



## Sacrococcygeal teratoma associated with hindgut duplication: A case report & review of literature<sup>☆</sup>



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### ABSTRACT

The index case report describes a previously unreported coexistence of sacrococcygeal teratoma and a tubular duplication of the rectum. The child presented with a large sacrococcygeal mass. He was treated with pre-operative chemotherapy to reduce the bulk of the tumor and subsequently underwent surgical resection. Exploration surprisingly revealed an additional long tubular duplication of the rectum, which did not share any lumen with the parent gut. The duplication cyst and the residual tumor were excised. The histopathology report confirmed residual mature teratoma and the duplication cyst. Relevant literature is reviewed.

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### 1. Case report

A 2 year old boy presented with a large mass in the lower back, which was diagnosed as a sacrococcygeal teratoma. The alpha fetoprotein (AFP) level was markedly raised (55,279 ng/ml). CT scan (Fig. 1) showed a predominantly pre-sacral mass (Altman type 3) approximately 11 cm × 6 cm × 5 cm in size, pushing the bladder and rectum anteriorly, with loss of fat planes with the rectum. The patient initially received 3 cycles of cisplatin based chemotherapy with very good response. Hence, he received 2 more cycles. The post chemotherapy AFP level dropped to 2.66 ng/ml. A repeat CT scan showed significant decrease in the size of the tumor to approximately 4 cm in diameter, with compression but preservation of the fat plane between the tumor and recto-sigmoid colon. Hence, he was posted for surgical excision. While excising the coccyx during surgery, a segment of bowel measuring 8 cm × 1.5 cm was identified just posterior to the rectum. The initial suspicion was that this was the rectum, which was being pulled up. However, on careful dissection and palpation, an independent segment of bowel was identified. It had an independent mesentery with blood supply, and was closed at both ends, without any

communication with the rectum (Figs. 2 and 3). The segment was completely excised (Fig. 4), as was the residual tumor, along with the coccyx. The histopathology report showed a mature teratoma with no viable malignant cells seen in the presacral mass, and a colonic type of mucosa in the duplication cyst (Figs. 5 and 6).

### 2. Discussion

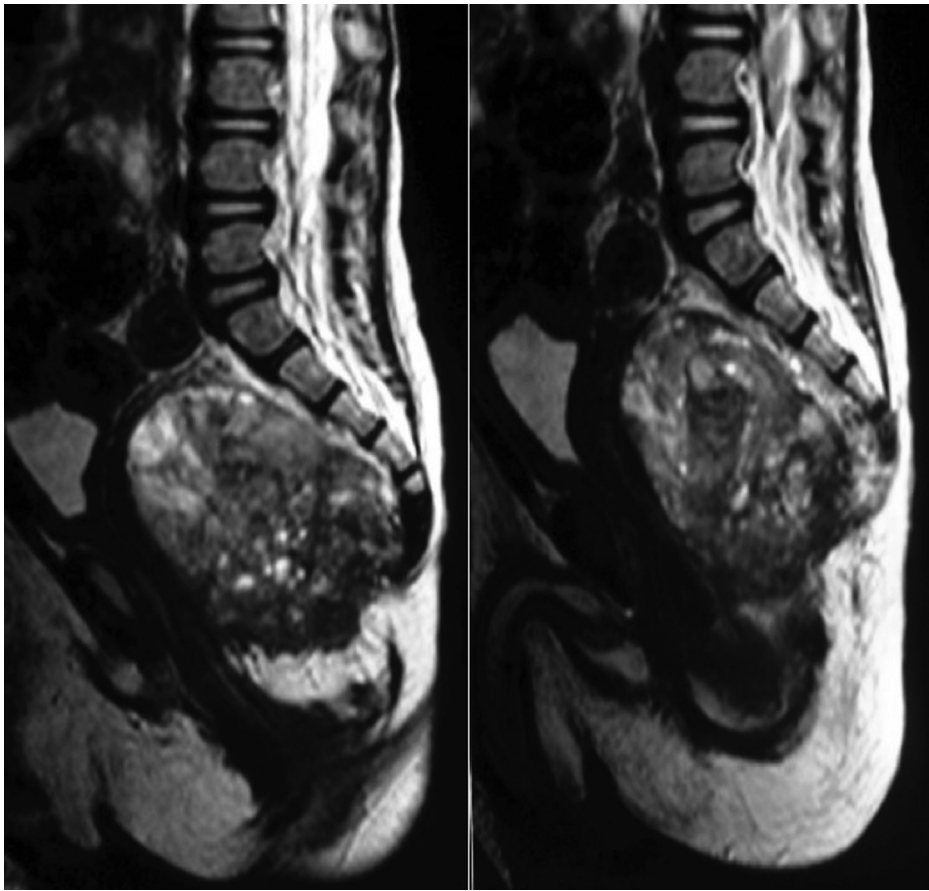
Sacrococcygeal teratomas (SCTs) present with large, predominantly external lesions at birth, and with obstructive symptoms in presacral growths. These comprise of 70% of all teratomas in childhood with a female preponderance [1] and seem to be originating from primordial pluripotent germ cells. Polyhydramnios is seen in 20% of cases antenatally [2] and nearly 80% can be diagnosed by an antenatal USG scan [3]. Incidence of malignancy is 10% at birth [4] and it increases with the delayed passage of time to treatment. Prognosis depends upon the size, age at diagnosis and treatment, histological type and complete removal of coccyx. Failure to remove the coccyx leads to a recurrence rate of 30–40% [5]. Recurrence after resection varies from 2 to 35% [6] and this may result from incomplete surgical excision with the presence of microscopic residues and tumor spillage.

