An Unusual Content in a Congenital Hernia - Complete Spleno –gonadal Fusion Band

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Abstract Six years old boy underwent elective inguinal exploration for left congenital hernia. Per- operatively, an elongated, purplish-red, fleshy band of tissue was found inside the sac, adherent to the upper pole of testis. Biopsy was taken and the wound closed. An MRI done after 4 weeks proved the origin of the band from spleen. Laparotomy and excision of the band was done. The histo-pathology of the specimen was reported as normal splenic tissue. The above features are consistent with a diagnosis of spleno - gonadal fusion (SGF).

Keywords Spleno-gonadal fusion · MRI for spleno-gonadal fusion · Splenic tissue in the testis · Splenic anomalies

Case Report

A 6 years old boy attended our OPD with complaints of a left inguino – scrotal swelling. The swelling was reducible and was present since 1 1/2 years of age. On initial examination the sac was felt along with the cord structures in the root of the

scrotum. Both testes were descended and the external genitalia was normal. He was diagnosed to have left congenital hernia and underwent elective inguinal exploration. The sac was identified, separated from cord structures and opened. An elongated, purplish-red, fleshy band of tissue was found inside the sac, adherent to the upper pole of testis (Fig. 1). Further exploration through the internal ring revealed this tissue was found extending towards the left upper quadrant of the abdomen, trans-peritoneally. Some large vessels were seen on the surface of the band. A provisional diagnosis of spleno gonadal fusion was made. However, as the relative contribution of the vessels from the band to the vascularity of the testis could not be determined and a pre-operative consent for orchiectomy was not available, it was decided to take a biopsy from this tissue. A wedge biopsy was taken and the wound was closed. The histo-pathology of the biopsy specimen was reported as normal splenic tissue.

An MRI scan was done (4 weeks after the initial surgery) to delineate the soft tissue & vascular anatomy. A long cord-like extension of splenic tissue was seen from the inferior pole of the spleen to the level of the left inguinal canal (Fig. 2). It was located just deep to the parietal wall and was anterior to the descending colon and the left ilio-psoas. The signal intensity of this cord of tissue was seen paralleling the spleen in all sequences. A thin vascular channel could also be traced from the splenic hilum into the cord of tissue, further confirming its origin from the spleen. The left testis was small in size and was closely apposed to the inferior end of this cord of splenic tissue at the level of the inguinal canal. The above features were consistent with a diagnosis of complete spleno-gonadal fusion (SGF). A dopplerultrasonogram was also done which showed the presence of good, independent vascular supply to the testis via the vessels in the cord structures.

A mini -laparotomy and left inguinal exploration was carried out. The band was found arising from the inferior

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Fig. 1 Band of tissue adherent to the upper pole of left testis

pole of the spleen, traversing anterior to the left colon and entering the left inguinal region (Fig. 3). The lower end was adherent to the upper pole of testis. However the tunica albuginea was found intact. A vascular clamp was applied across the band and the vascularity of the testis was ascertained. The band was surrounded by loose areolar tissue and could be easily excised in toto and the ends were over-sewn. The completely excised band measured 18 cm in length and was 21 mm thick (Fig. 4). The histopathology was consistent with normal splenic tissue. Patient made an uneventful recovery. A final diagnosis of complete spleno-gonadal fusion (SGF) was made.

Fig. 2 MR imaging study showing the cord like extension from the spleen to the left inguinal canal

Fig. 3 The band arising from the interior pole of the spleen at laparotomy

Discussion

Spleno-gonadal fusion is a rare congenital malformation in which the spleen is abnormally connected to the gonads or rarely to the mesonephric structures like vas or epididymis [1]. More than 150 cases have been described in world literature so far. Putschar&Manion classified spleno-gonadal fusion into two types, viz. continuous and discontinuous [2]. In the continuous variant, the spleen is connected to the gonads by a fibrous cord, a splenic cord or a beaded cord of mixed fibrous and splenic tissue. In the discontinuous type, the ectopic tissue is attached to the gonads, but no connection with the orthotropic spleen is identified [2]. The two forms appear to occur with relatively equal frequency. About one fifth of the continuous type cases are associated with other major congenital abnormalities, such as limb defects (ectromelia, peromelia),

Fig. 4 The completely excised band

micrognathia, cardiac defects, etc. [3]. The male: female ratio is reported to be about 15:1 [3].

It is believed that the adherence of the splenic primodium to structures derived from the mesonephric ridge occurs between fifth and eighth week of gestation (before the gonadal descent and at the time when the splenic analage lies in close proximity to the left gonad during rotation of the dorsal mesogastrium [4]. The descent of the testis seems to draw out the developing spleen into a long band in some cases or to detach a portion of the splenic primodium and carry it down with the descending testis in others [4].

SGF typically presents as left sided scrotal swelling, left inguinal hernia or cryptorchidism [3]. Other presentations include painful scrotal swelling secondary to mumps and viral infections or bowel obstruction caused by the cordlike structure seen in the continuous form [5]. The diagnosis is usually made on the operating table during surgery for hernia, undescended testis or testicular mass. Though a sonogram would be in order for the workup of a testicular mass, a definite diagnosis cannot be made solely on the basis of sonographic findings [6]. Some authors have resorted to using ⁹⁹Tc – Sulfur colloid imaging to identify ectopic splenic tissue [7].

An MRI scan is valuable as it gives the necessary soft tissue and vascular delineation. The MR appearance of spleno-gonadal fusion has not been described in literature so far. The characteristic signals of the fusion band paralleling the signal of the spleen helps in identifying the tissue. The availability of respiration-triggered sequences, aid in exquisite anatomic delineation, which is not possible with other imaging modalities.

In the discontinuous form, surgical exploration is often necessary to rule out a malignancy in a suspected testicular mass. An intra-operative frozen section can aid in the diagnosis [8]. If normal splenic tissue deposits are reported, they can be left alone. Symptomatic masses of splenic tissue can usually be dissected away from the tunica albuginea. Orchiectomy should be avoided for these benign lesions. Unfortunately, 37% of the reported cases are associated with orchiectomy [6].

Available literature on the treatment complete symptomatic SGF is limited; many of these lesions are detected incidentally during laparotomy for other indications or at autopsy [3]. Colonic obstruction secondary to extrinsic compression by the fleshy band has been reported. It may also serve as an axis for small bowel volvulus and knotting, etc. These bands are symptomatic in patients with ITP and hemolytic anaemias. They can be excised with minimal morbidity to the patients. Laparoscopic excision of the band is also a viable option.

Conclusion

SGF is a rare congenital anomaly usually presenting as an inguinal hernia or testicular mass. The diagnosis is usually made on the operating table. A prior knowledge about this condition avoids unnecessary orchiectomy.

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