CASE REPORT OPEN ACCESS

Congenital Bilateral Spigelian-Cryptorchidism Syndrome? A Case Report

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ABSTRACT

Background: There is ongoing debate on nomenclature of congenital Spigelian hernia (SH) associated with undescended testis (UDT) in infants due to its frequent reporting in last two decades despite rarity of the condition. It is commonly reported as unilateral entity. To the best of our knowledge, only 4 bilateral cases have been reported.

Case Report: We report a case of bilateral congenital Spigelian cryptorchidism syndrome in a small infant who was dealt surgically with good outcome.

Conclusion: The main differentiating point from inguinal hernia is direction of herniation in SH is laterally and upwards while it is downwards towards scrotum in inguinal hernia.

Key words: Congenital Spigelian hernia; Undescended testes; Bilateral cryptorchidism

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INTRODUCTION

Spigelian hernia is (SH) caused by a defect in abdominal wall in semilunar line above or below the umbilicus.[1] It is a well-known entity in adults and usually post-traumatic but rarely reported in children. SH when associated with an undescended testis (UDT) is thought to be congenital and reported in 53% of SHs in a recent review. The frequency of reporting is increased in last 2 decades.[2] This report focuses on surgical findings in a recently managed case of bilateral SH with UDT to help answer queries regarding nature of this association. This is the 5th bilateral case in English literature to best of our knowledge.

CASE REPORT

A 28-day-old male infant presented to us with complain of bilateral abdominal swelling on crying and bilateral empty hemiscrotum noticed since birth. He was product of consanguineous marriage, pre-term (35 weeks); delivered by Cesarean section done for fetal distress. Antenatal ultrasounds were unremarkable for fetal anomalies. Postnatal course was significant for physiological jaundice for which he presented to a tertiary care center at 10 days of age where he was advised conservative treatment for jaundice and follow up after 6 months for bilateral impalpable testes. No remark of hernia was made. On physical examination, there were prominent bulges present bilaterally in iliac regions, right being larger than left and hypoplastic scrotum with a normal appearing uncircumcised penis (Fig. 1A). On palpation, right hernia contained reducible bowel while left contained reducible gonad. The baby also had right incomplete and left complete simple syndactyly of third web spaces of hands. Due to unusual location from inguinal hernias, a literature search was done which revealed it

to be Spigelian hernia. Ultrasound examination showed both testes within hernia sac.

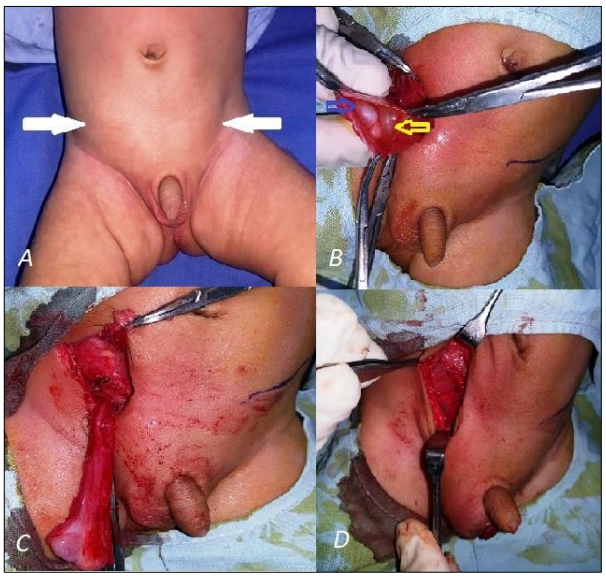

Figure 1 A) White arrows showing right and left Spigelian hernias and bilateral empty hemiscrotum. B) On opening sac, blue arrow shows right testis (no gubernaculum), yellow arrow shows small bowel loop. C) showing closure of hernia loose enough for exit of spermatic cord. D) Showing completed repair of external oblique and subcutaneous orchiopexy.

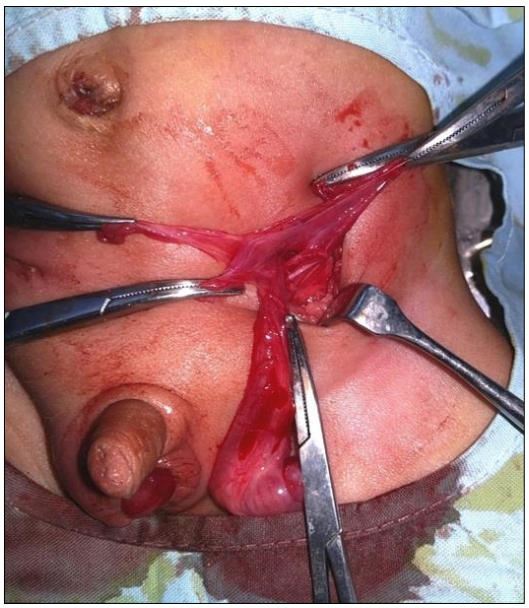

Figure 2: Showing same procedure done on left side. Note enough cord length.

