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Congenital Spigelian hernia and ipsilateral cryptorchidism: a new syndrome?

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Abstract: Spigelian hernia (SH) is a rare ventral interstitial hernia occurring through a defect in the transversus abdominis aponeurosis (Spigelian fascia). Spigelian fascia is found between the lateral border of the rectus abdominis muscle and the semilunar line, which extends from the costal cartilage to the pubic tubercle. In other words, Spigelian line is where the transversus abdominis muscle ends in an aponeurosis characterized by a congenital or acquired defect in the Spigelian aponeurosis. Pediatric cases of SH are either congenital or acquired due to trauma, previous surgery or increased intra-abdominal pressure. SH in combination with ipsilateral cryptorchidism may constitute a new syndrome, as such cases are extremely rare in the literature This new syndrome is characterized by the following congenital, ipsilateral disturbances: SH, absence of inguinal canal and gubernaculum and the homolateral testis found within the Spigelian hernia sac (a hernia sac containing undescended testis). The aim of this study is to emphasize some typical findings of this specific entity, and, hence, the necessity for a thorough investigation of the origin of the SH.

Key words: Spigelian hernia, transversus abdominis aponeurosis (Spigelian fascia), arcuate line or linea semicircularis (Douglas), cryptorchidism, child.

Background

SH is a rare condition, characterized by a ventral interstitial hernia occurring through a slit-like defect in the transversus abdominis aponeurosis (Spigelian fascia) [1]. Compared to other types of hernias, SH accounts for 1–2% of all hernias, while it constitutes about 0.1–0.2% of all abdominal wall hernias [2–4]. A defect in the transversalis fascia, 2–7 cm in diameter, may be found [5].

SHs occur along the semilunar line, which was first described by Adrian van der Spieghel, a Belgian anatomist (1578–1625) [2, 6]. SH was initially described by Josef Kinkosh in 1764. A typical patient's profile is of a 50 year old overweight patient with associated disease [2, 7, 8]. Its occurrence as a congenital hernia was firstly described in 1935 in infants or children and since 2012, 54 cases of SH have been reported [1, 2].

Hernia sac usually contains, apart from preperitoneal fat, in most cases testis, followed by small intestine, sigmoid colon, omentum, gallbladder, stomach, Meckel's diverticulum, ovary, while herniation of the bladder has been rarely reported in children [6, 9]. A defect in the Spigelian fascia combined with a concomitant incarceration of a hollow organ can lead to the development of Richter hernia [2].

After comprehensive research of the relevant literature in pediatric population, it has been reported that 11–16% of all cases of SHs may involve multiple hernias found ipsilaterally or bilaterally [5, 6]. In children, SHs are usually left-sided and are more frequent in males than females (M/F ratio = 2.7/1). On the contrary, in adulthood, they are most usually found on the right side and are more frequent in females (M/F ratio = 0.75/1) [10].

Surgical anatomy

The transversus abdominis muscle becomes aponeurotic at the semilunar line which extends from the ninth costal cartilage to the pubic tubercle. The aponeurosis between the semilunar line and the lateral edge of the rectus muscle is called Spigelian aponeurosis [5]. It is the layer between the rectus abdominis medially and the semilunar line laterally.

In the upper abdominal wall, the Spigelian aponeurosis is posterior to the rectus muscle. Superior to the umbilical region, the aponeurosis of the internal oblique crosses over the transverse abdominal muscle, in a perpendicular fashion. Inferior to the umbilical region, these aponeuroses run parallel to each other.

Spigelian fascia in its upper two thirds is reinforced by prolongation of muscular fibers of transversus abdominis. It's widest portion in the abdominal wall (about 6 cm) is found above the line joining two anterior superior iliac spines, and it extends above arcuate line of Douglas [2].

Spigelian fascia or Linea semilunaris is found in between the lateral border of the rectus abdominis muscle and the semilunar line, which extends from the ninth costal cartilage to the pubic tubercle. It is also found laterally to the junction of the arcuate and semilunar lines and is formed by the fibrous union of the rectus sheath and the anterior abdominal wall muscles below the level of the umbilicus.

