J Ped Surg Case Reports 8 (2016) 22–26

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Congenital prepubic sinus: A case report and review of the literature

Tatsuma Sakaguchi [a,*], Yoshinori Hamada [a], Yusuke Nakamura [a], Hiroshi Hamada [a], Takeshi Shirai [a], Hirotsugu Yanai [b], Masanori Kon [b]

[a] Division of Pediatric Surgery, Department of Surgery, Kansai Medical University, 2-5-1, Hirakata, Osaka 573-1191, Japan
[b] Department of Surgery, Kansai Medical University, 2-5-1, Hirakata, Osaka 573-1191, Japan

ARTICLE INFO

Article history:
Received 28 February 2016
Accepted 21 March 2016

Key words:
Congenital prepubic sinus
Ultrasound
Magnetic resonance imaging

ABSTRACT

Congenital prepubic sinus (CPS) is an extremely rare anomaly, which is often associated with purulent discharge from a midline opening overlying the pubis. CPS was first described by Campbell et al. in 1987 and they suggested that it might represent a variation in normal embryological development. Several theories have been proposed regarding the pathogenesis of CPS. However, the etiology of CPS is still unclear because the anatomical and pathological features of CPS often differ from each other. We report a case of CPS and review the literature to improve the global understanding of CPS.

1. Case report

A female neonate had a small midline opening overlying the pubis since birth. Purulent discharge was observed 1 month after birth and it then improved. After 2 years and 6 months, she was referred to our department for recurrence of discharge. She had no other symptoms and no evidence of urinary tract infection. A local examination showed a prepubic sinus with brown discharge and reddening around the sinus (Fig. 1). An ultrasound (US) examination revealed a subcutaneous cyst in front of the pubis that measured 15 × 13 × 11 mm (Fig. 2). Magnetic resonance imaging (MRI) showed a cystic lesion and thin tract to the deeper side in the subcutaneous layer (Fig. 3). We could not insert a plastic tube far enough into the sinus tract for conventional fistulography. Three months later, after the inflammation was relieved, we performed an operation. The opening of the skin was connected to the subcutaneous cyst. The sinus tract attached to the cyst extended to the pubis (Fig. 4). We injected indigo carmine into the sinus to confirm that the sinus was obstructed in front of the pubis and the sinus was excised. Histologically, the distal lumen of the sinus was lined with stratified squamous epithelium and the proximal lumen was lined with transitional epithelium (Fig. 5). The postoperative course has been uneventful without recurrence.

2. Discussion

To the best of our knowledge, there are 29 reports of CPS in the English literature (44 cases; Table 1) including 23 males and 21 females, aged 1 month to 22 years old [1–30]. CPS most commonly presents in infancy with discharge from a midline opening between the dorsal penile root/clitoris to the suprapubic region. Sinus tracts

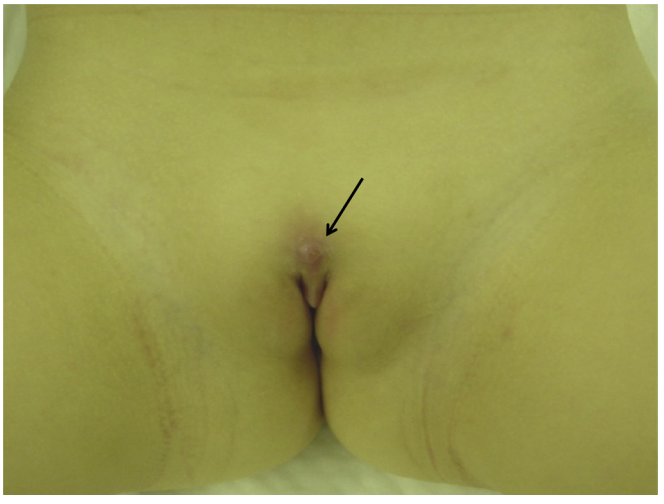

Fig. 1. Midline prepubic opening with a reddish appearance and discharge (black arrow).

