Sacrococcygeal Teratoma with Recto-Sigmoid Fecolith in Adult

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ABSTRACT

Sacrococcygeal tumors are present most frequently in infancy and childhood. Incidence in the newborn is 1 in 40000 with a female to male ratio of 10:1. It is rare in adults. Less than a hundred cases of teratoma in adults have been documented in literature. Constipation is a common complain; that is refractory to treatment or results in complications such as fecal impaction warrants consideration of occult causes. In most cases, it is diagnosed in childhood, with most descriptions being found in the pediatric literature. It is extremely uncommon for the syndrome to present in adulthood. We report a case of sacrococcygeal teratoma in adult, which has treated in our hospital. The presented case has shown a large thin walled cystic pre-sacral, retro-rectal cystic mass along the intra-luminal recto-sigmoid rounded ball of fecolith. Here, we highlighted the association of sacrococcygeal cystic teratoma in adult male with fecolith, which is rarely documented. So, we should also search the cause of constipation in adulthood, not only in paediatric age-group; that is refractory to treatment or results in complications such as fecal impaction.

Key words: Fecolith; Hirschsprung's disease; sacrococcygeal; teratoma

Introduction

The presacral mass, sacral bony abnormality and anorectal malformation was first described by Bryant in 1838.[1] Kennedy in 1926[2] and Ashcraft and Holder in 1974[3] further characterized the condition, with Currarino and associates[4] in 1981 recognizing the triad as occurring by a common embryogenic mechanism. It is usually diagnosed in pediatric age group. Therefore, we sought to describe the presacral mass, sacral agenesis and recto-sigmoid fecal ball in adult patient.

Case Report

A 35-year-old man was referred to radiodiagnosis department with a left lower quadrant mobile mass and a history of lifelong constipation. He had chronic painful abdominal cramps and on-off mild distension but he was healthy. He claimed to have never had a spontaneous bowel movement. He denied a change in his bowel habit, unusual weight loss or rectal bleeding, and he had no history of bladder or sexual dysfunction. He had no family history of similar bowel symptoms. Examination revealed a distended lower abdomen with a spherical, freely mobile mass in the right lower quadrant.

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The patient underwent the abdomino-pelvic ultrasound examination and shown a large well defined low level echogenic cystic lesion in pelvic and right lower quadrant abdomen [Figure 1a]. Another rounded echogenic lesion with distal acoustic shadowing seen eccentric within large cystic lesion [Figure 1b]. Urinary bladder anteriorally shifted by large pelvic cystic mass. Computed tomography (CT) of the patient's pelvis and abdomen confirmed a large highly dense fluid filled well defined cystic lesion in pre-sacral and retro-rectal area with anterio-cranially displaced urinary bladder and bowel [Figure 2a and b]. It also revealed partial sacral agenesis; [Figure 3] and it is probably presacral cystic teratoma. A fecal impacted large ball was seen at the rectosigmoid junction with mild dilatation of proximal large bowel in the left lower quadrant [Figure 2a and b]. On surgical examination, we found a large presacral thick fluid filled cystic lesion. It was firmly adherent to pre-sacrococcygeal region.

The cyst contents were aspirated and lesion was curetted from sacrococcyx [Figure 4]. The rectosigmoid fecal ball was withdrawn per rectal [Figure 5a and b].

Discussion

Sacral agenesis in a triad of anorectal malformation, sacral bony abnormality and a presacral mass (which may be a meningocele, a teratoma or an enteric cyst) is known as Currarino syndrome.[4] In our case no significant anorectal malformation was identified.

