Triorchidism - A Case Report of a Rare Congenital Anomaly

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INTRODUCTION

Polyorchidism is a rare congenital anomaly usually found on the left side and frequently associated with inguinal hernia, testicular torsion, and cryptorchidism. An increased risk of testicular malignancy has been reported in cases of Polyorchidism.[1] It may be detected as a scrotal mass in children or may go undetected till adult life and found incidentally. Different classifications of supernumerary testis exist depending on the location of the extra testis and its relationship to the native testis and the vas deferens. The most commonly occurring presentation is one of an extra testis or triorchidism on the left side. We report here a case of Triorchidism in a young male patient who was clinically suspected to have an epididymal cyst.

Polyorchidism, or supernumerary testicle is the presence of more than two testes, most frequently presenting as triorchidism[1] It is an extremely rare congenital anomaly of the urogenital system and there are only about a hundred documented cases in literature. We describe here a rare case of Triorchidism in a 21-year-old male patient who presented with long standing left sided painless scrotal swelling, clinically suspected to be an epididymal cyst and evaluated using high resolution Ultrasonography including Colour Doppler and Magnetic Resonance Imaging (MRI). Ultrasound demonstrated the supernumerary testis to be located posterosuperior to the native left testis with each having their own epididymal and vas deferens connection. MRI confirmed the presence of supernumerary testicle with an abnormal signal intensity on T2W images appearing homogenously hypointense in contrast to the hyperintense signal on T2W images of the two normal testicles leading to the suspicion of the occurrence of seminomatous transformation of the testicle. The finding of necrotic enlarged iliac, inguinal, pre and para aortic lymph nodes both on ultrasonographic and MRI evaluation further supported the suspicion of a seminomatous transformation.

PRESENTATION OF CASE

A 21-year-old male patient presented to the Surgical Out Patient Department (OPD) with complaints of long standing left sided painless scrotal swelling. He had no history of trauma, fever or local tenderness at the site. Physical examination revealed an ovoid, non-tender, mobile lump at the upper pole of left testicle. He was clinically suspected to have an epididymal cyst and was referred to the Radiology department for confirmation by ultrasonography. Grey scale and colour doppler ultrasonography of the scrotum demonstrated evidence of three testis, one in the right hemi scrotum and two in the left hemi scrotum. Both the testicles in the left hemi scrotum namely the supernumerary and the native testicle had their own epididymis and vas deferens. The supernumerary testicle was located posterosuperior to the native testis within the left hemi scrotum and showed diffuse homogeneous decreased echotexture (hypoechoic) in comparison to the native left testis on grey scale ultrasound examination while colour doppler examination revealed a significant diffuse increase in vascularity. The other two testicles were normal both in echotexture and vascularity.

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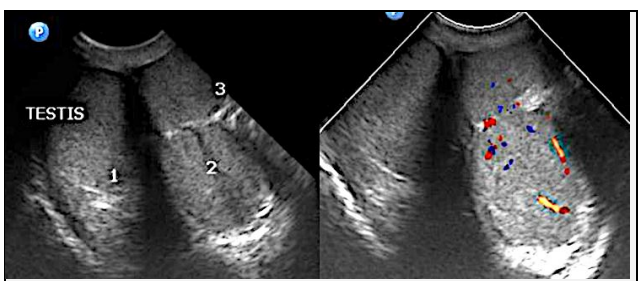
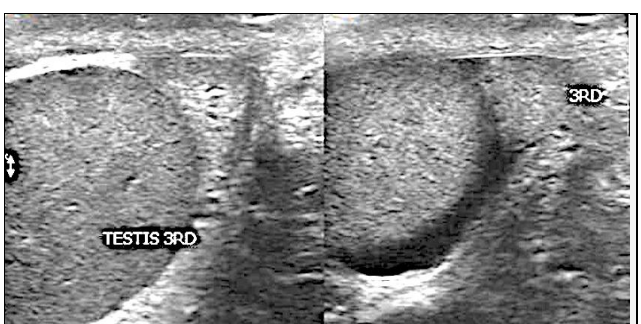
Ultrasound findings of triorchidism was confirmed using MRI (1.5 tesla). MRI of the scrotal region revealed two testis one in either scrotal sac showing normal signal intensity (hypointense on T1W images and hyperintense on T2W images). The supernumerary third testis in the left scrotal sac showed homogeneous hypointensity on T2W images with normal signal intensity of the epididymis. Abdominal screening using ultrasonography and MRI revealed multiple enlarged iliac, inguinal, pre and para aortic lymph nodes. On ultrasonographic examination these lymph nodes revealed complete loss of morphology and shape and appeared hypoechoic with loss of the normal echogenic fatty hilum suggestive of necrosis within the lymph nodes. These nodes revealed decreased intensity on MRI. The patient was advised further evaluation with histopathological examination in order to rule out seminomatous transformation of the supernumerary testis. However, patient refused admission and so was lost for further follow up.

