

Late Paraplegia In Diastematomyelia

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Key words: congenital spinal malformation, progressive paraparesis

IMAJ 2000;2:944-945

Diastematomyelia is a congenital dysraphism of the spinal cord in which the affected segment is longitudinally divided by a band of fibrous tissue, cartilage and bone [1]. This kind of malformation is classified into two types according to the composition of the dural coverings and intervening mesenchymal tissue. Type I malformations comprise two dural sacs, each of which contains a single spinal hemisection, and a bony or fibrocartilaginous septum. Type II malformations consist of a single dural sac and intradural fibrous bands. They support the theory of non-involution of a firm midline structure that prevents the fusion of the separated parts [2]. Diastematomyelia may be clinically silent, unrecognized into adulthood [3], or it presents with cutaneous findings such as hairy patch, dimple, hemangioma, subcutaneous mass or teratoma at or near the level of the lesion in neurologically asymptomatic patients [4]. Computed axial tomography demonstrates sagittal bony spurs more clearly than does plain X-ray film, and magnetic resonance imaging also shows a well-pronounced bony septum eventually associated with split cords [5].

Our case report describes a young man who developed severe paraparesis in his 30th year after repetitive falls. We discuss the methods used to discover the previously silent diastematomyelia.

Case Description

A 36 year old man with a long history of low back pain, mild weakness in his lower limbs and recurrent falls was hospitalized with acute exacerbation of pain and weakness. Five years earlier

he began to complain of difficulties in urination, and in the last 3 years his sexual function was affected. Motor examination demonstrated severe paraparesis with normal muscular tone. Sensory examination revealed anesthesia from L3 level on the right side and L1 level on the left. Thermal and vibratory sensation was reduced in the right leg and absent in the left leg. Abdominal reflexes were active and symmetric, but cremaster reflexes were absent.

Patellar tendon reflexes were symmetrically reduced, and the achilles reflexes were increased, mostly on the right side. Babinsky and Chaddock pathological signs could not be elicited. Anal sphincter tone was preserved. Straight leg raise testing was positive at 30 degrees of hip flexion on both sides. The patient was independent in activities of daily living and transfers and was able to walk very slowly with a walker. T11-L4 CAT scan revealed a fine bony septum toward the spinal canal, with a sagittal cleft in the spinal cord at the L3 vertebral level, transversal spinal stenosis at T11 and sagittal spinal stenosis at L1-L2 [Figure 1]. Sagittal T1 proton density and T2 magnetic resonance images revealed splitting of the spinal cord with myelomalacia at level L2 and disc hernia with spinal stenosis at D12-L1; the spinal cord extended till vertebra L4. Somatosensory evoked potential demonstrated a severe conduction dis-



Figure 1. CT scan showing fine bony septum with a sagittal cleft in the spinal cord at L3 level.

order in the sensory pathways in response to right and left tibialis posterior nerve stimulation. Intravenous pyelography showed a middle residual volume in the bladder after voiding.

The urethrocytogram showed no pathological changes in the bladder wall, no voluntary voiding and no reflux. The patient refused urodynamic evaluation.

Comment

The etiology of the symptoms of diastematomyelia in adult patients is not clearly understood. Low back pain, which was the chief complaint in our adult patient, was found to be associated with a tethered or fixed-in-position spinal cord. This kind of fixation occurs when adhesions associated with the split spinal cord attach to a solid septum, or exit through the dorsal cerebral spinal fluid space. This would explain why our patient's spinal cord

terminates at level L4. Neurological symptoms such as paraparesis have been thought to result from movement of the fixated spinal cord within the spinal canal, with subsequent local injury to the neural elements from traction by the dentate ligament or transfixing bony spur [3]. Presumably this deterioration occurred also because of tethering or traction of the spinal cord, extending to vertebra L4, and secondary to falls as described by the patient.

CT, MRI and SSEP were the tests of choice for proving the existence of pathological manifestations inside the

SSEP = somatosensory evoked potential

lumbar spine. Resection of the spur should be performed in patients who have otherwise irreversible progressive neurological manifestations. Patients without neurological manifestations should be carefully observed, and if progression is noted a resection should be performed [5].

The present case provides documentation of a diastematomyelia, unknown until adulthood and discovered by the appearance of neurological symptoms after repetitive falls. Early recognition followed by surgical resection of this spinal defect could prevent late paraparesis.