In over 80% of cases, presentation of Currarino syndrome occurs in infancy or childhood[5] Symptoms such as intractable constipation, bowel obstruction in infancy, urinary retention, incontinence, and infection are frequently associated with this condition. Although rare, there have been reports of the syndrome present in adulthood. In females, additional symptoms such as dysmenorrhea, obstructed labor, perianal sepsis, or low back pain may also constitute the initial complaint.[6] Patients with Currarino syndrome are frequently present with other anomalies including hydronephrosis, vesicoureteric reflux, duplex ureters, congenital single kidney, urinary incontinence, voiding dysfunction, tethered cord, bicornuate uterus, subseptate vagina, and rectovaginal fistulas.[6] Our patient did not have kidney, ureter, and urinary bladder anomalies.

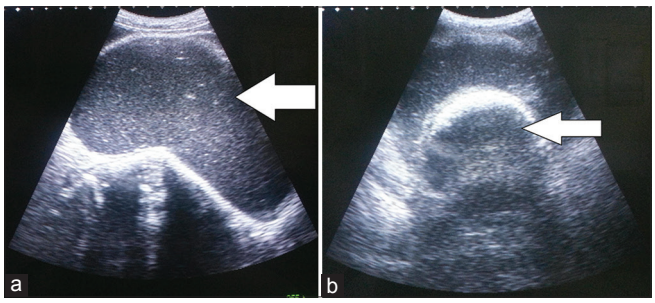

Figure 1: (a) USG image of pelvis showing a large well defined low level echogenic cystic lesion (arrow). (b) USG image of pelvis showing another adjacent rounded echogenic lesion (arrow) with distal acoustic shadowing

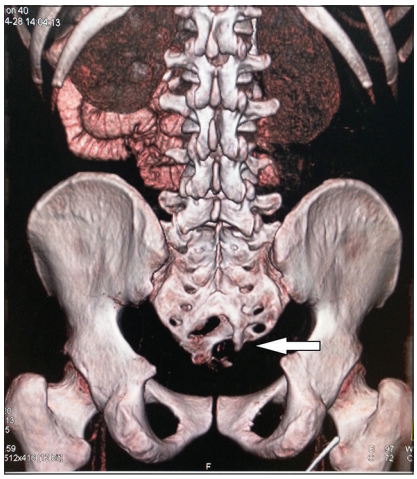

Figure 3: Inspace CT image revealed partial sacral agenesis (arrow)

In their review of the literature, Lynch and associates found that 16% of patients with this disease presented with bowel obstruction in infancy.[7] In those, who do present with this disease in adulthood, diagnosis is usually made in 1 of 3 situations.

However, in our own review, we did not find any adults presenting with a massive fecolith, with large presacral mass without anorectal anomalies similar to our patient. Therefore, we believe that this type of presentation in adults is quite rare.

The syndrome, recently linked to the HLXB9 homeobox gene, is familial in approximately half of all cases and often demonstrates in an incomplete form. Family screening has been suggested as a routine part of management to allow identification of asymptomatic family members at risk.[6,7] Our case does not have family history of such type of syndrome.

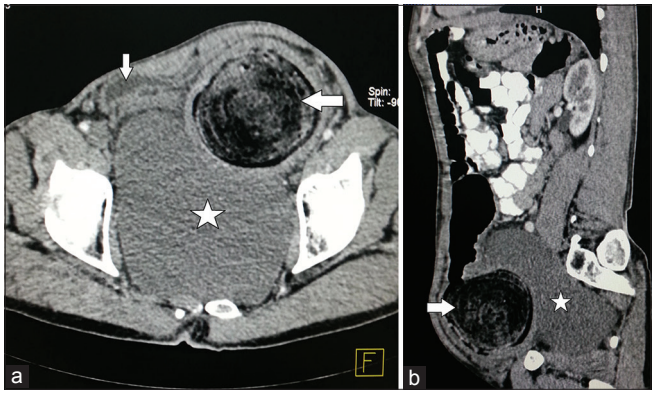

Figure 2: (a) CT axial image of pelvis showing a large high dense fluid filled well defined cystic lesion in pre-sacral and retro-rectal area (star) with faecal impacted large ball (horizontal arrow) and right anterior-superior shifting urinary bladder (vertical arrow). (b) CT sagittal image of pelvis showing a large high dense fluid filled well defined cystic lesion in pre-sacral and retro-rectal area (star) with faecal impacted large ball (arrow)