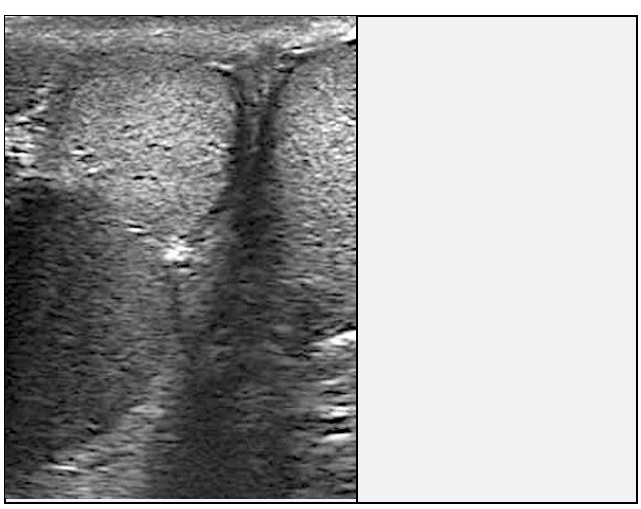
Ultrasound with Doppler

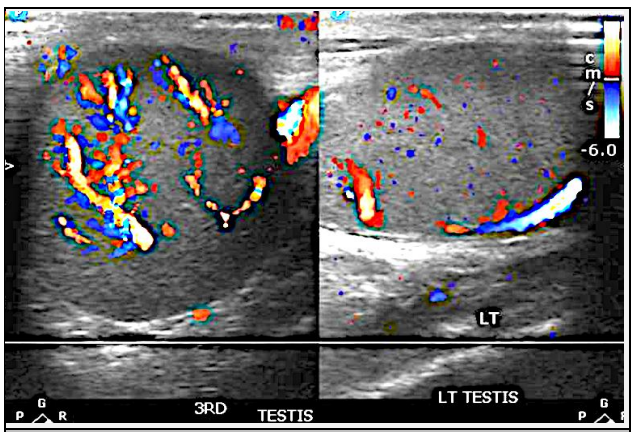


Sonography Demonstrated Evidence of Three Testes.

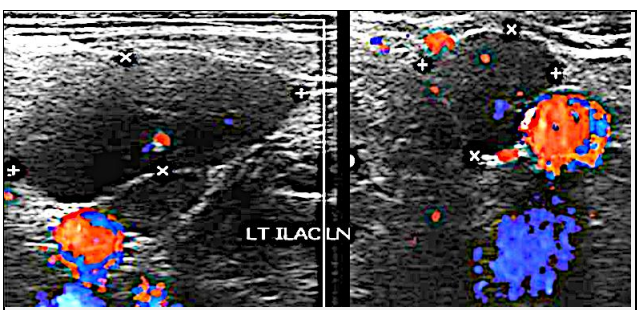


Third Testis Showed Diffuse Homogenous Decreased Echotexture (Hypoechoic) as Compared to Other Two Normal Testes

