Multiparametric ultrasound findings in a patient with polyorchidism

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Abstract

Polyorchidism is a rare condition usually incidentally discovered in young patients investigated with ultrasound for unrelated reasons. It is characterized by the presence of unilateral or, rarely, bilateral supernumerary testes which, depending on the type of polyorchidism, may have their own epididymis and vas deferens. Ultrasound, including B-mode and color Doppler technique, represents the primary imaging modality for the evaluation of scrotal diseases, including the characterization of supernumerary testes, which normally appear identical to the ipsilateral testicular parenchyma on every ultrasonographic technique performed. The role of MRI is thus limited to confirming ultrasonographic findings and excluding the presence of malignancy. Contrast-enhanced ultrasound is a recently introduced ultrasonographic technique providing detailed and sensitive visualization of the perfusion pattern of structures. It can thus be added to the conventional ultrasonographic examination in order to establish the diagnosis, negating the use of more expensive and time-consuming imaging modalities. We present a young patient with an incidentally found supernumerary testis, where new distinctive ultrasonographic findings, like the identification of transmediastinal vessels and a cystic appendage along with the perfusion pattern on contrast-enhanced ultrasound, established the diagnosis of polyorchidism.

Keywords

Polyorchidism, contrast-enhanced ultrasound, pediatric, scrotum

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Introduction

The term “polyorchidism” derives from the Greek words “poly-” meaning multiple and “orchi-” meaning testis, referring to the presence of more than two testes usually within the left hemiscrotum.^{1,2} It is a very rare congenital anomaly with a little less than 200 cases reported.² In most patients with polyorchidism there is only one supernumerary testis (triorchidism),¹ while there are patients with bilateral supernumerary testes reported.^{2,3} Extra testes are usually intrascrotal, situated either superior or inferior to the ipsilateral normal testis, but in some cases they are found abutting or being completely separated from a maldescended testis, or they may present as maldescended themselves located outside the scrotum.¹

Ultrasound (US) is the first-line modality for the evaluation of scrotal masses including polyorchidism.⁴ It is believed that cryptorchid supernumerary testes entail an increased risk for malignant transformation,

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justifying ultrasonographic surveillance.² Contrast-enhanced ultrasound (CEUS) is a useful complementary ultrasonographic technique, which could be potentially used alternatively to establish the diagnosis of a supernumerary testis and exclude the presence of neoplasia. Among CEUS advantages are good tolerability, safety, dynamic evaluation of perfusion and the potential to reach a diagnosis in the same visit with conventional ultrasound, reducing patient anxiety and possibly negating the need for more time consuming and expensive techniques such as MRI.⁵

In this case report, we present new distinctive ultrasonographic findings of a supernumerary testis in a patient with polyorchidism, where CEUS was used to evaluate the perfusion pattern of the incidentally found lesion. Among these findings, we describe the identification of transmediastinal blood vessels and a cystic appendage in the supernumerary testis. To our knowledge, this is the first report of a supernumerary testis with a cystic appendage and the use of CEUS for the characterization of polyorchidism.

Case presentation

A 15-year-old boy was presented to the Pediatric Surgery Outpatient Department complaining of pain in the left hemiscrotum. The pain was mainly located in the area of the body and tail of epididymis. His medical history included congenital torsion of the contralateral testis necessitating orchiectomy. The boy was referred for scrotal ultrasound, performed with an Aloka ProSound alpha 7 (Hitachi Aloka Medical Ltd), with a frequency range 5–13 MHz linear matrix array transducer with harmonics and 36 mm scan width. In the area of pain in the left hemiscrotum, US revealed hyperemia of the body and tail of epididymis. Note was also made of an 8 × 11 mm ovoid mass

situated superior to the ipsilateral testis and attached to the head of epididymis (Figure 1(a)). On B-mode technique, the mass showed echogenicity similar to that of the normal testicular parenchyma. The head of epididymis appeared hyperechoic in comparison with the mass and the testis (Figure 1(a)). The left testis itself measured 24 × 30 mm. Power Doppler technique was used to assess the lesion's vascularity demonstrating a similar pattern of blood flow signals with the normal testis. Within the lesion, an obliquely oriented blood vessel was identified, resembling normal testis' transmediastinal blood vessels (Figure 1(b)). A small cystic structure with posterior acoustic enhancement was found attached to the superior pole of the lesion (Figure 1(b)). There was only one epididymis identified inside the left hemiscrotum. The boy was treated conservatively for the epididymitis.

