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A CASE PRESENTATION AND LITERATURE REVIEW OF RARE SPLENO-GONADAL FUSION FOUND DURING EXPLORING THE PATIENT FOR CONGENITAL HYDROCELE.

Surgery

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ABSTRACT

Spenogonadal fusion is a rare congenital anomaly in which there is a fusion of Spleen and the gonad or meso nephric derivatives. Till date approximately 200 cases is been recorded. The condition was first reported by Bostroem in 1883. Very few cases has been diagnosed pre operatively many of them are accidently found during the operative procedures. Few a times is been mis diagnosed at a malignant tumour of testis. A case is presented and reviewed.

KEYWORDS

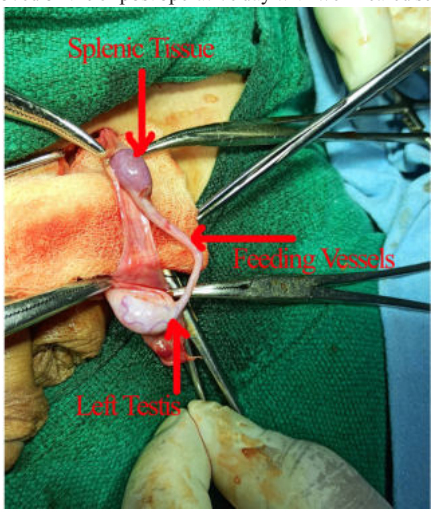
1. CASE REPORT

A 5 year old boy came to our surgery opd with complaint of left sided scrotal swelling since birth. He was delivered by normal vaginal delivery and was of normal development for his age. Physical examination was unremarkable on local examination left testis was enlarged compared to right side. Patient mother was aware of his swelling since birth but was adviced for elective procedure. No any hernia was present. Cough impulse was negative. Get above swelling was possible. Swelling was brilliantly transilluminating. No Any palpable mass was noted except the swelling. Swelling was not separately palpable. Patient was investigated he has o positive blood group with 14gm/dl of hemoglobin and normal liver function and renal function tests and was undergone ultra sound. Ultra sound was suggestive of gross amount of fluid in the left trans vaginal sac with testis showing normal size, shape, echo texture and vascularity.

On exploration left sided groin incision was placed and inguinal canal was entered and processus vaginalis was dissected, which is the hydrocele sac, freed from the vas deference and vessels. Sac was opened and examined. On examination a fleshy, smooth margined reddish-brown structure was identified posteriorly. With slight blunt dissection and traction, it was found to be lying in close relation with the testicular vessels, with an intervening plane of loose areolar tissue. It was possible to follow it retro peritoneally up to the lower pole of the left kidney using the blunt dissection. The sausage shaped structure was 3cm in length and 2cm width. On examination it had separate feeding vessels. They were ligated and specimen was retrieved completely. The procedure was concluded by the high ligation of the sac and closure. Sterile dressing was placed. Whole procedure went uneventful patient was shifted to recovery post operatively.

Histopathology was suggestive of normal splenic tissue.

Patient was discharged on the 3rd post operative day and skin sutures were removed on the 6th post operative day with well healed scar.



2. DISCUSSION

approximately 200 cases of spleno gonadal fusion have been reported since the first description of this condition by the bostroem in 1883[1]. The first detailed report was given by Pommer in 1889[2]. In 1956 splenogonadal fusion was categorized into continuous and dis continuous types by the Putschar and Manion[3] in a review of 30 cases. In 1990 Carragher[4] published a detailed literature review including 123 cases of spleno gonadal fusion.

2.2 PRESETATION

Splenogonadal fusion is most commonly and incidental discovery during routine groin exploration for an undescended testis or hernia. Of the cases reported cases nearly 17% were diagnosed at autopsy[5]. The most common presentation is that of a testicular swelling[4]. Of 137 cases reviewed by the Karaman and Gonzales, 37% underwent an unnecessary orchidectomy because of suspicion of a primary testicular neoplasm[5]. Of total number of cases only four were reported with a malignant testicular neoplasm. Other presentation include that of an acute painful scrotal pain. Acute torsion of splenic rest mumps, leukemia and mono nucleosis and traumatic rupture of ectopic spleen. Bowel obstruction due to and intraperitoneal cord secondary to continuous splenogonadal fusion was noted by Hines and Eggum[10]. Sripathi reported a case presenting with macro orchidism. Very few cases were suspected or diagnosed preoperatively. One of those was reported by kadlic[12] in 1983. Another 3 cases were diagnosed by 99mTc-Sulpher colloid liver spleen scan, one of them during work up of a patient with an undescended left testicle. And associated limb mal formations[13]. And the 2 during the evaluation of a abdominal mass [14 15]. Patel diagnosed a case by ultrasonography when he followed a tubular process arising from the upper pole of the spleen to the upper pole of undescended testis. The left testis is far more commonly involved than the right side. Only 3 case (2%) had a discontinuous right sided splenogonadal fusion and were all males [17 18].