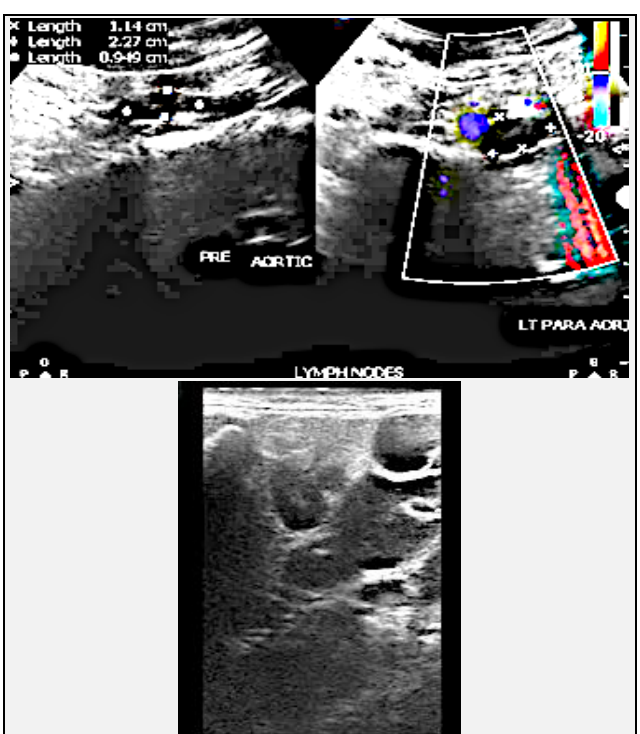




Third Testis Showed Diffuse Homogenous Decreased Echotexture (Hypoechoic) and Colour Doppler Study Showed Diffuse Increase in Vascularity in The Third Testis



USG Revealed Multiple Enlarged Iliac and Inguinal Lymph Nodes. On Ultrasound, These Lymph Nodes Exhibited Complete Loss of Morphology, Shape and Hypoechoic with Loss of Hilar Echotexture Suggestive of Necrotic Lymph Nodes



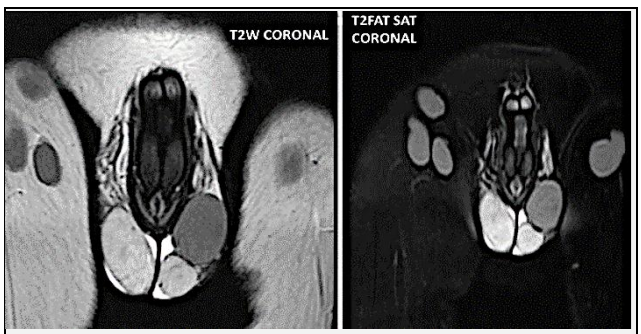
Screening of the Abdomen on USG Revealed Multiple Enlarged Pre and Para Aortic Lymph Nodes with Loss of Fat Hilum Suggestive of Necrotic Lymph Nodes

Sonography demonstrated evidence of three testis with three epididymis. Third testis showed diffuse homogenous decreased echotexture (hypoechoic) as compared to other two

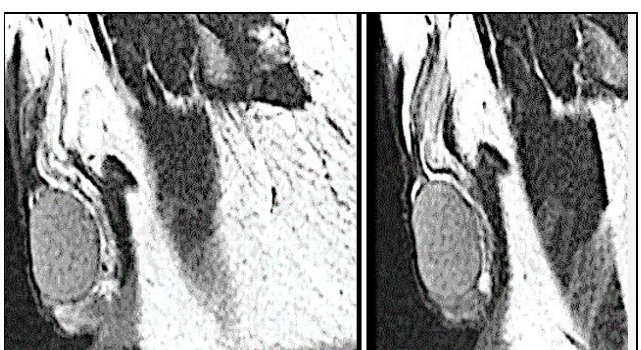
normal testis. Colour doppler study showed diffuse increase in vascularity in the third testis.

Screening of the abdomen on USG revealed multiple enlarged iliac, inguinal, pre para aortic lymph nodes. On ultrasound these lymph nodes exhibited complete loss of morphology, shape and hypoechoic with loss of hilar echotexture suggestive of necrotic lymph nodes.

