

Case Report

Splenogonadal Fusion in a Boy: Case Report and Review of literature

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ABSTRACT

We report an unusual case of splenogonadal fusion in a four-year-old boy with history of bilateral undescended testes. He had presented to us when he was one month old with an empty scrotum. On clinical examination he had bilateral undescended testes. The right testis was palpable in the inguinal region and the left was not palpable. No other anomaly was detected on clinical examination. Right orchidopexy was done at three years of age. Left impalpable testis was subsequently managed by left groin exploration which revealed no testis. Hence exploratory

laparotomy was done. An intrabdominal left testis was found which had two abnormal-looking fleshy masses on its superior pole. Excision biopsy of these nodules was done and first stage orchidopexy was performed as it was difficult to mobilize the testis up to the left scrotum. Biopsy of the nodules revealed splenogonadal fusion; hence we were justified in saving the testis. Second-stage orchidopexy was done after three months to fix the testis in the left scrotum.

KEY WORDS: impalpable, intraabdominal, splenogonadal fusion, testis

INTRODUCTION

Splenogonadal fusion is a rare congenital anomaly, which is defined as an abnormal connection between the spleen and gonad or derivatives of mesonephros. It was first described by Boestrom in 1883 and later reported in detail by Pommer in 1989^[1].

It occurs most commonly in the left gonad in males. To date, approximately 150 cases have been reported in the literature. Preoperative diagnosis is possible only in a few cases and unnecessary orchidectomy is often performed because of suspicion of neoplasm^[2]. Cryptorchidism has been associated with splenogonadal fusion^[3]. We report a case of a child with an impalpable left testis with splenogonadal fusion and review the relevant literature.

CASE REPORT

A one-month-old boy was referred to us because of an empty scrotum. On clinical examination he had bilateral undescended testes with right palpable and left impalpable testis. He had a normal phallus and no other anomaly was noticed on clinical examination. An ultrasound examination showed normal-sized right testis in the inguinal canal but

the left testis was not found in the groin or abdomen. He was advised to undergo right orchidopexy first at one year of age but he did not come back to us till he was three and half years old. Right orchidopexy was planned first and normal-sized right testis was fixed in the scrotum. Three months later, left groin exploration by transverse crease incision was done. The left testis or cord structures were not present in the inguinal canal; hence incision was extended laterally and the abdominal cavity was entered after opening the peritoneum. The left testis was present in the peritoneal cavity deeper to the internal inguinal ring. It looked abnormal because of two fleshy masses on its superior pole. The first nodule was discrete and 1x1 cm size, attached to the cord in close proximity with the superior testicular pole. The second nodule diffusely merged with the superior pole of the testis near the head of epididymis. A suspicion of malignancy was considered intraoperatively on gross examination but it was decided to wait for the biopsy report to avoid unnecessary orchidectomy. The first nodule was excised without damage to any cord structure. Longitudinal excisional biopsy of the second nodule was done so as to include adjacent normal-looking testicular tissue. The testis was mobilized on its cord

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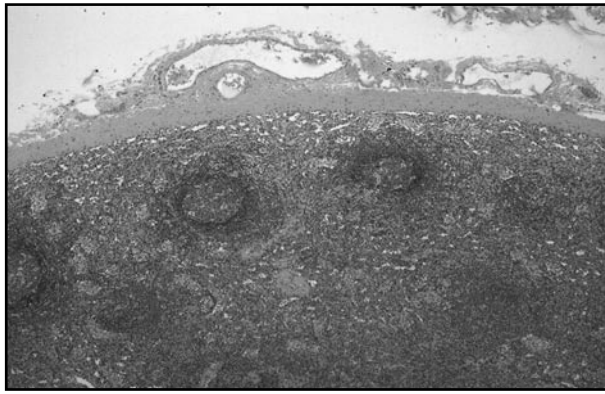


Fig. 1: Nodule on the upper pole of the testis comprising of ectopic spleen. (Hematoxylin and Eosin stain, 4 X)