* Corresponding author.
E-mail address: sakaguct@hirakata.kmu.ac.jp (T. Sakaguchi).

http://dx.doi.org/10.1016/j.epsc.2016.03.012

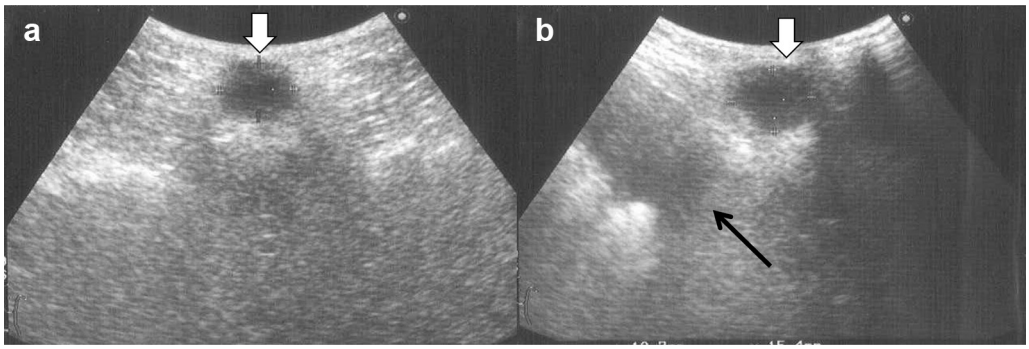

Fig. 2. US examination. (a) Horizontal scan. A subcutaneous cyst (white arrow) can be seen. (b) Sagittal scan. A subcutaneous cyst (white arrow) and bladder (black arrow) can be seen.

passed above the pubis in 16 cases, through the pubic symphysis in five cases, and below the pubis in six cases. The distal end of the sinus tract reached to the bladder wall in 25 cases, the umbilicus in five cases, the urethra in two cases, the retropubic space in five cases, the pubic symphysis in two cases, and the prepubic space in four cases. Histologically, only squamous or stratified squamous epithelium was reported in 15 cases, only transitional epithelium in six cases, both squamous/stratified squamous and transitional/urothelial epithelium in 20 cases, and cylindrical or columnar epithelium in four cases. Smooth muscle bundles surrounded the sinus in 18 cases. Connection of the sinus with the bladder was reported in two cases.

Several embryological theories have been proposed for the development of CPS. The first theory considers a localized failure of midline fusion in the lower abdominal wall [1]. During the 4th gestational week, closure of the anterior abdominal wall is complete. The cephalic border of the cloacal membrane, moving downward from the base of the umbilical cord, results in formation of the infra-umbilical anterior abdominal wall by fusion of the lateral folds. Any closure defect will cause anomalies, including omphalocele, bladder exstrophy, and epispadias. At the 9th week, a persistent cloacal membrane above the genital tubercle interrupts complete fusion, causing various anomalies, including diastasis of the pubic symphysis, bifid clitoris, separation of the corpus cavernosum, and epispadias [31]. Some cases of CPS were accompanied by diastasis of the pubic symphysis [1,18]. The presence of stratified squamous epithelium in the entire tract might support this theory.