MRI Images

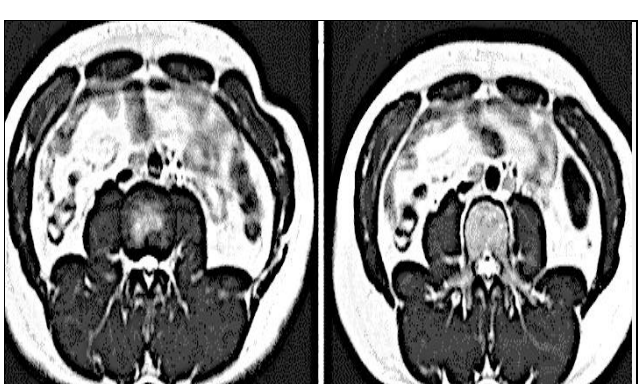


MRI of the Scrotal Region Revealed Two Testes Which Were Showing Normal Signal Intensity [hypointense on T1W Images and hyperintense on T2W Images]. Third Testis Showed Homogenous Hypointensity on T2W Images with Normal Epididymis

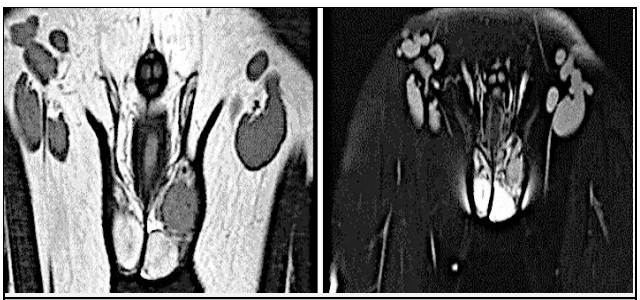


MRI of the Scrotal Region Shows Third Testis to Be Having Homogenous Hypointensity with Normal Epididymis

MRI of the scrotal region revealed two testis which were showing normal signal intensity [hypointense on T1W images and hyperintense on T2W images]. Third testis showed homogenous hypointensity on T2W images with normal epididymis. Screening of abdomen showed multiple enlarged inguinal, iliac, pre and para-aortic and aortocaval lymph nodes.



Screening of Abdomen Showed Multiple Enlarged Para-Aortic and Aortocaval Lymph Nodes



MRI of the Scrotal Region Shows Third Testis to Be Having Homogenous Hypointensity and Multiple Enlarged Inguinal and Iliac Lymph Nodes

DISCUSSION

Polyorchidism is an extremely rare congenital anomaly of the urogenital system and there are only about a hundred documented cases in literature.[2] The exact aetiology of Polyorchidism is unclear. The primordial testis develops from the medial aspect of the primitive genital ridge at about 6 weeks of embryological life in a normal embryo while the epididymis and vas deferens develop from the Wolffian duct at about 8 weeks of embryological life. Several theories regarding the mechanism of occurrence of Polyorchidism have been postulated. Duplication or longitudinal vs transverse division of the urogenital ridge possibly due to development of peritoneal bands, anomalous appropriation of cells, development of peritoneal folding and incomplete degeneration of a portion of the mesonephros are all theories proposed to explain the development of Polyorchidism. The most accepted theory out of these explains the cause to be anomalous division of the embryonal genital ridge either with or without the mesonephros before the 8th week of embryonic life caused by a local accident or by development of peritoneal bands. The supernumerary testis's communication with the epididymis and vas deferens may or may not be retained depending on the plane of segmentation and the level of division. Polyorchidism has been classified by Leung based on embryological development.[3,4]

Type I: Supernumerary testis lacks an epididymis and vas deferens as a small part of the genital ridge having no contact with the mesonephric duct is separated by the division.

Type II: Supernumerary testis has its own epididymis as the genital ridge division occurs at the site of attachment of the primordial gonads to the metanephric ducts.

Type III: Supernumerary testis has its own epididymis and shares the vas deferens with the native testis which is due to incomplete longitudinal division of the genital ridge and the proximal portion of the mesonephric duct or complete transverse division of the genital ridge and the mesonephros.