Etiology

SH can be congenital or acquired. Although SHs in adulthood are considered to be acquired due to trauma, previous abdominal surgery or increased intra-abdominal pressure, cases found in pediatric population are highly suspected as congenital [11]. SHs develop due to defects in the Spigelian fascia; the aponeurosis of the internal oblique crosses over the transverses in a perpendicular way. Inferior to the umbilical region, these aponeuroses run parallel to each other and these parallel fibers form a weak barrier that is susceptible to protruding peritoneal sacs or extraperitoneal fat. These hernias almost always develop at the level or below the arcuate line, possibly due to the lack of posterior rectus sheath at this level.

SH is due to a congenital deformity in the development of the abdominal wall because of a structural change of the internal oblique and transversus abdominis muscles, neurovascular openings in the fascia, infiltration of the abdominal wall layers with preperitoneal fat and muscle paralysis [5, 12].

Concomitance of congenital midline defects or abdominal wall defects such as gastroschisis, umbilical hernia, omphalocele, bladder or cloacal exstrophy with SHs in pediatric population reinforce the above mentioned theory [13].

In the field of genetics, defective elastic fiber synthesis due to deletion of gene 7q1123, collagen synthesis disorder due to gene 2 defect and other co-existing genetic disorders, such as Williams syndrome, predispose to the development of SH [14].

Acquired Spigelian hernias have a real risk of strangulation due to sharp Spigelian fascial margin around the defect developed because of trauma, previous operation, chronic increased intra-abdominal pressure, constipation, imperforate anus, ascites and ventriculoperitoneal shunt, to name but a few predisposing factors.

Cryptorchidism assosiated with Spigelian hernia

After thorough investigation of the relevant literature, it has been suggested that SH is associated with ipsilateral cryptorchidism at about 28%–75% of male pediatric patients [4–6, 12, 15]. The testis is found within the hernia sac in 87% of those patients [4].

SH combined with ipsilateral cryptorchidism may be part of a new syndrome, as such cases are extremely rare in the literature This new syndrome is characterized

by the following congenital, ipsilateral disturbances: SH, absence of inguinal canal and gubernaculum and the homolateral testis found within the Spigelian hernia sac (a hernia sac containing undescended testis) [4, 16].

The underlying mechanism leading to the development of this syndrome remains unknown, triggering a vivid discussion in the relevant literature. The first indication may be an ipsilateral ectopic testis and the subsequent conversion into Spigelian hernia or the absence of a gubernaculum.

Raveenthiran and colleagues attempted to interpret the development of a low SH by considering the ectopic position of the testis as the primary event. Then the SH develops, preventing the completion of the descend of the ipsilateral testicle, as it is believed, that in the context of increased intra-abdominal pressure, the testicle follows the path of lower resistance, in other words towards the direction of the aponeurotic defect of the Spigelian line [9, 17].

Rushfeldt *et al.* attributed the occurrence of aplasia of the inguinal canal ipsilaterally to a failure in the development of a gubernaculum, and as a consequence the testis will not be able to descend from its intra-abdominal position to the scrotum. The defect in the Spigelian fascia may be due to the arrest of the testis that induces a kind of rescue canal through a lavender area in the abdominal wall in the absence of an inguinal canal [4, 18].

Classification

The classification depends on the localization of the hernia's orifice. There are three types of SH:

- the orifice of the SH in which hernia sac protrudes is found within the fibers of the transverse abdominal muscle and the internal abdominal muscle.
- the orifice of the SH in which hernia sac protrudes is found between the external oblique muscle and the internal oblique muscle. This occurs due to the direct relationship between the aponeurotic defect with the Spigelian line and the aponeurosis of the internal oblique muscle.
- the orifice of the SH in which hernia sac protrudes is the subcutaneous fat, in the presence of aponeurotic defects in the transversus abdominis aponeurosis (Spigelian fascia), internal and external oblique aponeurosis.

According to the localization of the defect, congenital SH is classified as "low" and "high". SH develops in the fascia below the level of the umbilicus, laterally to the junction of the semilunar and arcuate lines. It lies mostly on the "SH belt". Hernias develop within this "SH" belt. Most SHs (85–90%) are found cranially and laterally to the epigastric inferior vessels, although they feature occasionally medial and caudal localization under the name "low Spigelian hernias" [2, 4, 18].

Spigelian fascia in its upper two thirds is reinforced by prolongation of muscular fibers of transversus abdominis. It averts herniation through this area. It is called "high SH" when it occurs in the upper half.