The incidence of various congenital malformations associated with sacrococcygeal teratomas ranges from 5% to 26% [5,7,8]. Of these, anorectal and genital malformations are of prime concern. The association of sacrococcygeal teratoma with anorectal malformations was described as early as 1935 [9]. During the third week of

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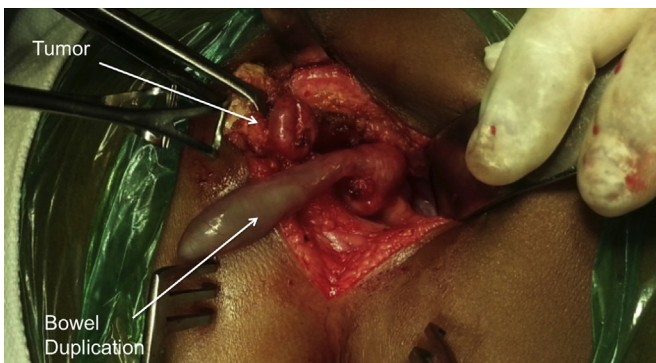
**Fig 1.** CT scan showing a Sacrococcygeal Teratoma with a large presacral component.

embryonic life, the genital folds unite to form the genital tubercle by migration of mesenchymal cells from the primitive streak region around the cloacal membrane. Between the fourth and seventh weeks, the cloaca is subdivided by the urorectal septum to form the anorectal canal and the primitive urogenital sinus. A growing sacrococcygeal teratoma at this stage of intrauterine life could thus encroach between the layers of the cloacal membrane and prevent descent and fusion of the urorectal septum to the cloacal membrane, resulting in a high anorectal malformation with a rectourethral fistula. The physical presence of a teratoma could also prevent fusion of the genital folds, resulting in a bifid scrotum and/or hypospadias [9].

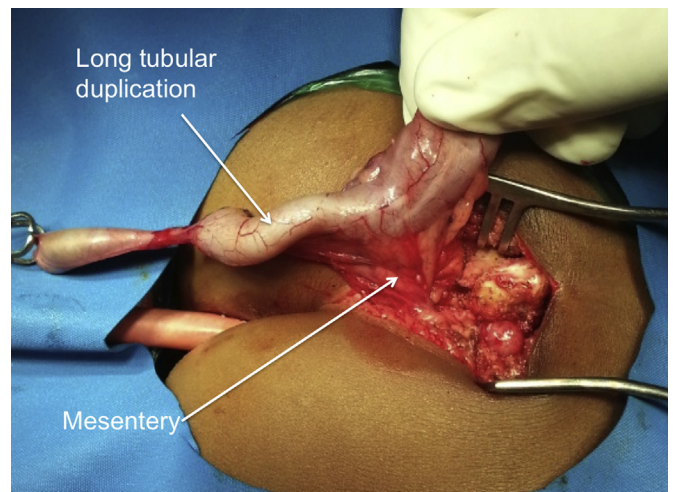
Other associated congenital anomalies include spinal dysraphism, sacral agenesis, dislocation of the hips caused by a large

tumor and meningocele [10]. Rarely, cardiac anomalies such as a ventricular septal defect or gastrointestinal anomalies other than imperforate anus have been described [11]. Vogl and Riel reported a case with anorectal malformation, sacral dysplasia, and a presacral mass which has been described as Currarino's triad [12,13]. Lahdenne et al. reported vertebral abnormalities in 80% of their 45 patients with benign sacrococcygeal teratoma [14].