Due to concern of bowel incarceration in previous reports, we decided to operate upon him at 35 days of age. On exploration on right side, the hernia sac was found deep to external oblique aponeurosis. On opening it was found to contain a small bowel loop and right testis (Fig. 1B). The defect was near 1.5x1.5 cm. After reducing bowel, the processus vaginalis was separated from cord structures and purse string ligature (4/0 vicryl) applied with excision of redundant sac (Fig. 1C). The spermatic cord had enough length for tension free orchiopexy through subcutaneous route (Fig. 1D). The external oblique fascia was closed with 4/0 vicryl suture in interrupted fashion and caudal most stiches not applied to prevent strangulation of exiting spermatic cord. A subcutaneous tunnel created with hemostat and testis brought in scrotum and fixed in subdartos pouch without constraint. We did not specifically look for inguinal canal. Same procedure was done on left side (Fig. 2).

Important findings were absence of gubernaculum on both sides as shown and sufficient cord length. Postoperative course was uneventful in terms of infection and atrophy and he is doing well at 2.5 month follow up.

DISCUSSION

SH occurs in spigelian zone (also known as semilunar line) at any point along its length and is defined as zone of transition between lateral border of rectus sheath and aponeurosis of transversus abdominis.[1] It is usually found in adults as a result of increased intra-abdominal pressure or trauma. In pediatric population it is considered congenital when it is associated with cryptorchidism.[3]

There is considerable controversy regarding nature of association of UDT with SH due to coexistence of two anomalies in surgical field but limited number of cases reported to statistically prove an association between them. Nevertheless, the surgeon is tempted to speculate whether UDT caused SH or SH has led to UDT. Previously held view was that hernia occurred due to weakness in abdominal wall and testis takes path of least resistance and follows hernia as in exomphalos and gastroschisis but it fails to explain all ipsilateral cases of SH and UDT. [4] Other proposal was that the testis was ectopic in abdominal wall and dragged with it processus vaginalis which turns into hernia with increased abdominal pressure. However, almost all cases of ectopic testis are attributed to mis-attachment of gubernaculum; which is not found in all reported cases of SH/UDT.[5] Another speculation was made by Rushfeldt et al where he said that absence of gubernaculum led to absence of inguinal canal and testis used SH as escape point.[6] However reason of coexistence of both anomalies cannot be explained by this hypothesis and inguinal canal cannot be absent since it is present before testicular descent in both genders. [7] The term syndrome here is

unjustified since mere coexistence of SH and UDT is not enough but a known or presumed common cause has to be present. None of the hypotheses described above point towards a common cause; rather each try to explain whether SH caused UDT or vice versa. [8]

Bilateral SH/UDT are exceedingly rare and only four cases have been described. [2] First bilateral SP+UDT was reported in 2003 in which the 1 month old infant presented with incarcerated right hernia and operated emergently. The bowel was saved and right orchiopexy was done without details of gubernaculum or inguinal canal. Left side was operated at 10 month of age. [9] In two cases of Durham, one patient underwent staged repair; right hernia mesh repair with orchiopexy at 13 months of age followed by left hernia repair without mesh and orchiopexy at 16 months. Both testes were brought in scrotum through new internal rings medial to inferior epigastric vessels. The other patient underwent single stage bilateral orchiopexy at 14 months with same technique. They reported absence of gubernacula and inguinal canals in both cases. [10] Fourth case from Italy was first neonate to be operated upon had associated Aplasia cutis. He underwent bilateral extra peritoneal orchiopexy and the authors did not comment on gubernaculum and inguinal canal. [11]

Our case concurs with surgical findings of other reports where gubernaculum was not found attached to testis. However we have not looked for inguinal canal since it would have needed additional exploration of hernia defect which we cannot do since it has ethical implications.

In conclusion, the pathogenesis of congenital SH and UDT remains elusive and due to frequent coexistence, some authors have called it a syndrome. Accurate observation of clinical findings can lead to correct diagnosis and surgical management of this rare condition even in a secondary care hospital. The main differentiating point from inguinal hernia is direction of herniation in SH is laterally and upwards while it is downwards towards scrotum in inguinal hernia. Till significant association is found it is better to term it “coexistent SH and UDT” rather than syndrome.

Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

Authors’ Contribution: All the authors contributed fully in concept, literature review, and drafting of the manuscript and approved the final version of this manuscript.

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