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Currarino syndrome with intramedullary spinal cord abscess related communication between the tethered cord and a presacral mass: A case report

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ABSTRACT

We herein report the case of a 21-day-old boy in which the detection of an intramedullary spinal cord abscess led to the diagnosis of Currarino syndrome (CS). He had a complete phenotype of CS, including sacral agenesis, an anorectal malformation, a presacral mass, and spinal cord malformations. In addition, he had an intramedullary spinal cord abscess. Intramedullary spinal cord abscess is rare in CS and is thought to require immediate intervention. Therefore, we additionally reviewed the available literature and discussed the therapeutic approach for CS with an intramedullary cord abscess.

Currarino syndrome (CS) is a hereditary disorder characterized as the triad of an anorectal malformation, sacral bony defect, and presacral mass. This syndrome was first described by Currarino et al. [1]. And CS is now known to be associated with the genetic mutation of *HLXB9* located at 7q36 [2]. The clinical phenotype of CS is variable. Some patients with CS do not exhibit the complete triad [3].

We herein report a rare case of a 21-day-old boy with Currarino syndrome manifesting as an intramedullary spinal cord abscess and review the treatment of CS with an intramedullary spinal cord abscess.

1. Case report

A 21-day-old boy was referred to a previous hospital due to a high fever. He was diagnosed as a urinary tract infection with a neurogenic bladder, and ceftriaxone was administered. Because the high fever persisted, he was transferred to our hospital and was diagnosed as an intramedullary spinal cord abscess by the lumbar puncture and magnetic resonance imaging (MRI) (Fig. 1). The cerebrospinal fluid was cloudy and cell count of the cerebrospinal fluid was 13,356/mm^3 (mononuclear cells: 1368/mm^3; polymorphonuclear cells: 11988/mm^3). Antibiotic therapy, including both panipenem/betamipron and gentamicin, was initiated and gentamicin was administered for two weeks. Panipenem/betamipron was changed to cefotaxime and administered for four weeks. Thereafter the patient's infection improved.

On admission to our department, funnel anus was also identified and neurogenic bladder was suspected, because he required urethral catheterization for urination. The X-ray film findings showed a hemisacrum, also known as a "scimitar sign," and MRI revealed the presacral tumor which was suspected to be a multicystic lipomatous tumor. In addition, thin slice image computed tomography (CT) was performed to allow for the precise evaluation of the anatomical findings. The CT showed the narrow continuity between the presacral tumor and rectal cavity; the presacral tumor continued to the spinal cord abscess through the Tethered cord. Similarly, a barium enema showed the fistula between the presacral tumor and rectum in addition to anal stenosis (Fig. 2).

We first performed colostomy at the transverse colon to prepare for the repair of the anorectal malformation. We did not select a one-stage treatment, including the resection of the presacral mass

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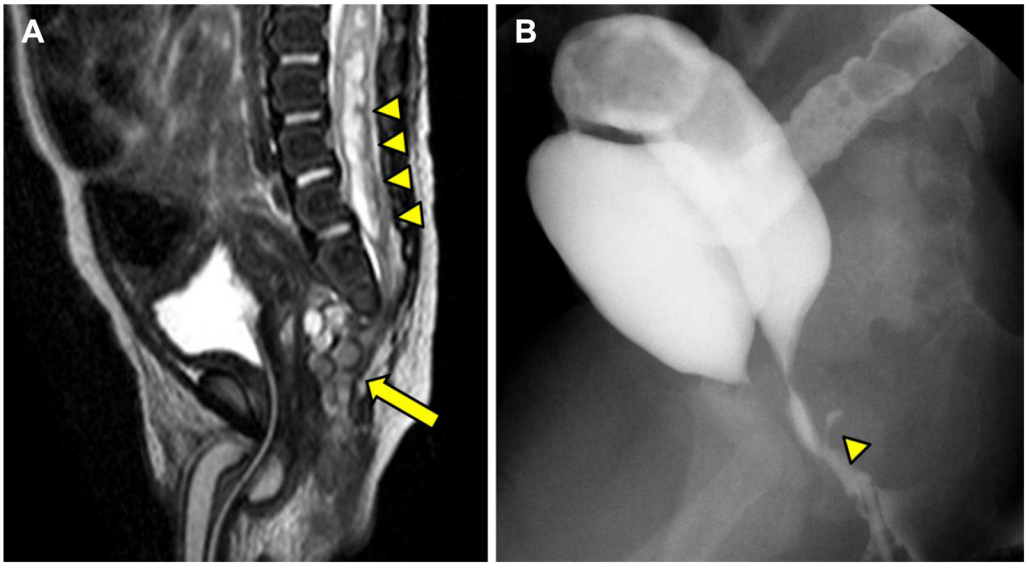

Fig. 1. A: sagittal T2-weighted magnetic resonance image shows a sacral abnormality and the presence of a multicystic presacral mass (arrow) and intramedullary spinal cord abscess (arrow head). B: The lateral view of the barium enema was suspected in the fistula between the rectum and the presacral mass (arrowhead).

and neurosurgery, because the treatment was thought to involve a high-risk of the recurrence of the abscess or meningitis. In fact, the colostomy prevented the recurrence of the spinal cord abscess after the presacral abscess improved.

Additionally, the repair of the Tethered cord was performed on the boy at 50 days of age and no recurrence has been observed following the repairing operation. At 6 months of age, the patient received presacral tumor extirpation through the posterior sagittal approach, and the rectal stenosis, 1.5 cm in length, was completely resected. We were not able to maintain his native anal mucosa because the anal stenosis was extremely rigid and the anal canal was less than 8Fr in diameter. The sacral bony defect that was detected preoperatively was not the coccyx bone. The gross specimen consisted of a 3.5 × 2.7 × 2.0 cm soft tumor with multiple cystic components (Fig. 2). The pathological findings showed the lipomatous tissue of the tumor was a mature teratoma with a cystic lesion. He was discharged on postoperative day 8. His colostomy was closed at one years of age. After these treatments, no recurrence of the presacral mass during a 1.5-year follow-up period has been observed. Neurogenic bladder was confirmed by a urodynamic study during the follow-up period. In addition, the patient currently requires the use of a glycerin enema for constipation, because the anal function was affected by the tethered cord.

2. Genetic examination

Informed consent for the genetic examination, only chromosome examinations, was obtained from the patient's parents. The genetic examination detected a mutation at the chromosomal region 7q34 in the present patient. Therefore, he was diagnosed with complete Currarino syndrome. We did not perform the familial screening because his parents did not consent to the screening genetic examinations.

3. Discussion

CS is a rare complex of congenital caudal anomalies, including the following three features: a sacral bony deformity, anorectal malformations, and a presacral mass. Since it was first described in

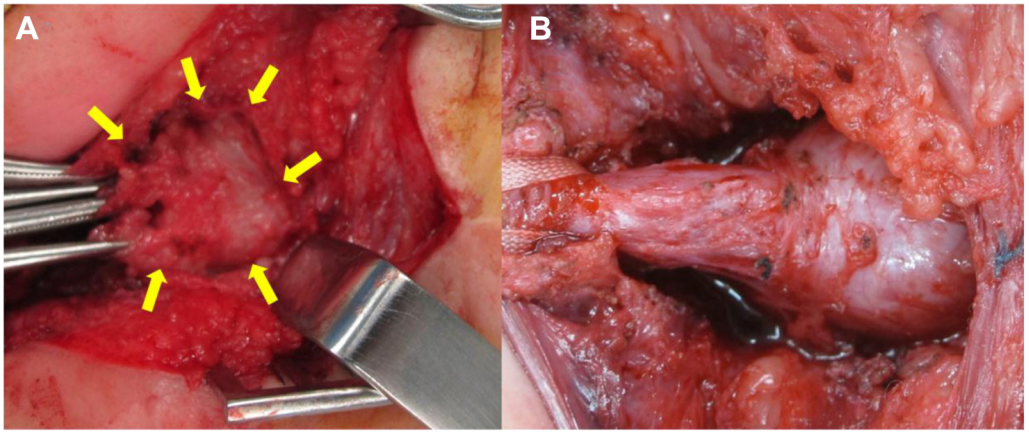

Fig. 2. A: A presacral mass was found at the bottom of the sacral bone and the gross specimen consisted of a 3.5 × 2.7 × 2.0 cm soft tissue (arrow) with multiple cystic components. B: After the removal of the presacral mass, anal stenosis was revealed and this rectal wall showed only mild fibrosis and chronic inflammation.