cysts frequently accompany ASM. The death of a woman[25] due to terato-

carcinoma 22 years after successful nonresectional surgical treatment of ASM has been reported. We think these facts justify removal of the cyst and any adjacent tumor during operation.

Table 3 summarizes the surgical approaches to ASM and their results. The abdominal route was first successfully employed by Roux[64] and later by Weber.[87] Patients should have complete mechanical and antibiotic bowel preparation preoperatively. The procedure involves developing the space between the rectum and sacrum, conserving the sigmoidal vessels until the stalk of the meningocele is identified bulging out of the sacrum. This may be transfixed when narrow, or cut and closed in a watertight manner from the sacral side with nonabsorbable sutures. Compression of the jugular veins will increase CSF pressure and show whether the closure is secure. The cyst is emptied and bluntly dissected out, as is any additional teratoma or dermoid cyst. Care must be taken not to injure the posterior rectal wall at this stage. If this occurs or is suspected, a complete diverting sigmoid colostomy should be done. Meticulous hemostasis is also necessary, since an infected hematoma may endanger the closure of the dura and result in life-threatening meningitis.

Thirty-two patients had surgical operations for ASM via the abdominal route. In only 17 of these 32 cases were the pathologic consequences of the ASM fully recognized before or during operation, with proper ligation of the stalk. Three of these patients died of meningitis.[39, 63, 83] Another patient[87] had meningitis postoperatively and was successfully treated. In one patient,[14] the stalk could not be reached from the abdominal route, and was later resected by a posterior approach.

Fifteen patients had abdominal operations without ligation of the stalk, or without proper preoperative or intraoperative diagnosis. The results were worse. Six of these patients died,[20, 35, 50, 81, 87] and others had numerous complications, including arachnoiditis with paraplegia,[77] CSF fistula,[12] and long-standing meningitis.[15, 19, 69, 78] In some of these cases the ASM was erroneously considered to be an ovarian or para-ovarian cyst and was partially excised, marsupialized, and anastomosed to the bladder, with devastating results.

Although it is rare, surgeons should consider the possibility of ASM when an unsuspected retroperitoneal cystic mass is encountered during surgical operation.

The posterior parasacral route, or Kraske's approach, was recommended by Pupovac in 1903.[62] It consists of a vertical incision near the midline extending from near the rectum to above the coccyx, removal of the coccyx, and exposure of the retrorectal space. This approach provides access to the cyst, but access to the stalk is limited when it emerges from high on the sacrum. Sixteen patients had successful surgical treatment using this approach. Two cases[23, 67] were complicated by CSF fistulas, which eventually healed.

Adson[2] introduced ligation of the stalk of the ASM after sacral laminectomy in 1938. With this method the communication is closed from the posterior aspect and there is no access to the cyst itself or any accompanying tumor. When a tumor is palpated by rectal examination after evacuation of the cyst at the time of operation it should be removed in another stage. This technique was employed in 19 cases, with no mortality. Three patients[4, 76] had wide stalks that could not be closed this way, and a second procedure with an abdominal approach was necessary.

Aspiration of an ASM as a diagnostic or therapeutic procedure is not only useless, since the cyst rapidly refills, but also extremely dangerous, since infection may develop within the cyst and extend to the

central nervous system. There is no way to insure a sterile aspiration done via the rectum or the vagina. Five patients died after such aspirations.[22, 56, 61, 72, 81, 84] Meningitis has even occurred after aspiration during laparotomy.[78] Similar considerations apply for procedures employing drainage of the cyst through the vagina or the rectum; these have resulted in an almost 100 per cent mortality rate.[42, 44, 79]

In summary, the results of surgical treatment for ASM are good when the diagnosis is made preoperatively, regardless of the approach used. Selection of the best surgical approach should be based on the width of the meningocele stalk and the level at which it emerges from the sacrum.

Summary

Five new cases of anterior sacral meningocele are presented, including one secondary to neurofibromatosis, a previously undescribed association. The literature is reviewed, drawing attention to the relationship between anterior sacral meningocele, sacral dysgenesis, and other congenital anomalies. Special consideration is given to the clinical features of this entity, as well as to the techniques and results of surgical management.

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Announcement

The American College of Surgeons' 63rd Annual Clinical Congress will be held in Dallas, Texas, October 17–21, 1977.

Included in the program are several specialty sessions devoted to colorectal surgery:

Colon and Rectal Problems in Infants and Children — Interdisciplinary. Alfred A. deLorimier, M.D., San Francisco, Moderator

Complications of Anorectal Surgery — Prevention and Management. James A. Ferguson, M.D., Grand Rapids, Moderator

Emergency Management of Colonic and Rectal Problems. John H. Remington, M.D., Rochester, New York, Moderator

Special Procedures in Colonic and Rectal Surgery. Burchard E. Winne, M.D., Toledo, Moderator

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