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Currarino Syndrome: Efficacy of CT

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Currarino 症候群：CT の有用性

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今回，我々は，Currarino 症候群を呈した2症例を経験した．Currarino 症候群は，1）anorectal stenosis，2）sacral defect，3）presacral mass，を呈するものである．presacral mass としては，anterior maningo-cele，teratoma，enteric cyst 及びその混合型が過去に報告されている．今回，我々が経験した2症例のうち1症例は anterior meningocele，もう1症例は teratoma であった．これらは CT によって，ほぼ確診が得られた．また，triad のもつ意義として，文献学的にこの疾患群は遺伝的に発生し得ることから，単に患者個人において三徴を見いだすだけではなく，家族内発生を予期することにも有用と思われる．

Introduction

Recently we have experienced two cases of the Currarino syndrome, which are presented herein with special reference to the findings of CT-scan and with a review of the pertinent literature.

The Currarino syndrome, described for the first time by Kennedy[1] in 1926, was defined by Currarino[2] in 1981 as one variety of the so-called "split notochord syndrome". Its clinical triad can be represented as

ASP; i.e., anorectal stenosis, sacro-coccygeal abnormality, and a presacral mass. Commonly the chief presenting symptom is constipation since birth.

Materials

A 5-year-old boy at the Keio University Hospital and an 18-day-old girl at the National Children's Hospital, both presented in 1984, have been studied.

Case report

Case 1: The 5-year-old boy had a gradually progressive tendency towards constipation and was noticed to have an abdominal distention at 10 months. He was diagnosed elsewhere as having an anal stenosis and underwent a diverting colostomy one month after that. A simultaneous transabdominal biopsy of the lowest accessible part of the rectal wall revealed the presence of normal ganglion cells; therefore, the possibility of Hirschsprung disease could be eliminated. The colostomy was closed 3 months thereafter because of a peristomal abscess. At any rate, he had established a normal pattern of bowel movement by then. He was discharged with slight fecal incontinence at the age of 1 year and 2 months. Thereafter, abdominal distention and constipation persisted, and inducing bowel movements with a bougie at home for almost one year did not improve the condition. At 3 years of age good bowel habits were aided by a teleminsoft suppository; however, the underlying condition still did not improve. Therefore, he was brought to our hospital at the age of five.

The results of the physical examination were otherwise within normal limits. An abdominal plain X-ray showd the fusion of the third and fourth sacral bones and a right-half defect of the lower part of the sacrococcygeal bone, a defect which appears to be consistent with the so-called scimitar sacrum (Fig. 1,2). A barium enema revealed an anorectal stenosis and compression from the back, suggesting the presence of a