Clinical evaluation

SH manifests as a lateral parietal swelling, which may be either painful or painless with positive cough impulse. It usually lies inferior to the umbilical region, adjacent to the iliac crest (Spigelian Hernia Belt or Zone). Rarely the hernia can enter the rectus sheath [2]. Diagnosis is easy if hernia is incarcerated, causing pain. The incidence of incarceration varies significantly between various case series with high potential for life-threatening complications. Almost 50% of pre-operatively diagnosed SHs are non-incarcerated [2, 19, 20].

Differential diagnosis of SHs includes the following: rectus sheath haematoma, parietal abscess, parietal lipoma, cystic lymphangioma, mesenchymal tumor and appendicitis [2, 6, 11, 19, 20]. Right-sided tenderness or discomfort due to SH may be presumed as acute abdomen (acute appendicitis) [12].

Low SHs may protrude from a small defect of the Spigelian fascia into Hesselbach's triangle. SH can be easily misdiagnosed as ipsilateral inguinal hernia [19, 21].

The incidence of SH's strangulation may reach up to 20% of all cases, due to a small defect (<2 cm) in the transversalis fascia [6, 22]. SHs are usually narrow necked, thus, the risk of strangulation is high enough. The latter is the reason why SHs should be operated soon after the documentation of diagnosis.

Diagnostic modalities

In doubtful cases of SH, ultrasonography (US) and computed tomography (CT) can help to establish the correct diagnosis preoperatively. A meticulous imaging examination may depict the fascial defect, the hernia sac with its blood supply and its contents.

Imaging should detect the localization of the ipsilateral testicle and confirm whether it is outside the scrotum or inside the hernia sac.

Torzilli *et al.* express the opinion that US may assist in the reduction of an incarcerated SH by applying force using the US probe itself, thus avoiding urgent surgical intervention [23].

Principles of management

SHs can be treated either through conventional open surgical technique or through laparoscopic modalities, which can be either transperitoneal — so called intraabdominal — or extraperitoneal [2, 6, 24].

Laparoscopy in children is not an acceptable therapeutic alternative, when there is strangulation of the hernia. In those cases, open surgical technique is considered as the treatment of choice.

In highly suspected cases of SH, surgery under general anesthesia involves the following steps:

- an inguinal skin crease (transverse) incision is made over the protrusion
- external oblique aponeurosis is incised in order to expose peritoneal sac
- the hernia sac is opened and then its content is reduced, if this is feasible
- in male infants with SH and suspected ipsilateral cryptorchidism, a thorough investigation of the testis is indicated as this might be found within the hernia sac
- the fascia defect is closed with 1–2 sutures in layer
- the abdominal wall is ligated in layers using non-absorbable sutures in each layer
- the preperitoneal space or above the fascia is closed in layers and the skin either with tacks or with a continuous suture.

In cases of simultaneous orchidopexy bilaterally, in which exploration of the inguinal region yields no evidence of the inguinal canal and gubernaculum, a new internal inguinal ring should be conducted through the abdominal wall and the testes should be brought down medially to the inferior epigastric vessels and just laterally to the pubic tubercle (technique reported by Durham and Ricketts) [1, 25].

Inan *et al.* performed a SH with concomitant ipsilateral cryptorchidism repair in a male neonate. Postoperatively, scrotal abscess along with atrophy of the recurrent testicle developed. They believe that these intra-operative surgical complications occurred either as a consequence of vascular damage of the testicle or due to compression of the testicle at the new internal ring, or even due to increased pressure of the elements of the spermatic cord [26].

They finally conclude that surgical repair of congenital SHs should be performed in the early period of life, in which the defect in the Spigelian aponeurosis should be corrected primarily, with conduction of orchidopexy secondarily. When the testis is found within the hernia sac and spermatic cord is not large enough, then the Fowler-Stephens technique can be performed simultaneously, as a primary orchidopexy alternative [26].

In cases of investigative laparotomy conduction due to high index of clinical suspicion for acute appendicitis, without intraoperative confirmation of the preoperative diagnosis, then the surgeon should exclude the presence low SHs (Spigelian Belt or Spigelian Zone) [19].

Finally, when the diagnosis of ipsilateral inguinal hernia is not confirmed intraoperatively, the surgeon should also exclude the presence of a low SH, as in cases of presence of aponeurotic defect at the level of Hasselbach's triad, they may be misdiagnosed as inguinal hernia [20].

Conflict of interest

None of the contributing authors have any conflict of interest, including specific financial interests or relationships and affiliations relevant to the subject matter or materials discussed in the manuscript.

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