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Human Bite on Penile Shaft from Oral Sex as a Portal of Entry for Streptococcal Toxic Shock Syndrome

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Key words: *Streptococcus*, toxic shock syndrome, human bite, penile shaft

IMAJ 2000;2:945-947

The medical community has long recognized *Streptococcus pyogenes*, or Group A *Streptococcus*, as a cause of a broad spectrum of acute illness (e.g., pharyngitis, scarlet fever, impetigo, erysipelas, necrotizing fasciitis) and of rheumatic fever and post-streptococcal glomerulonephritis, two non-suppurative sequelae [1]. Since the 1980s, there has been a marked increase in the reporting of highly invasive GAS infections associated with shock and organ failure, with or without necrotizing fasciitis. Such dramatic cases have been defined as StrepTSS, and have seized the attention of the general public as the "flesh-eating bacteria" [2]. We report the favorable

GAS = Group A *Streptococcus*

outcome of such a case of *Streptococcus* toxic shock syndrome, in which the portal of entry was a human bite on the penile shaft during oral sex.

Case Description

A 20 year old previously healthy male was admitted to our department because of high fever (40°C), chills and general malaise of 24 hours duration. He also complained of pain in the pelvic region around the base of the penile shaft. The patient stated that he had suffered slight trauma to the penis while receiving fellatio one day prior to the appearance of his local and systemic signs. The patient denied any respiratory, urinary, gastrointestinal or other complaints. His medical history was unremarkable. On physical exami-

nation he appeared septic, with fever of 40°C, heart rate 120/min, blood pressure 110/50, and 18 breaths/min. The penile shaft, pubis and the right inguinal region were tender, swollen and erythematous, with local lymphadenopathy. On the penile shaft, two minor open wounds (3 mm each) surrounded by a red halo with no apparent pus or discharge were noted. No other relevant physical findings were found. Initial laboratory findings showed leukocytosis of 21,000/mm³ with a left shift; arterial blood gases on room air showed normal oxygenation with acute respiratory alkalosis. Liver and kidney function tests were normal. Chest X-ray and electrocardiogram were normal. Based on the suspicion that this acute febrile dis-

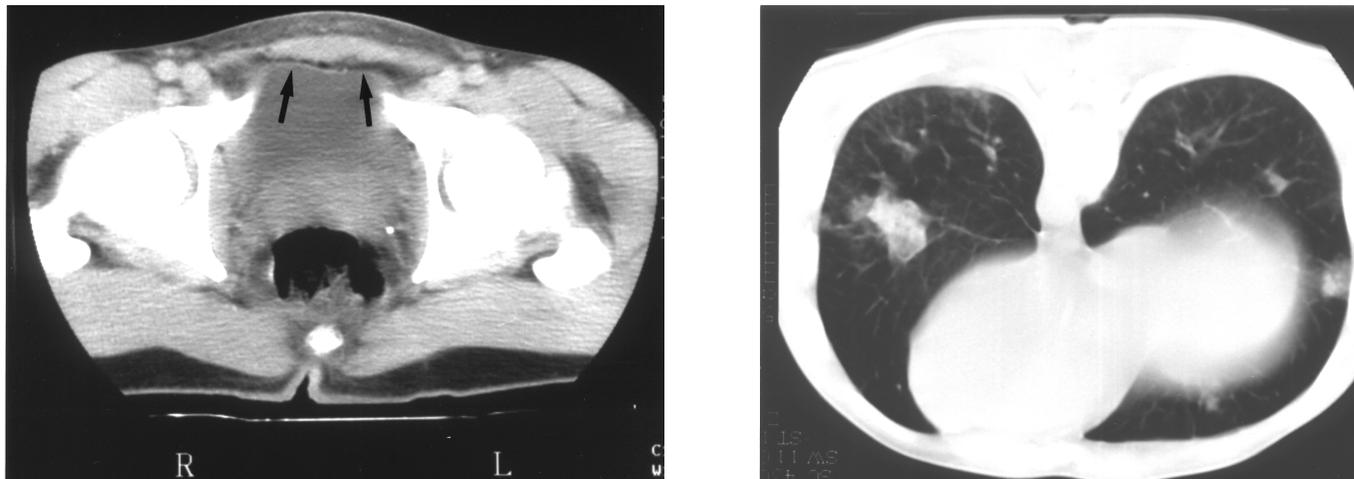


Figure 1. Axial CT of the lower abdomen and chest. **[A]** Thickening of the adipose layer, anterior to the rectus abdominus, with unclear margins of the muscle and its surrounding fascia. **[B]** Multiple bilateral lung lesions.

ease was caused by a human bite, the patient was started on fluids and broad-spectrum antibiotics (amoxicillin-clavulanic acid and vancomycin) to cover common pathogens of the human oral flora.

