Case Report

**Congenital Split Anal Sphincter Associated With Perineal Lipoma - A Rare Anorectal Malformation**

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**Abstract**

An 8-month-old female presented with an absent normal anal opening. She had a small opening located in the vestibule through which she passed stool. There was a midline perineal lipoma that had split the anal sphincter complex. She also had associated vaginal duplication, dermal sinus and spina bifida. Surgical repair was done by excising the midline lipoma, mobilizing the rectum and approximating the sphincter complex on either side. Post operatively, the child was continent and could contract her rectum. During follow up, the child developed secondary rectal ectasia (dilatation) due to anal stenosis for which anoplasty was done. She is continent for stool at 8 years follow up.

**Keywords**: Anorectal Malformation; Split Anal Sphincter; Perineal Lipoma; Partial Vaginal Duplication

**Introduction**

The appropriate positioning of the rectum within the anal sphincter complex is most crucial for continence. In rare situations where the sphincter complex itself is split, the management becomes challenging. Here we present a rare case of anorectal malformation where the split sphincter complex was associated with an intervening perineal lipoma and partial vaginal duplication.

**Case Report**

An 8 month old female presented with history of a small abnormally located anal opening. On examination, she had a perineal fistulous opening in the vestibule, placed on the left side, with a perineal lipoma in the midline between the split sphincter complex (Figure 1A,B). She also had partial vaginal duplication and a dermal sinus on the sacral region. Barium enema showed dilatation of recto sigmoid with narrowing of the rectum (Figure 1C). Magnetic resonance Imaging (MRI) of the spine depicted an intraspinal lipoma.

After proper bowel preparation, a single stage posterior sagital anorecto-vaginoplasty (PSARVP) with excision of perineal lipoma, approximation of sphincter and excision of dermal sinus was done (Figure 1D-F). A thick vaginal septum was present. The mucosa of left vagina was excised and the raw surface was approximated.

Postoperatively, she had mild dehiscence of the perineal body that healed with local care. The child was continent till 15 months follow up with good cosmetic result. Thereafter, the child developed mild constipation that was relieved with laxatives. At 18 months follow up the child required occasional suppositories and enemas. Barium enema suggested rectal ectasia. She was kept on dilatations to take
care of the anal stenosis. At 36 months follow up, she developed severe constipation and the Barium enema was suggestive of persisting rectal ectasia and anal stenosis. An anoplasty was done at 40 months follow up. The child responded well to the anoplasty and started passing stool once a day without any laxatives, 15 days after the anoplasty. She remains continent at 8 years follow up. No intervention has been done for the intraspinal lipoma as she does not have any symptoms due to it.

**Discussion**

The surgical principle of repairing an anorectal malformation involves proper central positioning of the rectum within the anal sphincter complex to attain continence. The anus develops by fusion of the anal tubercles and an external invagination, known as the proctodeum, which deepens toward the rectum but is separated from it by the anal membrane. Presence of an intervening tissue like a perineal lipoma may distort the sphincter anatomy. In the case described, the lipomatous tissue might have split the developing sphincter complex and caused it to split on both sides of the lipoma.

Though the diagnosis of a perineal lipoma is easy to make on inspection, yet this curious entity is not so common [1,2]. A large retrospective review identified only ten cases of perineal lipoma in more 2000 patients of ARM [2]. Recently, the entity has been diagnosed in the prenatal period on ultrasonography [3]. The differential diagnosis is a vascular malformation, infantile hemangioma, lipoblastoma, hamartoma, choristoma and ambiguous genitalia [2-5]. Perineal lipomas may arise from the midperineum but are usually lateral in location [3].

The association of a split anal sphincter as described here has not been reported earlier. While most cases in females are associated with anorectal malformation, those in males may be associated with scrotal anomalies and penoscrotal transposition [6-9]. Perineal lipoma occurring in isolation only requires simple excision [10].

The presence of unusual perineal masses in association with anorectal malformation adds to the complexity of the repair. The lipoma has to be removed carefully from between the muscle fibres while taking care to preserve the muscle complex during posterior sagittal anorectoplasty [2].

Though the case reported had good continence initially, thereafter she developed mild constipation requiring bowel management and later anoplasty. Others have also reported that the bowel function seems to be compromised in these lesions [1]. The case described here was managed without a colostomy. In a series of 6 cases of perineal lipomas, all patients had colostomies; 2/3 evaluated for bowel function required antegrade continent enema stoma to control constipation and soiling while 1 had soiling despite regular washouts [1]. In another large series, at a follow-up of 5 months to 12 years, 6 of the 10 lipoma patients were continent [2].

In the case reported, following anoplasty at 40 months follow up, the child is fully continent without any medications at a follow up of eight years. To conclude, careful delineation of the anatomy is important to successfully manage complex anorectal malformations.

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