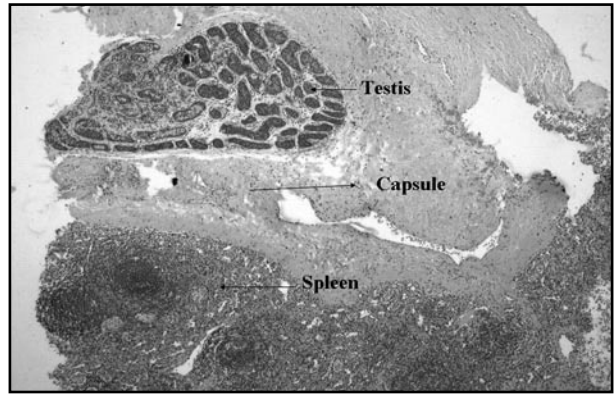


Fig. 2: Upper pole of left testis showing splenogonadal fusion. (Hematoxylin and Eosin stain, 4 X)

but it was not possible to bring it into the scrotum. Hence staged orchidopexy was planned and the left testis was fixed to the pubic tubercle. Histopathology report of the nodules came as splenogonadal fusion. The first nodule showed only ectopic splenic tissue (Fig. 1). The second nodule biopsy showed splenic and testicular tissues merging with each other by their capsules; hence a diagnosis of splenogonadal fusion was made (Fig. 2). Orthotopic spleen was normal and it was shown on subsequent ultrasound. Second stage orchidopexy was done three months later. The testis could be brought to the left scrotum this time without any tension. Both testes were well felt in the scrotum, on follow up visits.

DISCUSSION

Splenogonadal fusion is a rare entity. Several clinical reviews of this disorder have been published in the literature. It has been traditionally classified into continuous and discontinuous types^[4]. In the continuous type there is a direct connection between the principal spleen and gonad; the discontinuous type lacks any anatomical connection between ectopic spleen and native spleen. Both types occur with relatively equal frequency^[1]. Cryptorchidism is the most commonly associated anomaly involving 31% cases of splenogonadal fusion out of which 59% had bilateral undescended testes^[3]. The continuous type may be associated with other congenital anomalies like limb malformations, micrognathia, cardiac defects, cleft palate, anal anomaly, craniosynostosis and spina bifida. This reflects an embryological insult occurring between the 5th to 8th weeks of gestation^[5]. Although the exact etiology of splenogonadal fusion remains unknown, the most widely accepted etiology relates to events between the 5th and 8th weeks of gestation. Between five and six weeks of gestation splenic anlage forms in the dorsal mesogastrium. At the same time the gonadal ridge is formed between the dorsal mesogastrium and mesonephros on either side. As the stomach rotates to the left, splenic anlage also rotates and

lies in close proximity with the left gonadal ridge. Simple adhesion^[6] or mild inflammation^[7] between splenic tissue and gonad at this stage explains left presentation of splenogonadal fusion. The fact that in most cases the splenic capsule is intact supports the theory of simple fusion. It does not explain rare cases of right sided splenogonadal fusion^[6], intraovarian^[8] or intratesticular splenic tissue^[2].

Splenogonadal fusion occurs predominantly in males with a male- to- female ratio of 16:1. Out of 150 cases reported till now there were only 9 females; however, incidence in females could be underestimated because of inaccessibility of the female gonad to physical examination^[9]. This disorder can present at any age from birth to 80 years although the majority (82%) are below 30 years. Nearly half present below 10 years of age^[1,10]. Most commonly, this entity is an incidental discovery during routine groin exploration for an undescended testis or hernia^[10]. Approximately 17 % of reported cases were diagnosed at autopsy^[1]. Testicular swelling is the commonest presenting symptom. Another presentation is acute scrotal pain due to involvement of the ectopic spleen in various conditions like malaria^[11], torsion of splenic rest^[10], mumps, leukemia, mononucleosis^[12], traumatic rupture^[13] and bowel obstruction due to intraperitoneal cord of continuous splenogonadal fusion^[14]. Very rarely this condition is suspected or diagnosed preoperatively. Few imaging reports of this entity have appeared in the literature. Scintigraphy using agents labeled with technetium-99m has confirmed presence of splenic tissue in some reports^[15] and more recently radiologists have diagnosed this condition by ultrasound^[16, 17].

While no treatment is necessary when diagnosis is confirmed and no symptoms are present, in 37% of reported cases orchidectomy was done unnecessarily^[10]. There can be coincidental occurrence of testicular neoplasm in cases of splenogonadal fusion but till now no study has demonstrated a direct relationship between

splenogonadal fusion and development of testicular cancer^[18]. If surgical intervention is performed, orchidectomy is generally not indicated; splenic tissue usually can be dissected off the gonadal structures and the testis can be saved^[1, 10, 12, 13, 18].

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