The condition predominantly in the male patient with male to female ratio of 16:1. Only 9 females were reported in the literature; however inaccessibility to the female gonad by physical examination may distort the true prevalence.

Nearly half of the cases were reported below 10 years of age and others above years of age.

2.3 CLASSIFICATION

Since the report by putschar and manion spleno gonadal fusion has been classified into continuous and discontinuous type. The continuous type involves a direct anatominal connection between the principle spleen and the gonad, by a cord that may be a total splenic, beaded with multiple splenic nodules, or composed of a fibrous tissue. This cord crosses the peritoneal cavity or lies in the left paracolic gutter in vast majority of cases[23]. The discontinuous type lacks a connection between the ectopic and principle spleen. Le Raux and Heddle[24] suggested thata the latter type was simply a rare variant of an accessory spleen and there was no solid evidence that it shared the same etiology nor it is associated with the same congenital anomaly.

2.4 ETIOLOGY

The exact cause of this condition remains unknown. The current acceptable theory is that splenogonadal fusion occurs as a result of events between 5 and 8 weeks of gestation. During this period, several groups of cells derived from the coelomic epithelium and the mesenchyme of the mesogastrium aggregate to form the splenic anlage in the dorsal mesogastrium. This occurs at approximately the same time as the gonadal ridge is formed between the dorsal mesogastrium and the mesonephros on either side. During weeks 6 and 7, rotation of the stomach to the left and growth of the dorsal mesogastrium translocate the spleen to the left side of the abdominal cavity, bringing it into close proximity to the gonadal ridge. Two theories attempt to explain how the spleen fuses with the gonad. The first one is simple adhesion of the left surface of the dorsal mesogastrium and the ventral wall of the developing mesonephric-gonadal structures [3]. This theory is supported by the paragonadal localization of the aberrant splenic tissue and the presence of a splenic capsule in most cases. It does not explain, however, the presence of intraovarian [25] or an aberrant splenic mass beneath the tunica albugenia, nor does it explain right splenogonadal fusion [26]. The second theory postulates mild inflammation between the splenic tissue and the gonad leading to fusion [27]. A teratogenic theory was proposed after reports of one case born to a mother using promethazine R (antiemetic, phenothiazine derivative) during pregnancy [25]

2.5 ASSOCIATED ANOMALIES

In a review of 111 cases, cryptorchidism was the most common anomaly associated with splenogonadal fusion, involving 31% of the cases, of which 59% had bilateral undescended testes [28]. One third of the cases reviewed by Gouw et al [26] were associated with one or more congenital defects, excluding congenital inguinal hernia or cryptorchidism. Continuous splenogonadal fusion is usually associated with severe congenital anomalies and carries a 5-fold higher risk of having associated anomalies in comparison to the discontinuous type. The most common anomaly encountered was peromelia, constituting a subset of patients with relatively higher associated anomalies compared to patients with anomalies other than peromelia. Putschar and Manion [3] proposed that splenogonadal fusion with peromelia is a syndrome different to splenogo nadal fusion without limb defects, with 75% having at least one other anomaly in the former group compared to 15% in the latter [3,25]. Other associated anomalies in decreasing order of frequency are micrognathia, cardiac defects, cleft palate, anal anomalies, craniosynostosis, spina bifida, Mobius syndrome [4], microgastria, thoracopagus [8], and osteogenesis imperfecta [24]. Only 4 cases with the discontinuous type were found to have associated anomalies, one with peromelia and micrognathia and the rest with subclinical ventricular septal defects [8]. In a study performed on 351 embryos, Loomis et al [29] found that the limb buds, the mandible from Meckel's cartilage, and the splenic bud in the dorsal meso gastrium develop at nearly the same time and that any insult during this period may lead to such associated anomalies.

2.6 ASSOCIATED MALIGNANCY

In 1980, Falkowski and Carter [14] first described a 38-year-old male with bilateral impalpable testes, presenting with a left lower abdominal mass which was found to be a left testicular anaplastic seminoma with associated splenogonadal fusion. Since then, 3 more cases were reported describing primary testicular neoplasms with an associated splenogonadal fusion. All of the 4 reported cases were of adults with impalpable undescended testes [10,30,31] or a history of orchiopexy [22]. In all cases,the splenic tissue sent for histopathology was reported as normal splenic tissue. It appears that the coexistence of splenogonadal fusion with a testicular neoplasm is a coincidental finding in this group of cryptorchid patients, who bear a well-documented increased risk for testicular malignancy. Till now, no study has demonstrated a direct relationship between splenogonadal fusion and the development of testicular cancer.

2.7 CONCLUSION

Splenogonadal fusion is a rare congenital anomaly that is seldom diagnosed or suspected preoperatively. Knowledge of this anomaly may spare patients an unnecessary orchiectomy, on the basis of a suspected of a primary neoplasm of the gonad. Preoperative diagnosis was achieved using 99mTc-sulphur colloid liver-spleen scan, which demonstrated this anomaly nicely. In most cases, the splenic tissue may be dissected off the gonadal structures easily, and if there are any doubts concerning the nature of the swelling, an intraoperative frozen section may confirm the diagnosis.

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