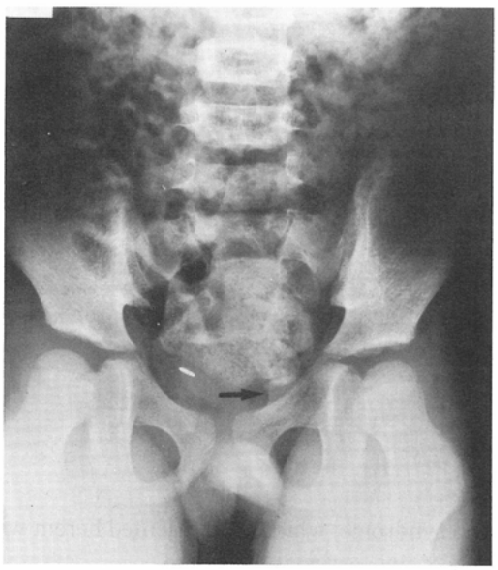

Fig. 1

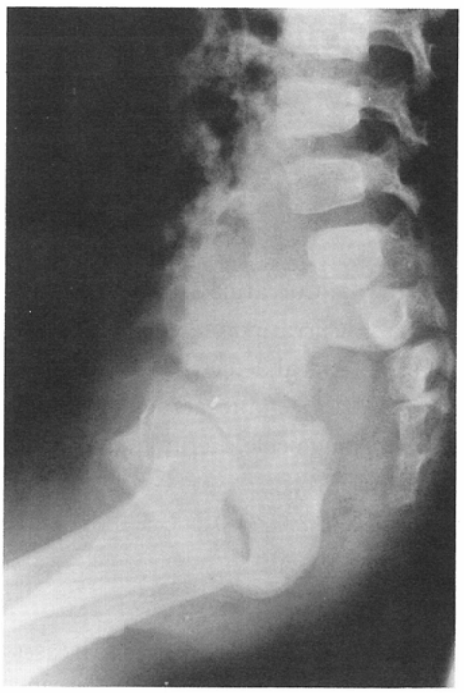

Fig. 2

Fig. 1 and 2 Case 1. Scimitar sacrum and fusion of lower sacrum are noted.

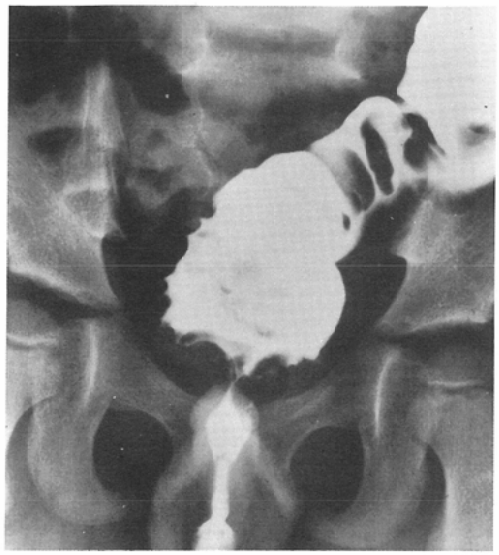

Fig. 3

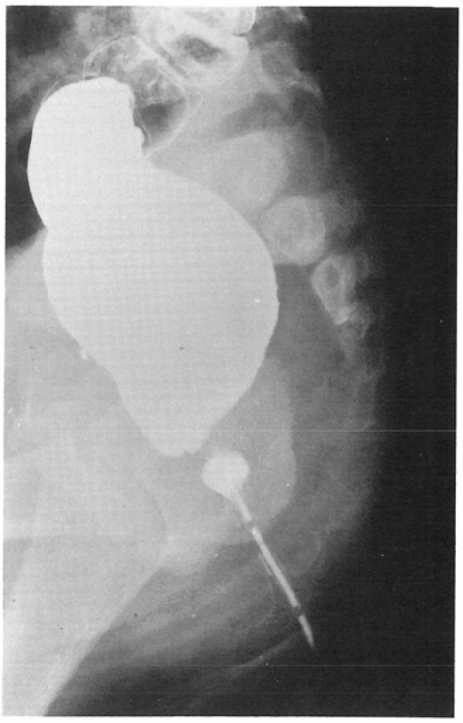

Fig. 4

Fig. 3 and 4　Case 1. Barium enema demonstrates anorectal stenosis and extrinsic compression on rectum from posterior aspect, suggesting presacral mass.

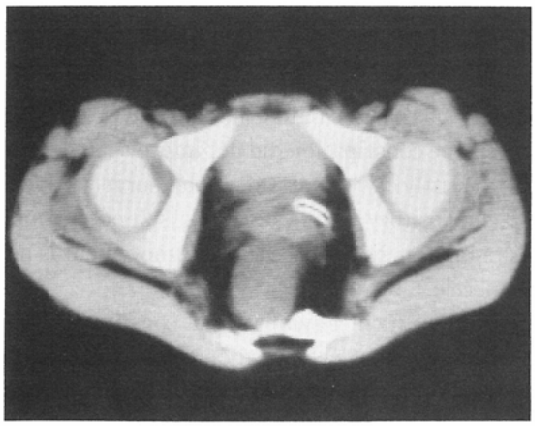

Fig. 5　Case 1. Myelo-CT shows CSF density mass impinging from the defect of sacrum.