Type IV: Complete duplication of the testis, epididymis and vas which is due to complete longitudinal division of the genital ridge and mesonephros.

Another classification was proposed by Singer et al, based on the anatomical and functional potential of the supernumerary testis.[3,4]

Type 1: Supernumerary testis communicates with the draining vas deferens and epididymis and hence has reproductive potential. (corresponding to Leung type II, III. IV).

Type 2: Supernumerary testis lacks attachment to a vas and an epididymis and hence has no reproductive potential. (corresponding to Leung type I).

Both the types are further subdivided into two depending on their location either within or outside the scrotal sac.

In most cases patients are asymptomatic and diagnosis is incidental. In a few patients the presenting symptom is a painless mass in the groin or scrotum. In others the primary disorder might be cryptorchidism, inguinal hernia, maldescended testis, infertility, torsion, hydrocele, varicocele, epididymitis or malignancy.

The supernumerary testis is commonly located within the scrotum either superior or inferior to the native testis. Malignant transformation within the supernumerary testis is known to occur, the most commonly reported neoplasms being embryonal carcinoma, germ cell tumour and seminoma.[4] Ultrasonography is diagnostic in most cases while confirmation of the diagnosis is made by MRI. The supernumerary testis on ultrasound examination appears as a scrotal mass located superior or inferior to the native testis and has an echotexture identical to that of the ipsilateral native testis. Flow characteristics on Colour Doppler sonography are also similar in both testicles.

MRI confirms ultrasound findings of the supernumerary testis which appears as a round or oval structure having the signal characteristics of testicular tissue showing intermediate signal intensity on T1 weighted images and high signal intensity on T2 weighted images.[4,5] MRI can also be used in the detection of a cryptorchid testis and in detecting the difference in signal intensity in case of tumour occurrence.

The case we have described comes under Type IV category according to Leung classification since there is complete duplication of testis, epididymis and vas deferens and Type 1 according to the classification by Singer et al. In this case the supernumerary testis showed diffuse homogeneous decreased echotexture (hypoechoic) in comparison to the native left testis on grey scale ultrasound examination while Colour Doppler examination revealed a significant diffuse increase in vascularity. MRI examination revealed homogeneous hypointensity of the supernumerary testis on T2W images. In addition abdominal screening by ultrasonography and MRI revealed multiple enlarged iliac, inguinal, pre and para aortic lymph nodes which showed features of necrosis on ultrasonography while MRI showed them to have decreased intensity. A possibility of seminomatous transformation of the supernumerary testis was considered in view of the abnormal imaging appearance of the supernumerary testis and the abdominal lymph nodes and the patient was so advised. Since patient refused admission, he was lost for further follow-up.

As mentioned in literature, Polyorchidism is currently treated conservatively in absence of associated complicating conditions like cryptorchidism, torsion or malignancy. Close observation by physical examination, tumor markers and imaging studies (ultrasonography and MRI) is advocated in such cases. Surgical treatment is advocated in presence of associated complications like cryptorchidism, torsion or malignancy. Orchidectomy was the advocated treatment for all patients in the pre-sonography era due to risk of torsion and malignancy especially in patients who had completed their families. Presently, owing to the advances in MRI imaging a more conservative approach is preferred.

CONCLUSIONS

Polyorchidism, or supernumerary testicle is an extremely rare congenital anomaly of the urogenital system, frequently presenting as triorchidism and most commonly located either superiorly or inferiorly to the native testis. Polyorchidism is most often diagnosed by ultrasonography due to its characteristic sonographic features. Magnetic resonance imaging is helpful in confirming the sonographic findings and in detecting associated complicating conditions like cryptorchidism or malignancy. Irrespective of its location malignant transformation can occur within the supernumerary testis.

Accurate diagnosis of Polyorchidism can be made solely on the basis of imaging due to the high resolution imaging modalities currently available. Characteristic imaging findings of the supernumerary testis helps in ultrasonographic diagnosis of the same and the use of MRI overrides the need for histopathological confirmation.

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