During the first day of follow-up the patient's condition deteriorated, as was manifested clinically by an acute confusional state, hypotension (90/55 mmHg), acute respiratory distress (40 breaths/min) and the use of inspiratory accessory breathing muscles. The pubic erythema spread rapidly upwards toward the umbilicus. Repeated laboratory investigation revealed elevated liver enzymes (alanine aminotransferase 225 u/L, aspartate aminotransferase 162 u/L). Arterial blood gases showed respiratory alkalosis and hypoxemia of 60 mmHg while breathing with an oxygen mask (40% oxygen). A computed tomography scan of the pelvis and abdomen revealed edema of the penile shaft and thickening of the adipose layer anterior to the rectus abdominus, with unclear margins of the muscle and its surrounding fascia, suggesting an inflammatory process of the connective tissue of the abdominal wall [Figure 1a]. A CT scan of the chest showed multiple lesions on the periphery of both lungs suspected as septic emboli [Figure 1b]. Group A β -hemolytic streptococci sensitive to penicillin were cultured from multiple blood specimens. The antibiotic treatment

was changed to a combination of high dose penicillin (24 million units/day) and clindamycin. During the following days of follow-up the patient gradually recovered.

Comment

StrepTSS is a particularly severe and rapidly progressive manifestation of GAS infection. Not all life-threatening streptococcal illnesses are StrepTSS. The Center for Disease Control and Prevention has formulated a set of criteria that must be met before the diagnosis can be made [3]. In our patient, isolation of GAS from the blood, hypotension (90/55 mmHg), elevated liver enzymes and adult respiratory distress syndrome fulfilled the criteria for the diagnosis of StrepTSS.

StrepTSS usually presents with an initial skin and soft tissue infection. Person-to-person contact readily transmits these streptococcal strains [4]. The streptococcal pyrogenic exotoxins are the mediators in the pathogenesis of this disorder. These specific strains also produce proteases whose virulent properties enable them to invade human tissues and bloodstream, causing a deadly cascade of inflammatory mediators. A typical initial presentation is that of flu-like symptoms. Within the next 3 days, a presentation of intense

StrepTSS = *Streptococcus* toxic shock syndrome

local pain, high fever, prostration, localized swelling, and erythema is common. What follows is a classic case of hypotension and symptoms of multi-organ failure. Laboratory investigations show organ dysfunction, white blood cell elevation with left shift, and declining hemoglobin and platelet counts. Blood cultures are positive in about 60% of StrepTSS patients. Prompt antimicrobial therapy is crucial. Empiric broad-spectrum coverage for septic shock should be administered initially, and once the etiology is confirmed high dose intravenous penicillin G is the drug of choice. A first generation cephalosporin or vancomycin is recommended for penicillin-allergic patients [1,2,5].

To our knowledge, this is the first recorded case of StrepTSS with such an unusual portal of entry. Based on the fact that approximately 30% of the population are carriers of group A β -hemolytic streptococci in their oral flora [4,5], physicians should be aware of this potentially deadly syndrome and its possible modes of transmission. Even the slightest suspicion is enough to warrant a full workup in these patients. The issue of public awareness of StrepTSS is important since the patient must immediately seek medical attention for such symptoms. It is also important to educate the public about the importance of the use of condoms as a method to prevent StrepTSS,

since such practices are considered relatively common in today's society.

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Neoadjuvant Chemoradiation Treatment of Locally Advanced Adenocarcinoma of the Duodenum

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Key words: adenocarcinoma, duodenum, neoadjuvant chemotherapy, survival

IMAJ 2000;2:947-948

Adenocarcinoma of the duodenum represents a rare neoplasm characterized by an indefinite symptomatology. Found in less than 0.05% of tumors at autopsy [1], it constitutes 0.35% of all tumors of the gastrointestinal tract [2]. Still, 75% of the primary tumors of the duodenum are adenocarcinomas, and the duodenum is the origin of approximately one-third to one-half of adenocarcinomas of the small bowel.

This low incidence renders optimal treatment strategies and prognosis unclear. The vague non-specific symptoms, together with its highly aggressive behavior contribute to the advanced stages upon presentation and the low odds of cure. Low resectability rates and high locoregional recurrence rate are largely responsible for the failure of surgery for localized duodenal adenocarcinoma.

It has been suggested that neoadjuvant chemotherapy may offer patients with peri-ampullary adenocarcinoma a better chance for resectability, a lower rate of local recurrence, and thus an improved survival. We report our recent experience with two patients who presented with locally advanced adenocarcinoma of the duodenum.

Tomographic scanning and endoscopic ultrasound revealed these tumors to be non-resectable, but neoadjuvant chemoradiation treatment enabled curative resection.

Case Description

Case 1

A 27 year old male was admitted to our department for evaluation of mild abdominal pain, nausea and weakness of 10 months duration. His abdominal examination was normal but hemoglobin was 7 g/dl. Endoscopy revealed a large ulcerated tumor in the second part of the duodenum, and biopsies from the tumor revealed a poorly differentiated adenocarcinoma. Endoscopic sonography discerned a T3N1 tumor, partially surrounding the vena cava. Spiral computed tomography visualized a soft tissue mass circumscribing the second part of the duodenum, contiguous with the retroperitoneum and pressing against and partially surrounding the vena cava [Figure 1A].