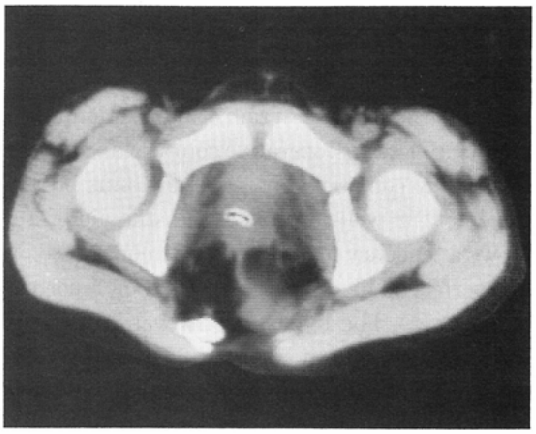

Fig. 6　Case 1. CT. Lower level than Fig. 5. There are coccygeal deviation and the fistula between the mass and rectum.

presacral mass (Fig. 3, 4). CT-scan clearly demonstrated an anterior meningocele of cerebrospinal-fluid density protruding from the defective sacral body, displacing the rectum (Fig. 5, 6). An anal-pressure stdy showed the dentate line present 3 cm from the anal ring, the rectoanal reflex therearound being intact. No urination difficulty was noted. Myelography showed a balloon-shaped cyst communicating at the upper

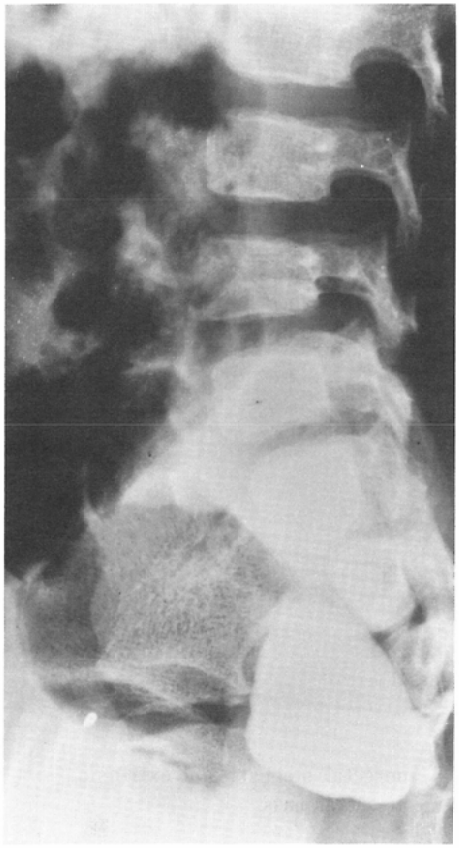

Fig. 7 Case 1. Myelogram shows anterior meningocele.

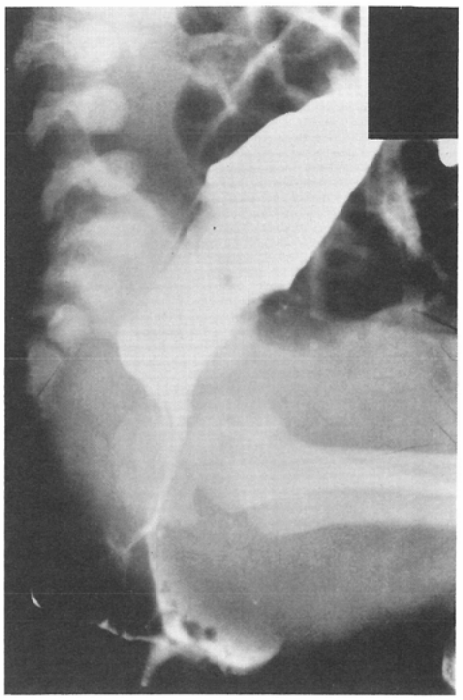

Fig. 8 Case 2. Barium enema reveals anorectal stenosis with suggestion of presacral mass.

level of the fourth sacral radix (Fig. 7). Meningomyelocelectomy was carried out at the age of five years and 5 months. The postoperative course was uneventful; however, the constipation did not improve, and there was a new complication; the anal tonus became flaccid. Presently, two years after the surgery, bowel movement is well-controlled with the aid of a teleminsoft suppository once a day. The sphincter tonus shows a tendency to improve, as demonstrated by a rctal examinaiton; however, the bowel movement is not still perfect. A histological examination reported lipomeningiocele.