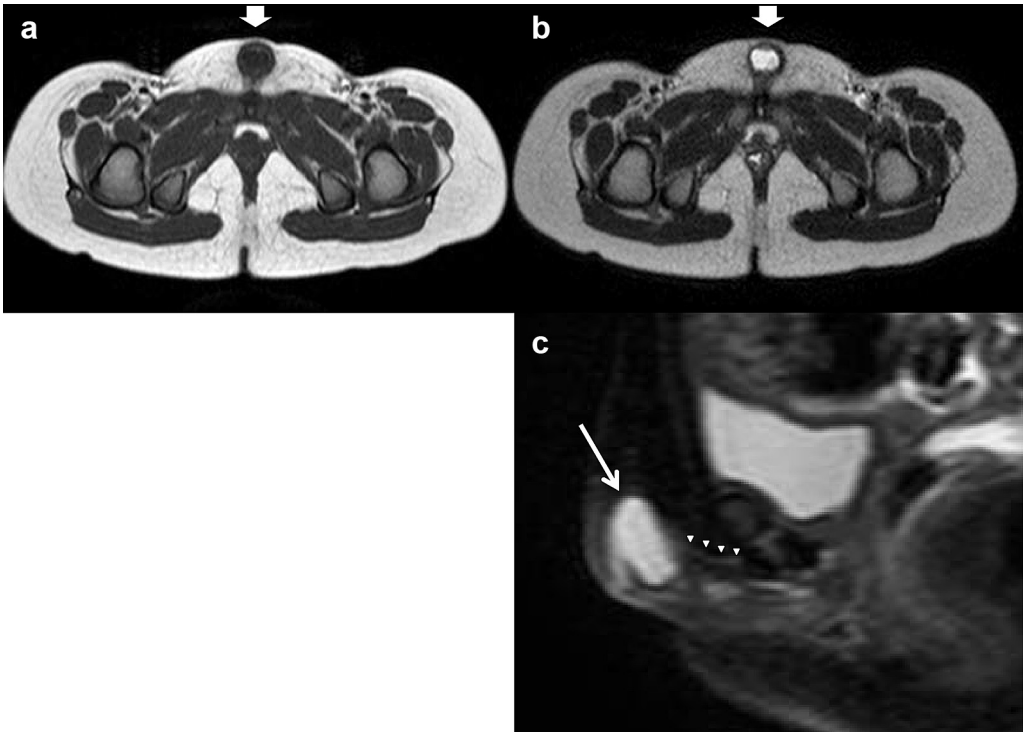

Fig. 3. MRI. (a) Horizontal scan of T1-weighted imaging. A subcutaneous cyst (white arrow) is shown. (b) Horizontal scan of T2-weighted imaging. A subcutaneous cyst (white arrow) can be seen. (c) Sagittal scan of T2-weighted imaging. A subcutaneous cyst (white arrow) and thin tract in the subcutaneous layer (white triangle) can be seen.

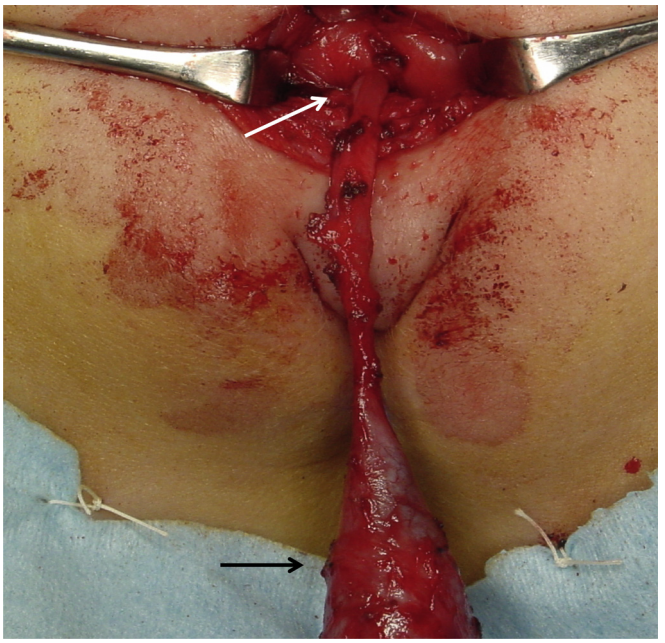

Fig. 4. View of the operation. A subcutaneous cyst (black arrow) and sinus tract extending to the pubis are shown (white arrow).

The second theory considers a variant of dorsal duplication of the urethra [1]. The urethra develops from the urogenital sinus to meet the ectodermic glandular plate [31]. An anomaly at any of the levels may lead to complete or incomplete dorsal urethral duplication. The ending of the sinus by a fibrous tract constantly links to the bladder or to the urethra. The presence of a transitional urothelium in the proximal part of the sinus favors this theory [8]. Stephens described three types of dorsal urethral duplication according to the anatomy [32]. Type 1 is a complete or incomplete tandem channel that runs parallel to the normal urethra from the glans to the bladder, and joins the urethra or ends blindly. Type 2 is an epispadiac type of channel from the dorsum of the penis to the bladder or it joins the urethra at some point. Type 3 is a dermoid sinus that simulates an accessory urethra, but tracks from the base of the penis in front of the pelvic urethra and bladder behind the pubic symphysis to or toward the umbilicus. The anatomy of our case is similar with the type 3 variant of Stephen's classification.

The third theory considers CPS as a congenital fistula of the primitive urogenital sinus. Soares-Oliveira et al. classified CPS with three anatomical types as follows: high, toward the urachal remnant; middle, to the bladder; and low, to the prostatic urethra [15]. From our review, 25 cases were the middle type, five cases were the high type, and two cases were the low type. The fourth theory considers that a residual cloacal membrane and umbilicophallic groove cause CPS. If the umbilicophallic groove traps a small portion of the cloacal membrane while it moves downward from the base of the umbilical cord, the residual cloacal membrane in the umbilicophallic groove may be turned inside and tubularized during longitudinal growth of the embryo [19].

Complete excision of the sinus is necessary to prevent recurrence of symptoms and late malignant change [4]. We could not perform conventional fistulography in our patient. However, preoperative MRI, especially T2-weighted imaging, was useful to display the tract extending to the pubis.