Differential diagnosis of the incidentally found lesion included the rare paratesticular tumours with adenomatoid tumor being the commonest, discontinuous form of splenogonadal fusion and the very rare entity of wandering spleen. A CEUS examination was decided on to further characterize the lesion by a detailed evaluation of its vascularity. The patient underwent a follow-up ultrasound 5 days after his presentation and conservative treatment. Conventional ultrasound confirmed the location of the head of epididymis between the left testis and the lesion (Figure 2(a)) along with its normal size and echogenicity. Power Doppler technique showed lower vascularity of the epididymis compared to the testis and the lesion while bridging vessels could be seen at the lesion's lower pole (Figure 2(b)). The lesion's vascularity was also evaluated using eFlow technique (Figure 2(c))—a flow visualization, color-based technique characterized by spatial resolution higher than color Doppler. Following parental informed consent,

Figure 1. (a) Initial US examination identified an ovoid mass of similar echogenicity and smaller size than the ipsilateral testis, situated within the left hemiscrotum. (b) Power Doppler technique identified an oblique blood vessel resembling testicular transmediastinal blood vessels. Note was also made of a small cystic structure with posterior acoustic enhancement, attached to the lesion's superior pole (arrowhead in 1b).

Figure 2. (a) Follow-up ultrasonographic examination showing the epididymis with normal size and echogenicity, clearly situated between the left testis and the ovoid lesion. (b) Power Doppler technique identified again the transmediastinal blood vessel along with some “bridging” vessels (arrowhead in 2b). (c) eFlow technique image confirming the vascularity pattern of the lesion.

Figure 3. (a) Following the bolus intravenous administration of 2.4 mL of US contrast agent, low MI harmonic CEUS images revealed moving microbubbles within the lesion in a similar enhancement pattern of the lesion and the ipsilateral testis on both arterial and delayed phase. (Also see supplementary video with the online version of this article at: <http://journals.sagepub.com/home/ULT>) (b) Increased contrast was noted between the perfused parenchyma and the non-enhancing cystic appendix.

an intravenous cannula was placed in the antecubital fossa and 2.4 mL of SonoVue (Bracco, Milan, Italy) were administered with a bolus injection. The contrast-enhanced part of the exam was performed with the same linear probe and a user-defined, low MI and harmonic protocol. There were moving microbubbles identified within the lesion, which showed similar enhancement pattern to the ipsilateral normal testis, in the arterial, venous, and delayed phase (Figure 3 and supplementary video—available with the online version of this article at: <http://journals.sagepub.com/home/ULT>). As a result, the possibility of a tumor or ectopic spleen was low, whereas the diagnosis of a supernumerary testis was considered likely. Surgical resection and histologic examination of the lesion confirmed the diagnosis of a supernumerary hypoplastic testis with a cystic appendage (Figure 4).

Figure 4. Intraoperative photograph showing the left testis and the lesion representing a supernumerary testis with a small cystic appendage.

Discussion

Polyorchidism is usually diagnosed in adolescents or young adults presenting with a painless scrotal mass or incidentally in patients examined for hydrocele, epididymitis, infertility, or other unrelated causes.^{1,2,6} Alternatively, it can be diagnosed in the emergency setting when complicated by torsion of the hemiscrotum or the supernumerary testis alone.⁷ In some cases, microlithiasis affects patients with polyorchidism although no association is established so far.³ The occurrence of malignant tumors like seminoma or germ cell tumors in supernumerary testis has been described and explains the potential need for follow-up with ultrasonographic surveillance of these patients.⁸

