Splenogonadal fusion: A very rare congenital anomaly in the differential diagnosis of a testicular mass

Splenogonadal füzyon: Testis kitlesi ayırıcı tanısında nadir bir konjenital anomali

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ABSTRACT

Splenogonadal fusion is a rare congenital anomaly characterized by congenital fusion between the spleen and testicular tissue. In the literature, there are approximately 175 reported cases, and most of them are associated with cryptorchidism. In this article, we report an unusual case of splenogonadal fusion that was pre-diagnosed as a solid testicular mass in a patient who underwent orchiectomy.

Key words: Splenogonadal fusion; testis; testicular cancer.

ÖZET

Splenogonadal füzyon dalak ve testis dokularının konjenital füzyonu ile karakterize çok nadir bir anomalidir. Literatürde bildirilmiş yaklaşık 175 vaka bulunmaktadır. Vakaların çoğunda inmemiş testis öyküsü bulunmaktadır. Bu çalışmada, testiste solid kitle tespit edilen ve orşiyektomi sonrası patolojisi spleneogonadal füzyon olarak rapor edilen 20 yaşında bir olguyu bildirmekteyiz.

Anahtar kelimeler: Splenogonadal füzyon; testis; testis kanseri.

Introduction

Splenogonadal fusion is a rare congenital anomaly characterized by fusion of the spleen and a gonad. In the literature, there are approximately 175 reported cases, and most of them are associated with cryptorchidism.[1] Splenogonadal fusion is classified either as continuous, in which there is a direct anatomical connection between the spleen and the gonad, or discontinuous, in which there is no direct connection between these two organs.[2] Testicular solid masses in young adults are presumed to be malignant, and surgical treatment is typically warranted. In this article, we report an unusual case of splenogonadal fusion that was pre-diagnosed as a solid testicular mass.

Case presentation

A 20-year-old male in the military service was admitted to the urology clinic with left scrotal pain and a palpable mass. He claimed that the pain started after a vigorous physical exercise session, however, the palpable mass had been present since childhood. The patient did not report any trauma, and had no history of cryptorchidism.

Physical examination revealed no signs of inflammation in the scrotum. The mass was located inside the scrotum just beneath the epididymis, and it was slightly painful on palpation. The border between the solid mass and the epididymis was vague.

Scrotal color Doppler ultrasound revealed that the 3 cm hypoechoic, homogenous mass was located intratesticularly just under the epididymis. Marked hypervascularity, and increased blood flow were noted within the mass. No sign of inflammation was reported. Serum alpha-fetoprotein, beta HCG and lactate dehydrogenase levels were normal. Chest roentgenograms and other laboratory findings were within normal range.

According to the clinical signs, the mass was suspected to be a malignant or even benign testicular tumor. The patient was informed about the clinical picture in full detail, and informed consent form was signed by both the

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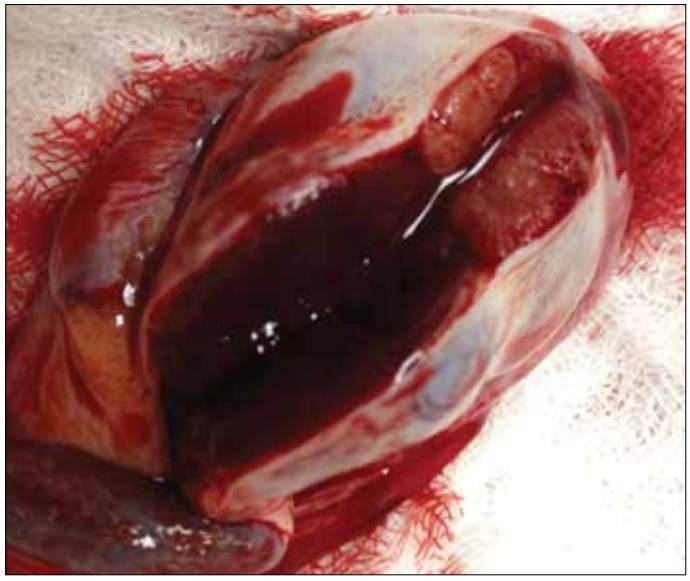

Figure 1. Gross appearance of the specimen. The lesion was dark red in color and resembled liver tissue