Due to the local advancement of the tumor, neoadjuvant chemotherapy was elected (the patient refused radiotherapy). The patient received three

courses of treatment, each consisting of cisplatin 30 mg/m² on the first 3 days and Gemzar 1,250 mg/m² on days 1, 8 and 15. Toxicity was monitored following this regimen.

Repeated CT showed a significant response with almost no evidence of residual tumor [Figure 1B]. Repeated endoscopic ultrasonography demonstrated a T0N1 tumor. Laparoscopic exploration ascertained resectability of the pancreatico-duodenal complex, and Whipple's operation was performed. During surgery extensive desmoplastic and fibrotic reaction was noted around the second part of the duodenum and the vena cava. The post-operative course was uneventful.

Histopathological examination of the surgical specimen found no residual tumor in the duodenum but a large area of fibrosis within its wall. Dissected lymph nodes showed only reactive changes. Twenty-four months after surgery the patient is well, with no evidence of disease recurrence on repeated scans.

Case 2

A 47 year old woman was admitted to our ward for evaluation of epigastric



Figure 1A. Pre-chemotherapy CT scan revealing a far advanced tumor of the duodenum.

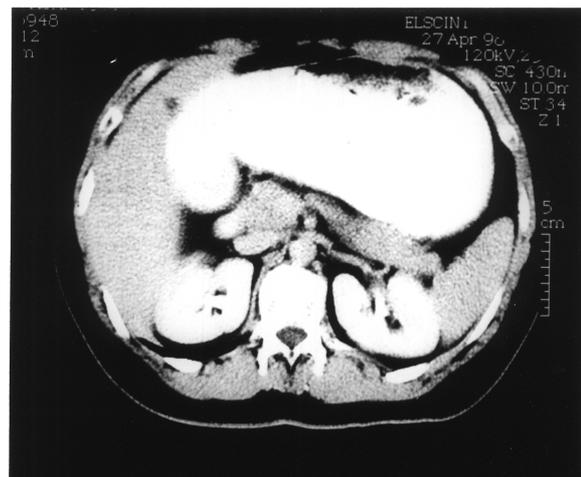


Figure 1B. Post-chemotherapy CT scan showing complete disappearance of the tumor.

pain, nausea and weight loss. No pathology was found on physical examination and in the subsequent laboratory workup, but endoscopy revealed a large ulcerated polyploid lesion in the second part of the duodenum. Biopsies taken showed a poorly differentiated adenocarcinoma of the duodenum that was graded by endoscopic ultrasonography as T3N1. Spiral CT showed a duodenal mass penetrating to the pancreatic head with some devastation of the bile ducts.

Due to the tumor's large size, neoadjuvant chemoradiation was elected. The patient received three courses of treatment as detailed above, and was

palliative procedures, such as bypass or biliary intubation, could be instituted.

This is especially true when using accurate pre-operative imaging such as high quality thin-section contrast-enhanced spiral CT. The adjunct of endoscopic ultrasonography has further increased the pre-operative staging [4]. Thus, the patients presented here were considered non-resectable.

Neoadjuvant chemoradiotherapy is an evolving strategy for patients with tumors of the peri-ampullary region. Yeung et al. [5] published their experience of this regimen in five patients with duodenal malignancies, and concluded that resection was enabled in

monitored for toxicity. Consequently, since the CT scan showed good response to the chemotherapy, radiation was initiated and dosed at 300 cGy daily for 10 days. After completion of the therapy, the CT scan revealed that the duodenal tumor has disappeared. Laparoscopic exploration revealed neither locally advanced disease nor metastases, and Whipple's operation was performed. The resected specimen disclosed a 1 cm tumor of the duodenum not extending beyond the muscularis propria. Twelve months post-surgery the patient is well.

Comment

Carcinoma of the duodenum comprises only 4% of all peri-ampullary cancers. Resection of these tumors offers the best survival rates. Sexe et al. [3] calculated the projected 5 year survival after resection to be 23%. They found that more than 50% of duodenal malignancies were non-resectable, and therefore only

80% of the patients following neoadjuvant chemoradiation. They also noticed that after this treatment no residual tumor was found, and suggested that a multicenter study be undertaken to evaluate the need for surgery after chemoradiation.

Similarly, our patients demonstrated that neoadjuvant chemotherapy or chemoradiotherapy could affect down-staging of the tumors and enable the safe complete resection of the affected organ and its surrounding, thereby offering the patient the best chance for survival.

In conclusion, we strongly recommend neoadjuvant chemoradiation for patients with advanced duodenal adenocarcinoma. We agree that only a multi-institutional study [5] will resolve whether chemoradiation alone is sufficient for the treatment of this malignancy. The decision to initiate neoadjuvant chemoradiation should be based on staging of the tumor utilizing spiral CT scan, endoscopic ultrasound and/or laparoscopic exploration.

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