Duplications of the colon and rectum constitute about 17% of all enteric duplications. Hindgut duplications associated with SCT have



**Fig 2.** Duplicated bowel segment seen below the coccyx.



**Fig 3.** Duplicated bowel segment delivered during dissection.





Fig 4. Resected specimen of duplicated bowel showing both ends blind.

never been reported. Embryological genesis of duplications suggest that these may arise as an attempt of abortive twinning and the pluripotent germinal cells may have a role to play in the occurrence of both the anomalies at the same time of embryological calendar. SCT then could represent an abortive attempt toward the formation

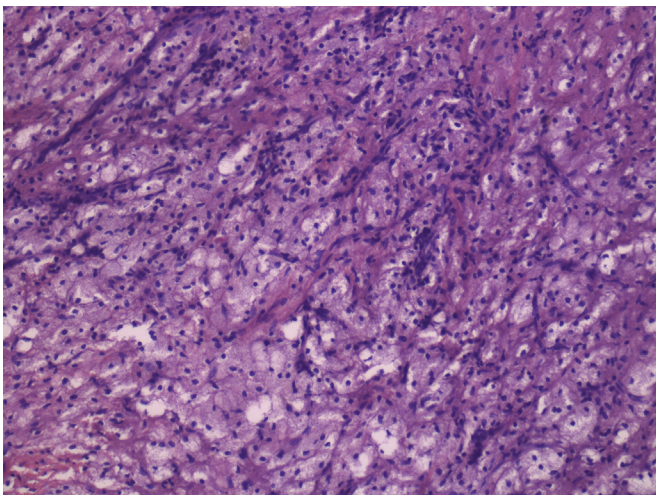


Fig 5. Histopathology confirming mature teratoma.

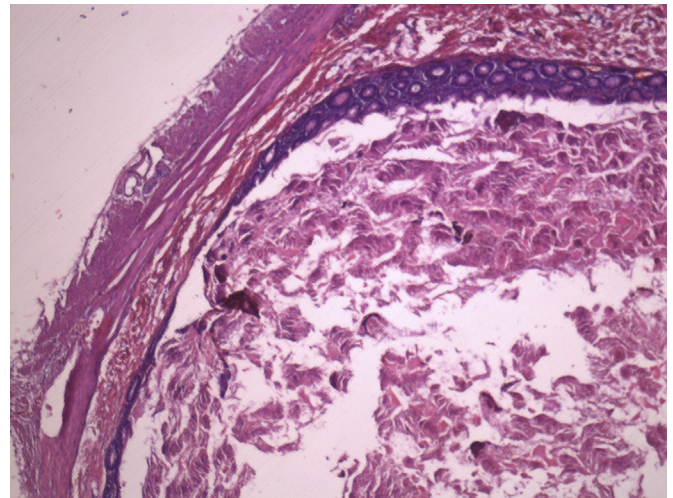


Fig 6. Histopathology of duplication cyst showing colonic mucosa.

of fetus in fetu and the rectal duplication is also an event indicative of failed twinning. They have also been reported in conjoined twins [15,16]. Colonic duplications contain colonic mucosa and on rare occasions gastric mucosa may be found within.

Owing to the anatomical location of the duplications in the presacral region, they can lead to urinary and gastrointestinal obstructive symptoms and have even been reported to mimic sacrococcygeal tumors such as teratoma or dermoid cysts [17]. A cystic mass may be present or rectal obstruction may get precipitated. They can be confused with perirectal abscesses and fistulas in the event of infections and can even lead to bleeding and prolapse.

The Diagnosis of hindgut rectal duplications may be difficult and may be missed even after thorough investigations [18] as it happened in the index case. Short or cystic duplications can be diagnosed only at the time of surgery. In the index case, the tubular duplication did not share any blood supply or wall with native bowel. The patient is being followed up with serial monitoring of AFP levels and repeat CT scans.

A thorough and diligent review of literature failed to reveal any previously reported cases of tubular intestinal duplication associated with an SCT. The case report also marks the significance of the embryological link of abortive twinning with respect to intestinal duplication and mature teratomas.

### 3. Conclusion

Sacrococcygeal teratoma has been shown to be associated with a variety of genital and anorectal malformations. However, the presence of a tubular duplication of the hindgut along with an SCT is previously unreported in English literature. The aim is to highlight the significance of the embryological link of abortive twinning with intestinal duplication and mature teratomas. This association is difficult to predict on preoperative imaging studies.

### Conflict of interest

None.

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