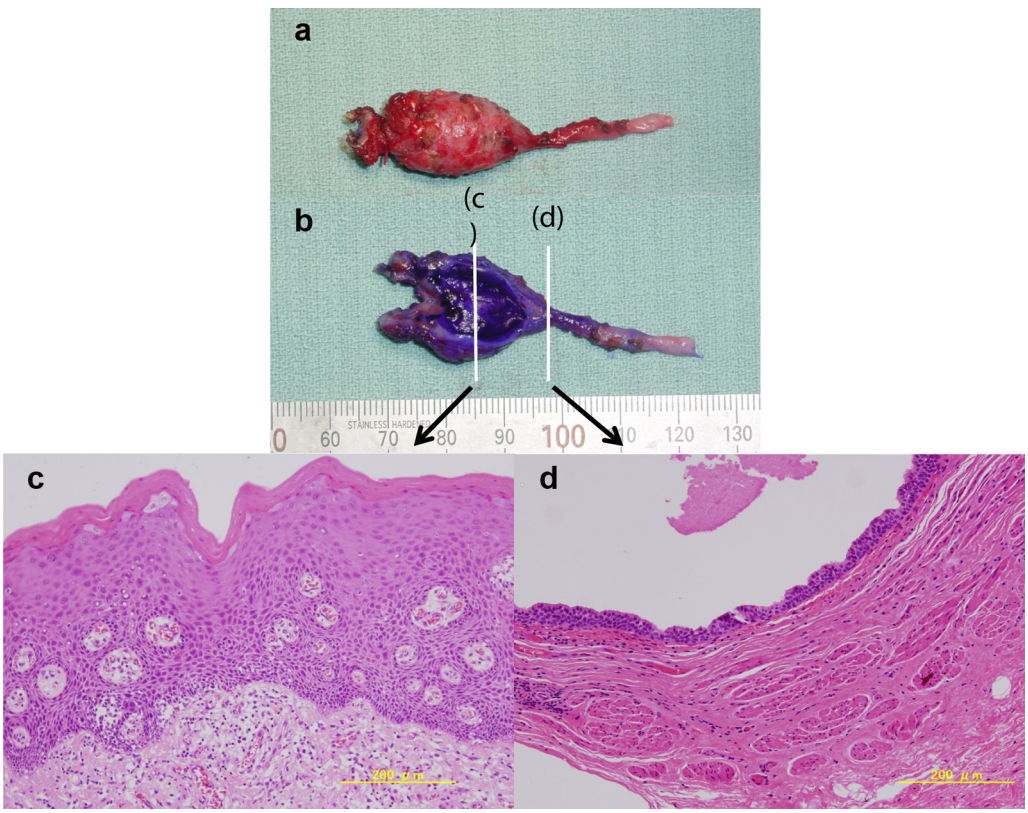

Fig. 5. (a) Excised sinus. (b) The lumen was dyed by indigo carmine. (c) Histologically, the distal lumen of the sinus is lined with stratified squamous epithelium. (d) The proximal lumen is lined with transitional epithelium.

Table 1
Reported cases of CPS.