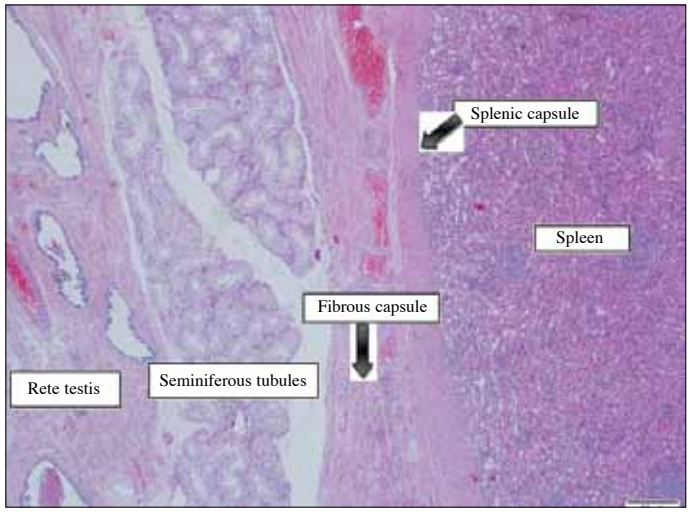

Figure 2. Microscopic examination: The splenic tissue was separated from the seminiferous tubules by a splenic capsule and vascular fibrous connective tissue

patient and a next of kin. Left inguinal orchiectomy was performed under regional anesthesia. As the surgical material was removed, immediate transaction of the testis was performed on the bench. The tumor was confined within the tunica vaginalis. The gross appearance of the specimen was very different from a typical testicular mass. It was dark red in color and resembled liver tissue (Figure 1). Macroscopic pathological examination demonstrated that the tumor was 3.6x1.6 cm in diameter, dark brown in color, solid and located in the upper pole. Microscopic examination of the specimen revealed neither a benign nor malignant tumor, and the lesion was reported as a splenogonadal fusion. The splenic tissue was separated from the seminiferous tubules by a splenic capsule and vascular fibrous connective tissue (Figure 2).

Discussion

Splenogonadal fusion is a rare congenital anomaly. It was first described by Bostroem in 1883,[3] and approximately 175 cases have been reported in the literature. The age of presentation is less than 10 years in half of the reported cases, and 82% of the cases occur in patients younger than 30 years.[4]

Splenogonadal fusion is postulated to result from the development of an abnormal attachment between the gonad and spleen during approximately the 5th and 8th weeks of embryonic life, when the organs are in close proximity to each other. When gonadal descent begins, the attached splenic tissue follows the gonadal path.[5] Splenogonadal fusion can either be continuous, in which the direct anatomical connection between the spleen and the gonad persists, or discontinuous, in which there is no direct connection between the two organs, and the splenic tissue is solitarily attached to the gonad.[2] In one third of cases, continuous splenogonadal fusion is associated with congenital anomalies such as cleft palate, cardiac defects, micrognathia, spina bifida, thoracopagus, osteogenesis imperfecta and extremity deformities.[2,4,6]

Splenogonadal fusion is typically diagnosed incidentally during surgery for inguinal hernia and/or cryptorchidism, which are the two most frequently associated anomalies. Cortes et al.[7] reported a 31% cryptorchidism rate in their review of 111 splenogonadal fusion cases. It is almost impossible to establish the diagnosis of splenogonadal fusion clinically. Moreover, in certain cases, including the one presented here, the clinical picture closely resembles a primary testicular neoplasm, and the patient may undergo an unnecessary orchiectomy.[8] However, in patients with a nonpalpable testis, radiologic evidence of a close relationship between the missing gonad and the spleen may arouse the suspicion of the presence of this anomaly.[3]

The unique features of the case presented here can be stated as follows. The patient did not present with cryptorchidism or hernia, which are commonly associated with splenogonadal fusion. As previously stated, splenogonadal fusion is typically discovered incidentally during surgery for cryptorchidism or hernia. Typically, the diagnosis is pathologic, and patients are rarely symptomatic. However, in our case, the patient was symptomatic. He had a palpable mass and suffered from pain in that testicle, particularly after exercise, which is uncommon in splenogonadal fusion.

Although the presentation of this case possesses the key diagnostic elements of testicular cancer (a palpable intrascrotal hypoechoic solid mass with marked vascularization), there are issues that render the diagnosis suspicious, including the pain

felt by the patient, particularly after heavy exercise, the presence of the mass dating back to childhood and negative tumor markers.

Also notable is the role of organ-preserving surgery in testicular neoplasms. The current literature indicates that testis sparing surgery can be considered for the treatment of small (less than 2 cm) suspicious nonpalpable lesions detected by ultrasonography as well as bilateral tumors and tumors in a solitary testis. The case presented here was apparently not a candidate for testis-sparing surgery, and the patient therefore underwent radical orchiectomy.

Splenogonadal fusion is a rare congenital anomaly that is frequently associated with cryptorchidism and hernia. Continuous splenogonadal fusion can be suspected, and properly diagnosed in young patients with cryptorchidism and associated anomalies. However, discontinuous splenogonadal fusion may be misdiagnosed as a malignant testicular mass with resultant unnecessary orchiectomy.

Informed Consent: Written informed consent was obtained from patients who participated in this case.

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