Case 2: The 18-day-old girl was brought in to our hospital with chief symptoms of constipation and abdominal distention since the 17th day after birth. A colostomy was constructed on the 25th day after birth. Abdominal scout films revealed the coccygeal bone to be ectopic and deformed. A barium enema showed an anorectal stricture and displacement from the vack (Fig. 8). The mass could be diagnosed as a teratoma based on the CT finding of its lipid density and the site involved. Ectopia of the coccygeal bone was also identified (Fig. 9, 10). The tumor was removed and an anorectal anastomosis was carried out on the 36th day after birth. The tumor removed was reported to be a mature teratoma. The postoperative course was generally uneventful. Her bowel-habit control had been aided by the use of an anal bougie. The colostomy was closed five months after birth. At present, two years after birth, she is suffering from slight constipation, but the condition is improving gradually. Now, no postoperative organic stricture is noted, and the sphincter tonus is intact.

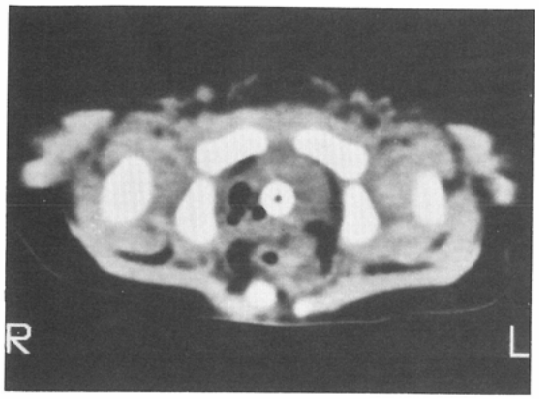

Fig. 9

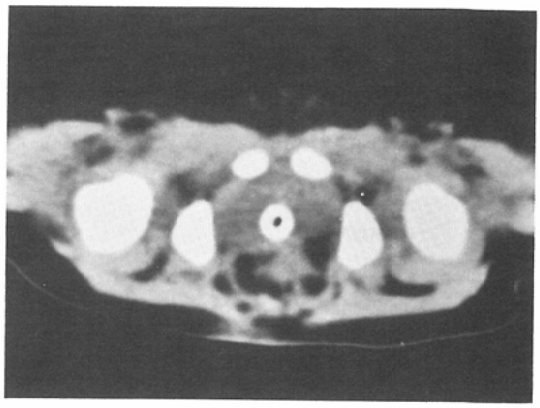

Fig. 10

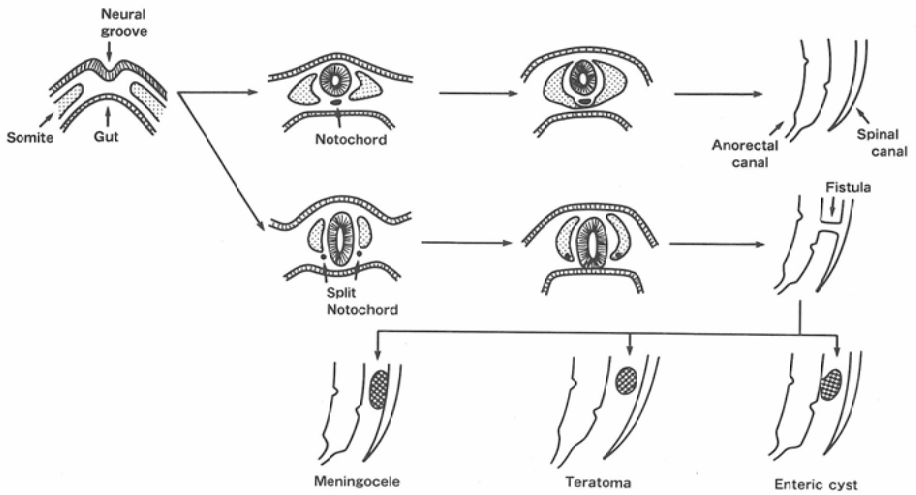

Fig. 11

Fig. 9 and 10　Case 2. CT demonstrated coccygeal deviation and presacral mass including fat density, suggestive of teratoma.

Discussion

To date fifty-six cases of Curarino syndrome consistent with the full triad have been reported[1)~17)]. The ages on established diagnosis range from the first day after birth to 89 years, and there is no significant difference in sex distribution (male 29, female 27 cases). Since birth, various degrees of constipation[1)~17)] were noted clinically. Other symptoms reported include fever, dysalgesia[4)], dysfunctional genital bleeding[4)], and so on due to bladder[4)6)8)12)13)16)] dysfunction, meningitis[6)15)], or perianoproctal abscess[5)8)11)16)13)]. Among these cases some asymptomatic cases have been reported.

As for the genetic background, Kirks[3)] reported that one of the triad can be detected in at least 50% of the members of the same family. Kennefick[4)] also reported that a sacral defect is transmitted as a dominant trait in some families, where a presacral mass is transmitted as a sex-linked dominant trait.

As for the embryologic background, Currarino[2)] explained as follows; The fetal notochord splitting causes the somites to fail to adhere between the neural ectoderm and the endoderm; therefore, there is a faulty vertebral development. On the otherhand, a fistula may form between the two structures. The