Figure 4: Photograph of curetted presacral mass from sacrococcyx after aspirated cyst contents

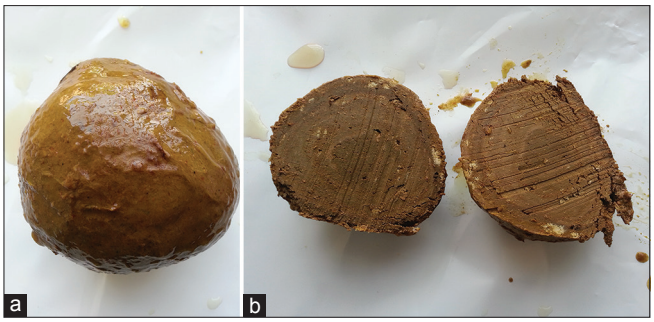

Figure 5: (a) Photograph of the rectosigmoid faecal ball. (b) Photograph of mid-cut section of the rectosigmoid faecal ball

When present, bowel obstructions associated with Currarino syndrome have been variably attributed to spinal cord tethering, obstruction from the presacral mass or obstruction due to rectal malformation.[7] Our patient did not suffer from a tethered cord and severe anal stenosis. There was a no fistula between the presacral mass and the colon. Although this patient was no focal neurologic deficit affecting a segment of his colon even though his bladder and sexual functions were spared. In our case, rectosigmoid impacted large fecal ball formation due to obstruction of bowel by large pre sacral mass. During surgery, it appeared that the large presacral mass was causing a blockage of the colon. There was pressure exerted by the presacral mass on the colon.

Regardless of the direct cause of constipation associated with Currarino syndrome, it is generally agreed that the majority of patients with this syndrome require surgical management to resolve the bowel symptoms. Most authors recommend resection and excision of the presacral mass in order to (1) Avoid the possibility of malignant degeneration of a teratoma, although this is a very uncommon occurrence;[5-9] (2) avoid infection and meningitis arising from an existing rectal–presacral mass fistula, an accidental presacral mass rupture, or an accidental presacral mass, puncture,[4,10-12] and (3) relieve external pressure on the rectum.[4,7,13]

However, surgical excision poses several risks including postoperative infection, meningitis and nerve damage resulting in possible bladder and sexual dysfunction.[4,5,14,15] Proper treatment of Currarino syndrome varies according to the severity of the case. It has been suggested that disease of milder severity can be successfully managed by conservative treatment.

Given our patient's age at presentation, the absence of a fistula between the rectum and the presacral mass, and the apparent of external pressure from the mass, we considered he was suffering from a severe form of the condition. Also, the statistics provided by others illustrate that our patient had little risk of malignant degeneration.[5-9]

Therefore, we believe that conservative treatment, including resection of the pre sacral mass with withdrawal per rectal faecal ball from rectosigmoid colon, was warranted. As demonstrated by the outcome of his management, this can result in improved bowel function.

Conclusion

We report this case in order to increase awareness of occult cause of constipation not only in pediatric age group; but also in adulthood. When approaching cases of long-standing refractory constipation, fecal impaction or a family history of similar problem should be considered. Identification of this allows both therapeutic and prophylactic management of anomalies.

Imaging in the form of CT or magnetic resonance imaging (MRI) should follow detection of sacral bony abnormalities on plain film to aid identification of associated presacral masses. In such cases like this, laprotomy and large presacral mass resection, as well as consultation with relevant disciplines regarding management of the presacral mass is recommended. Continued follow-up is needed to better characterize the long term outcome of surgery.

Here, we described an adult man, who presented with large presacral mass, sacral agenesis and a large recto-sigmoid fecolith; which was post surgically correlated.

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