Abstract

Introduction: The presence of more than 2 testicles is called polyorchidism. It is a rare anomaly. Less than 100 cases have been reported in the literature. Clinical Picture: A 51-year-old man complained of a painless right groin lump for a year. Physical examination revealed a discrete ovoid non-tender, firm but mobile lump. No cough impulse was demonstrated. Sonography demonstrated a soft tissue structure within the tunica vaginalis inferior to the deep inguinal ring. It demonstrated similar echotexture and vascular flow as the normal testis, but smaller in size. The findings were compatible with a diagnosis of polyorchidism. Treatment: The patient was not keen for surgery and was followed up conservatively. Conclusion: Differential diagnosis of a solid extratesticular mass should include polyorchidism.

Key words: Testis, Ultrasonography, Urogenital abnormalities

Introduction

The presence of more than 2 testicles is called polyorchidism. It is a rare anomaly. Less than 100 cases have been reported in the literature. Triorchidism is the most common presentation.1 The left side is predominantly affected. Approximately 50% of cases are seen between 15 and 25 years of age.2 About 75% of supernumerary testes are intrascrotal, with another 20% located in the inguinal canal and 5% in the retroperitoneal space.3 A prevalence of left-sided lesions (left-to-right ratio of approximately 3:1) has been observed.4 The majority of the patients are asymptomatic and present with painless groin or testicular masses. We describe a case of triorchidism in a 51-year-old man. To our knowledge, this is the first reported case in Singapore.

Case Report

A 51-year-old man complained of a right groin lump for a year. It was painless and stable in size. The patient was otherwise well, without a history of acute trauma, viral disease or infection. Physical examination revealed a discrete ovoid non-tender but mobile lump. No cough impulse was demonstrated. Alpha-fetoprotein (α-FP) and prostate-specific antigen (PSA) levels were normal, measuring 4.16 ug/L (1.00 to 10.00 ug/L) and 0.35 ug/L (0.00 to 4.00 ug/L), respectively. Sonography revealed a soft tissue structure with similar echotexture and vascular flow as the normal testis within the tunica vaginalis inferior to the deep inguinal ring (Fig. 1). The soft tissue structure measured approximately 2.8 x 1.3 x 2.5 cm, and the normal right and left testis measured 3.9 x 1.9 x 2.1 cm and 3.8 x 1.9 x 2.3 cm, respectively. There was no focal abnormal echogeneity that was suspicious of malignancy. Both the left and right testicles were unremarkable (Fig. 2). The findings were compatible with a diagnosis of supernumerary testis. A moderate degree of varicocele was also noted. The patient was not keen for surgery and was followed up conservatively. The lesion has remained stable 1 year after the initial clinical consult.

Discussion

The aetiology of polyorchidism is thought to be due to accidental longitudinal or transverse division of the genital ridge, with or without the mesonephros, before the 8th week of gestation, either through local accident or development of peritoneal bands.2 Depending on the segmentation plane and site, supernumerary testes may develop with a common or single epididymis and vas deferens. In most cases, the
epididymis and vas deferens are shared or missing. Mastroeni et al proposed a functional classification based on embryogenic development. In type I, the supernumerary testis lacks an epididymis and vas deferens. The split-off part of the primordial gonad does not communicate with the mesonephric tubules from which the epididymis develops. In type II, the supernumerary testis is linked to the regular testis by a common epididymis and shares a common vas deferens with it. The division of the genital ridge occurs in the region where the primordial gonads are attached to the mesonephric ducts, although the latter are not divided (incomplete division). In type III, the supernumerary testis has its own epididymis, but shares the vas deferens with the regular testis. This variant results from a complete transverse division of the genital ridge. Chromosomal abnormalities, including chromosome 21 long-arm deletion, have also been implicated in some cases.

In the evaluation of a scrotal mass, it is useful to distinguish the lesion into intratesticular or extratesticular and cystic or solid mass. Cystic extratesticular masses, such as hydroceles, epididymal cysts and varicoceles, are benign. Extratesticular solid masses are also mostly benign, with the prevalence of malignancy being approximately 3%. The most common extratesticular neoplasms are lipomas and adenomatoid tumours. They commonly arise from the spermatic cord and epididymis, respectively. Paratesticular masses comprise a group of lesions that are extratesticular, but are not easily categorised as originating from one of the other paratesticular tissues, namely, the epididymis, spermatic cord and fascia. These lesions include hernias, scrotal calculi, fibrous pseudotumours and polorchidism. They often appear as a mass within the tunica vaginalis.

On sonography, supernumerary testes demonstrate similar echo texture and vascular flow pattern as normal testes. Normalcy of testicular architecture and location are tantamount to a diagnosis of a normal variant. Magnetic resonance (MR) imaging can be helpful in clarifying the diagnosis if sonographic findings are inconclusive. The supernumerary testes have the same MR imaging characteristics as normal testes. They have intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images, with each testis having a tunica albuginea of low signal intensity.

Polyorchidism is associated with hydrocele, varicocele, indirect hernia (20%) and torsion (7%). Polyorchidism has also been found in patients with coexisting cryptorchidism (15%), multiple malformation syndromes (7%) and chromosomal anomalies (3%). Infertility (20%) is also a common finding. Thirty-seven per cent of the
testes lack spermatogenesis, tubular atrophy or Sertoli cell pattern (without Leydig cells). \(^4\)

Malignancy or malignant degeneration (seminoma, teratoma and choriocarcinoma) appears to occur in approximately 4% to 7% of cases. \(^7\) However, the true malignant potential of supernumerary testes is difficult to establish due to the rarity of the condition. It is also complicated by its coexistence with congenital risk factors, such as cryptorchidism. Spranger et al \(^2\) argued that testicular malignancies have a preponderance for the right side, unlike the left-sided predominance of polyorchidism. The explanation for this discrepancy is not evident. In any case, most of these testes are histologically normal and they argued that it is reasonable to follow these cases clinically.

Treatment option may either be watchful waiting or surgery. Surgical exploration is performed to ensure normalcy and to exclude tumour when clinically suspicious. In the absence of any concomitant disorder and if testicular tumour can be ruled out by sonography or MR imaging, surgical exploration with biopsy is unnecessary.

Conclusion

The diagnosis of polyorchidism is usually incidental. It is a diagnosis of exclusion requiring a high index of suspicion. The differential diagnosis of a solid extratesticular mass should include polyorchidism. Our case highlights the importance of a high index of suspicion amongst clinicians to the possibility of polyorchidism in the clinical work up of a solid extratesticular mass. Our patient presented with paratesticular lump of similar echo-texture and configuration to a testis on ultrasonography with no suspicious focal lesion. When clinical finding and imaging detected no focus suggestive of malignancy, the patient can be followed up conservatively. Our patient was contented with watchful waiting and has been stable 1 year post initial clinical consult.

REFERENCES