Table 1

Prognosis \ Presacral mass	Meningocele	Teratoma	Enteric cyst	Mixed	Unknown
do well	3	6	1	1	0
meningitis	1	0	1	0	0
abscess	1	2	0	0	0
constipation	3*	1*	0	0	0
recurrence	0	1	0	0	0
urinary tract disturbance	0	5	0	0	0
unknown	15	3	0	3	1
Total (48)	23	18	2	4	1

*includes our cases

remainder of the endoderm of the fistula may give rise to an enteric cyst. The remainder of the ectoderm element may give rise to an anterior meningocele. A teratoma is formed by the two elements left, with mesodermal elements being brought into the space (Fig. 11).

Based on this genetic and embryologic information, one of the triad can predict the potential concomitant presence of the other two. As this syndrome has a strong familiar preponderance, therefore, the first child being affected predicts that it is highly possible that a second child may develop at least part of the syndrome. Therefore, if such a syndrome is suspected, a barium enema and CT-scan will make possible an early diagnosis; thereby preventing chronic constipation and memingitis.

The prognosis of the patients with a presacral mass reviewed is shown in Table 1. Meningocele carried some postoperatie problems, such as meningitis and constipation. Meningitis might develop due to operative contamination or a remaining fistula. The high incidence of constipation is most likely to be attributed to the injury and compression of the cauda equina. On the other hand, a teratoma carries a problem of recurrence. The enteric cysts could not be fully evaluated because of the limited number of cases.

As for the diagnostic procedure, an abdominal plain X-ray, taken as the first maneuver on children with a chief complaint of constipation since birth, will show a deformed sacral bone. Subsequently, a barium enema will show an anorectal stenosis and retrorectal compression. Then, a CT scan will demonstrated a presacral mass. The CT scan, especially if made using a myelo-CT scan, makes possible an almost complete identification of a presacral mass; therefore, it appears to be very useful. That is, teratoma can be diagnosed by the finding of a mass with a fatty and calcified density; meningocele shows an enhancement in accord with CSF by myelo-CT. Lacking the above findings, an intestinal connection may suggest an enteric cyst.

Two cases of Currarino syndrome were reported here in with a review of the literature.

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