Congenital perineal hernia

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A R T I C L E  I N F O
Keywords:
Perineal hernia
Pelvic hernia
Congenital
Rare hernia
Rectal hernia

A B S T R A C T
Perineal hernias can occur primarily or secondary to a pelvic operation. Among primary perineal hernias, the congenital type is very rare, with less than 10 cases reported in the literature. This is a case report of a healthy female child presenting with a congenital posterior perineal hernia who underwent successful delayed operative repair using a perineal approach.

Level of evidence: Level V – case report.

1. Background

Perineal hernias result from weaknesses or defects in the pelvic floor musculature. They can be classified as congenital or acquired [1]. Acquired perineal hernias can be further subdivided into primary and secondary categories [1]. Primary acquired perineal hernias are associated with factors causing elevated intra-abdominal pressure, while secondary perineal hernias occur after pelvic operations such as abdominoperineal resections [2]. Anatomically, we distinguish anterior perineal hernias, or those that arise from defects in the urogenital diaphragm anterior to the paired superficial transverse perineal muscles, from posterior hernias, which occur as a defect within the levator ani muscles constituting the pelvic diaphragm or through defects resulting from failure of obturator internus and ilio coccygeus muscles to join [2]. Anterior perineal hernias occur in females only, while posterior perineal hernias may occur in both sexes [2]. We report a case of a rare congenital perineal hernia which was successfully repaired after delayed presentation.

2. Case report

A healthy 5-year-old female child was referred to surgical care for management of a symptomatic posterior perineal hernia. A left pericoccygeal bulge and buttock asymmetry had been noted by the mother since birth. She had been evaluated for this problem as a baby; however, she was lost to follow-up at the time. The patient had now reached an age where this bulge was disturbing her during ambulation and occasionally caused discomfort. The diagnosis of congenital perineal hernia was made based on clinical history and physical examination findings.

After informed consent was provided, the patient was booked for surgical repair. The patient was brought to the operating room and general endotracheal anesthesia was induced. She was placed prone on the operating table. A visible buttock asymmetry was noted, and a left-sided pericoccygeal defect was palpable, which occurred between the anus and the ischial tuberosity, suggesting a lower posterior perineal hernia (Fig. 1). On digital rectal examination, one could feel a defect in the 10 o’clock position and recreate protrusion of the upper rectum through this space. The defect was inferolateral to the sacroiliac junction, and medial to the ischial tuberosity (Figs. 2 and Fig. 3).

A 3-cm incision was made obliquely, along Langer’s lines of skin tension, above the bulge. This was carried down to the level of the pelvic diaphragm, where the defect was appreciated at the interface between the ilio coccygeus and the coccygeus muscles, at the lower border of the gluteus maximus. The hernia sac was dissected carefully off the edge of the defect and reduced. The defect was closed primarily in a simple interrupted fashion using a combination of 2-0 Ethibond® and 2-0 PDS-II® sutures. The skin was closed in a running subcuticular fashion using 5-0 Monocryl® sutures (Fig. 4). A transparent Tegaderm® dressing was applied over the wound after the skin was cleaned with a wet and dry sponge. The patient tolerated the procedure well, was extubated in the operating room and transferred to the recovery room in stable condition. She returned to the surgical ward shortly afterwards.

The patient’s in-hospital stay was uneventful. She resumed normal diet and activity right away. Her pain was well controlled with paracetamol. On post-operative day 2, her dressing was removed and she
was discharged home with planned follow-up in the surgical outpatient clinic.

At follow-up, the patient had no complaints and the wound had healed well. There was no evidence of surgical site infection, and the buttock asymmetry had resolved. The defect was no longer palpable. The patient had returned to school and was participating in all normal activities, without discomfort or limitations.

3. Discussion

Congenital perineal hernias are very rare, with only 10 cases described in the literature. Eight of those cases reported perineal hernias in children [3–9], while the other two described the association of congenital perineal hernias with chromosomal abnormalities in aborted fetuses, including Turner Syndrome and Trisomy 18 [10,11]. Not surprisingly, these patients can pose a diagnostic challenge to the surgeon who has not been exposed to this rare entity.

This type of hernia has been reported to present as a mass in the labium majus, a mass below the lower margin of gluteus maximus or a swelling between the anus and the ischial tuberosity, as was identified in this case, depending on the location of the defect [2]. Repair is indicated in congenital hernias or those who develop symptoms such as skin erosion, discomfort, pain, bowel obstruction or urinary obstruction [2,12–14]. Three surgical approaches have been described, including a trans-abdominal approach, a perineal approach, or a combined approach [2,13,15,16]. The transabdominal approach is preferred when laparotomy is being performed for another reason or for recurrent hernias. However, this is associated with less morbidity than the perineal approach, which avoids entering the peritoneal cavity. However, mobilization of adherent or incarcerated intra-abdominal organs is more difficult using the perineal approach, which is usually preferred for congenital perineal hernia repair. The combined approach offers the advantages of a better exposure and more robust pelvic musculature reconstruction, while allowing for reduction of herniating contents under direct vision. However, this procedure is much more extensive and associated with higher morbidity [2,12,13]. Laparoscopic techniques have been described more recently in cases of postoperative perineal hernia after abdominoperineal resection in adults [17–20].
4. Conclusion

Congenital perineal hernias are rare entities with very few reports present in the literature. It is feasible to repair these through a perineal incision with good results, even when the patient presents in a delayed fashion.

Conflict of interest

All authors declare no conflicts of interest.

Funding

No external funding was received for this work.

Acknowledgement

We gratefully acknowledge the contribution of Mr. Matt Evans and the McGill University Health Center Patient Education Office for their assistance with the creation of the illustration in Fig. 3.

Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.epsc.2018.01.016.

References


Fig. 4. Restored buttock symmetry following perineal hernia repair.