Year	Age	Gender	Symptom	Opening	Tract	Distal end	Histology (epithelium)	Histology (smooth muscle bundles)
1987	4M	F	Discharge	Prepubis	Above pubis	Bladder	Proximal transitional; distal squamous	+
	6M	M	Opening	Prepubis	Above pubis	Bladder	Stratified squamous	+
	2Y	F	Groin swelling	Above clitoris	Above pubis	Bladder	Proximal transitional; distal squamous	
1989	18M	F	Discharge	Suprapubis	Below pubis	Bladder	Transitional	
1990	10M	F	Discharge	Prepubis	Above pubis	Bladder	Squamous	
1992	2M	M	Polypoid opening	Prepubis	Through pubic symphysis	Bladder	Transitional	+
	2Y	F	Discharge	Prepubis	Above pubis	Bladder	Pseudstratified	
	2Y	F	Opening	Prepubis	Above pubis	Retropubic space	Transitional	+
1993	1M	F	Discharge	Prepubis	Below pubis	Umbilicus	Proximal transitional; distal squamous	
	1M	F	Discharge	Prepubis	Above pubis	Umbilicus	Proximal transitional; distal squamous	
1993	4M	F	Discharge	Above clitoris	Above pubis	Umbilicus	Stratified squamous	+
1994	11M	M	Discharge	Suprapubis	Above pubis	Bladder	Proximal transitional; distal squamous	+
	3Y	M	Red and swollen	Prepubis	Above pubis	Bladder	Squamous	+
1994	2M	F	Discharge	Above clitoris	Through pubic symphysis	Bladder	Stratified squamous	
1995	1Y	F	Discharge	Above clitoris	N.D.	Retropubic space	Squamous, transitional, columnar	
1996	8Y	F	Opening	Above clitoris	Below pubis	Retropubic space	Squamous and transitional	+
1997	10M	F	Pustule	Above clitoris	Below pubis	Urethra	Stratified squamous	
1997	10M	M	Discharge	Dorsal radix of penis	Above pubis	Bladder	Stratified squamous, cylindrical	
	5Y	M	Discharge	Prepubis	Above pubis	Bladder	Squamous	
	4Y	M	Discharge	Dorsal penile root	Above pubis	Bladder	Stratified squamous	
1998	5Y	M	Discharge	Suprapubis	N.D.	Abdominal wall	Squamous, columnar	
2001	2M	F	Discharge	Above clitoris	N.D.	Bladder	Proximal transitional; distal squamous	+
	3M	M	Discharge	Prepubis	N.D.	Bladder	Proximal transitional; distal squamous	+
	1M	M	Discharge	Prepubis	N.D.	Bladder	Proximal transitional; distal squamous	+
	14Y	M	Discharge	Dorsal penile root	N.D.	Prepubic space	Proximal transitional; distal squamous	+
	2M	F	Discharge	Prepubis	N.D.	Dorsal tunica albuginea clitoris	Squamous	
2002	8M	M	Discharge	Prepubis	Below pubis	Prostatic urethra	Stratified squamous	
	5M	M	Discharge	Prepubis	N.D.	Pubic symphysis	Stratified squamous	
2002	5Y	M	Discharge	Prepubis	N.D.	Retropubic space	Proximal ciliated columnar, middle transitional; distal stratified squamous	
2003	4Y	F	Discharge	Supurapubis	N.D.	Umbilicus	Transitional	
2003	2Y	M	Discharge	Dorsal radix of penis	N.D.	Bladder	Proximal transitional; distal squamous	+
2004	3M	M	Opening	Prepubis	Through pubic symphysis	Umbilicus	Proximal transitional; distal stratified squamous	+
	4Y	F	N.D.	N.D.	N.D.	Bladder	Transitional	+
2004	8Y	M	Discharge	Dorsal radix of penis	Above pubis	Bladder	Stratified squamous	+
2005	3Y	M	Discharge	Dorsal radix of penis	N.D.	Pubic symphysis	Stratified squamous	+
2006	12Y	F	Clitromrgaly	Above clitoris	N.D.	Retropubic space	Squamous	
2007	3Y	M	Discharge	Dorsal radix of penis	Above pubis	Bladder	Proximal urethral; distal squamous	
2010	4M	M	Discharge	Prepubis	Above pubis	Bladder	Stratified squamous, transitional, cylindrical	
2011	4Y	M	Discharge	Dorsal radix of penis	N.D.	Prepubic space	Squamous	
2013	9M	M	Discharge	Dorsal radix of penis	Below pubis	Bladder	Transitional	
2013	10M	F	Discharge	Prepubis	Through pubic symphysis	Bladder	Proximal transitional; distal stratified squamous	
2015	16Y	F	Clitoral and left labial swelling	N.D.	Through pubic symphysis	Bladder	Proximal transitional; distal squamous	+
2015	22Y	F	Fever and abdominal pain	Prepubis	Below pubis	Bladder	Proximal urethral; distal squamous	
2015	3Y	M	Discharge	Prepubis	Below pubis	Prepubic apace	Proximal urethral; distal squamous	
2016	2Y	F	Discharge	Prepubis	Above pubis	Umbilicus	Proximal urethral; distal squamous	

N.D. indicates not described.

Conflicts of interest

None.

References

[1] Campbell J, Beasley S, McMullin N, Hutson JM. Congenital prepubic sinus: possible variant of dorsal urethral duplication (Stephens type 2). J Urol 1987; 137(3):505–6.
[2] Crawford RA, Sethia KK, Fawcett DP. An unusual presentation of a urachal remnant. Br J Urol 1989;64:315–6.
[3] Rozanski TA, Kiesling Jr VJ, Tank ES. Congenital prepubic sinus. J Pediatr Surg 1990;25:1301.
[4] Lawson A, Corkery JJ. Prepubic sinus: an unusual urachal remnant. Br J Surg 1992;79:573.
[5] Groff DB. Suprapubic dermoid sinus. J Pediatr Surg 1993;28:242–3.
[6] Park WH, Choi SO, Park KK, Kwon KY. Prepubic dermoid sinus: possible variant of dorsal urethral duplication (Stephens type 3). J Pediatr Surg 1993; 28:1610–1.
[7] Komura J, Yano H, Kanazawa M, Tanaka Y, Takagishi T, