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An unusual cause for recurrent perianal sepsis in Currarino syndrome: Case report and review of the literature

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ABSTRACT

Currarino syndrome (CS) is a rare symptom complex comprising an anorectal malformation, a sacral bone abnormality and a presacral mass. Recurrent sepsis of the perianal area is a well-described post-operative morbidity. We report the case of a 4-year-old boy with CS and chronic perianal sepsis in the form of a draining sinus at the level of S5, which had plagued him since removal of a benign sacro-coccygeal teratoma at the age of 1 year. Normal alfa feto protein and beta-human chorionic gonadotropin levels ruled out malignant recurrence of a sacrococcygeal teratoma, whilst an MRI showed a large presacral abscess with no fistulous connection to the rectum. Biopsy and drainage of the abscess was performed through a posterior sagittal approach and pus was sent for microscopy, culture and sensitivity. Microscopy demonstrated the presence of acid-fast bacilli, confirming infection with *Mycobacterium tuberculosis* (TB). A review of the literature revealed paucity in the documentation of microbes related to this common complication with no previous reports of TB as the aetiological pathogen. This case is of particular interest as the patient was not immunocompromised, had no TB contacts or history of previous TB infection, and highlights the importance of actively excluding the cause of recurrent perianal sepsis, particularly in syndromes where sepsis is a common complication.

Currarino syndrome (CS) is a malformation complex characterized by 3 main features. These include an anorectal malformation (ARM), typically anal stenosis but may include any low-type ARM; a sacral defect, most commonly a scimitar sacrum affecting S2–S5 but usually sparing S1; and thirdly a presacral mass, most commonly an anterior myelomeningocoele followed by a sacro-coccygeal teratoma (SCT), enteric cysts, hamartomas or a combination of these [1–3].

The syndrome has been referred to as a triad because of these three distinguishing features, however multiple associated anomalies may or may not be present. These include urinary tract abnormalities, female genital tract abnormalities and a tethered spinal cord [4,5]. Consequently the term Currarino syndrome has gained widespread acceptance, replacing previous nomenclature which included caudal regression syndrome, hereditary sacral agenesis and ASP Complex (Anal atresia, Sacral abnormalities and a Presacral mass) [4,5].

SCT is encountered in 40% of patients with CS [3,6]. Although the risk of malignant transformation of SCT in CS is lower than that for isolated SCT (approximately 1%) [1] at least 10 reports of malignant change have been recorded in patients with CS, with a recommendation to remove the SCT as soon as possible within the first year of life [7]. Perianal abscess and recurrent infection of the perineum continue to plague these patients even after operative intervention [8]. This is likely due to unrecognized fistulous connections between the presacral space and rectum.

1. Case report

1.1. History

A four-year-old boy presented to our institution with a draining sinus at the level of S5. He had been treated at another facility as a

Abbreviations: CS, Currarino Syndrome; AFP, Alfa feto protein; B-HCG, Beta-human chorionic gonadotropin; TB, *Mycobacterium tuberculosis*; ARM, Anorectal malformation; SCT, Sacrococcygeal teratoma; GIT, Gastrointestinal tract; HLXB9, Homebox protein B9; MNX1, Motorneuron and panceas homebox 1; PSARP, Posterior Sagittal Anorectoplasty.

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neonate where he presented with intestinal obstruction secondary to an anorectal malformation for which he received a colostomy on day 2 of life. This was followed, in 2010, by a posterior sagittal anorectoplasty (PSARP) at 6 months of age. In 2011, a presacral mass was biopsied and found to be a benign SCT. In 2012, a posterior sagittal approach was used to excise the SCT and the colostomy was reversed that same year.

Since 2013, the child had undergone multiple incision and drainage procedures for recurrent presacral abscesses complicated by chronic sinus formation.

In 2015, the patient presented to our institution where the mother reported foul smelling pus draining from a sinus within the posterior sagittal scar. The pus had been draining for the past 2 years in between hospital admissions for incision and drainage procedures. The patient also reported constipation requiring medical therapy. Of note to the case was that he had no positive tuberculosis contacts.

1.2. Examination

The child appeared well and was thriving. He was apyrexial with a heart rate in the normal range for his age. There was no palpable lymphadenopathy. His chest was clear with good air entry bilaterally. Cardiac examination revealed no murmurs. His abdomen was soft with no palpable masses. Posteriorly, there was a midline scar with a small sinus draining pus at the level of S5. The perineal examination showed a posterior sagittal scar with a well-healed anoplasty.

1.3. Investigations

Renal ultrasound was normal. Blood tests revealed a normal white cell count and C-reactive protein. Beta-Human Chorionic Gonadotropin (B-HCG) and Alfa Feto Protien (AFP) levels were not elevated. HIV test was negative. A pelvic X-ray demonstrated a classic scimitar sacrum with a right-sided defect (Fig. 1).

An MRI demonstrated a midline pelvic-cutaneous fistula at the level of S5. There was intrapelvic extension with a presacral fluid filled cavity (3.4 × 3.3 × 6 cm). The fluid was heterogeneous and the wall of the cavity was rim enhancing suggesting an abscess. A second adjacent abscess was also noted (1.4 × 1.2 cm). No communication between presacral abscess and rectum could be elucidated (Figs. 2–4).

1.4. Operation

An incision with drainage and fistulectomy was performed through a posterior sagittal incision. No communication of the abscess to the bowel was noted intra-operatively. Pus was sent for microscopy, culture and sensitivity, which showed infection with Streptococcus group F and, surprisingly, a high concentration of acid-fast bacilli, consistent with the diagnosis of tuberculosis. The fistulous tract and a biopsy of the abscess wall were sent for histology, which confirmed that there was no recurrence of the SCT or other malignancy.

1.5. Treatment

The patient had already received co-amoxiclav and so was appropriately covered for the streptococcus, but was also started on standard treatment for extrapulmonary tuberculosis (TB). This included rifampicin, isoniazid, pyrazinamide and ethambutol, which he was to receive for 9 months.

2. Discussion

The first report of a patient with features resembling CS was first published by Kennedy in 1926 [9]. Subsequently, in 1974 Ashcroft and Holden reported a series of 17 patients with similar findings and proposed an autosomal dominant inheritance pattern [4]. It was only in 1981, that a radiologist, Guido Currarino, described the syndrome complex which now bears his name in its entirety, and postulated a common embryogenesis [2].

Currarino, noting the vertebral, neural and intestinal anomalies occurring at the same anatomical level, suggested abnormal endodermal adhesions and defects of the notochord. In normal early fetal life, the endodermal layer of the body stalk (which becomes the future gastrointestinal tract [GIT]) is closely approximated to the neural ectoderm. When the notochord appears, the somites engulf the notochord by extending forward and medially,

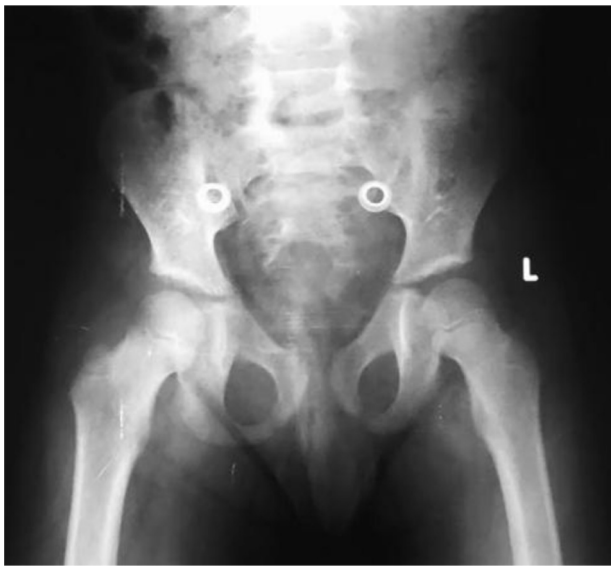

Fig. 1. AP pelvic X-ray demonstrating classic scimitar sacrum.

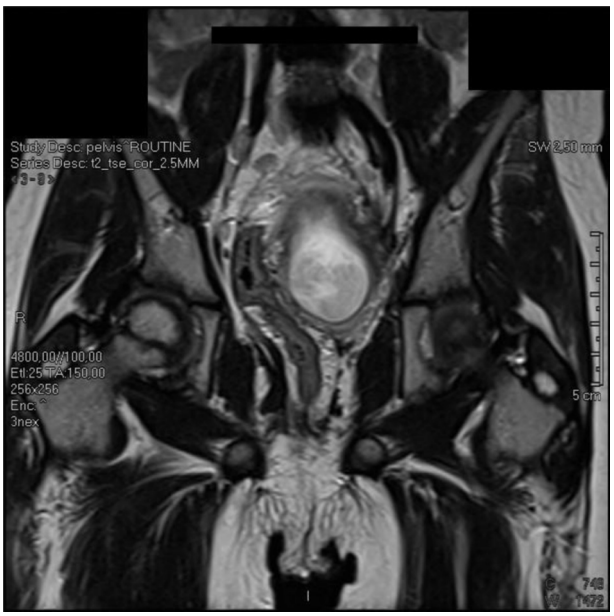

Fig. 2. Coronal section showing the presacral abscess to the left of the rectum.