The ultrasonographic hallmark of polyorchidism is the identification of a scrotal mass, which appears identical to the ipsilateral testis on every ultrasonographic technique performed, including echogenicity pattern on B-mode.^{1,6,9} This mass may be found attached to the ipsilateral testis or separated and located superiorly or inferiorly. It has been observed that the sum of lengths of the supernumerary and ipsilateral testis is approximately equal to that of the contralateral side testis, possibly due to the stipulated embryologic division theory.¹ The supernumerary testis may thus be of equal size to the ipsilateral testis or appear smaller and hypoplastic,¹⁰ as in our case. Color Doppler technique reveals blood flow signals within the mass similar to the ipsilateral normal testis, while in some cases “bridging” arteries may be seen connecting the two testes.^{1,9,11} The US findings may differ when the supernumerary testis is multicystic (rete testis) or when affected by microlithiasis. In the former case, multiple cysts of varying size can be seen within the extra testis while in the latter case note is made of fine echogenic foci.¹² There is also a report of a lobulated supernumerary testis with an internal septum.¹³

Differential diagnosis of polyorchidism includes extratesticular masses like lipomas, adenomatoid tumours or scrotal calculi.^{4,14,15} Polyorchidism should also be differentiated from the discontinuous type of splenogonadal fusion, a rare congenital anomaly where a mass of splenic tissue is abutting the testis mimicking a supernumerary testis. CEUS and elastography have been used to characterize splenogonadal fusion with the ectopic splenic tissue demonstrating marked contrast uptake during both the arterial and the delayed phase, similarly to spleen but in contrast with the testicular parenchyma.¹⁶

CEUS is performed after the intravenous administration of ultrasound contrast agents consisting of microbubbles. These agents are characterized by an excellent safety profile and remain strictly within the

blood vessels, acting as intravascular tracers. As a result, CEUS has been used to evaluate scrotal lesions, offering detailed visualization of vascularity with higher sensitivity compared to color Doppler technique, as it can detect microvessels as small as 2–7 μm . It can thus readily differentiate structures with no perfusion, like calculi or epidermoid cysts, from ectopic testicular or splenic parenchyma and neoplasias characterized by a different perfusion pattern.^{5,16–18} In our case report, we were able to demonstrate the similar enhancement pattern of an incidentally found extratesticular mass to the ipsilateral testis, helping to establish the diagnosis of polyorchidism. The diagnosis was also supported by the identification of a “transmediastinal artery” within the extratesticular lesion. This vessel can be found in half of normal testes, representing a branch of the testicular artery.⁴ It is thus a finding suggestive of normal testicular parenchyma and helps establish the diagnosis of a supernumerary testis, as it was the case in our patient. Finally, a small cystic appendage was identified attached to the supernumerary testis in our case; a finding not described previously. Testicular appendages represent embryonic remnants, which are commonly found attached to the testicular parenchyma and which may occasionally be cystic in nature.¹⁴ Our case highlights the identification of cystic appendages in supernumerary testes—a finding potentially demonstrating the nature of the incidentally discovered lesion.

Conclusion

Accurate and timely differential diagnosis of intrascrotal focal lesions is currently facilitated with the introduction of modern ultrasonographic techniques, including newer flow visualization techniques and primarily CEUS. These modalities provide detailed and sensitive evaluation of vascularity and perfusion patterns of incidentally found scrotal masses, helping to establish the diagnosis. The demonstration of a lesion’s perfusion pattern similar to testicular parenchyma on CEUS established the diagnosis of polyorchidism in this case report.

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Ethical approval

Written consent was obtained from the parents of the patient to publish the images and details of the case.

Guarantor

DS

Contributors

VR and MA researched the literature. VR and DR performed ultrasound and contrast-enhanced ultrasound examination and analyzed the data. DS examined the patient and performed surgery confirming histologic diagnosis. VR and MA wrote the first draft of the manuscript. VR and DR performed corrections and prepared the final version of the manuscript. All authors reviewed and approved the final version of the manuscript.

Supplemental Material

Supplementary material for this paper can be found at <http://journals.sagepub.com/doi/suppl/10.1177/1742271X16689808>

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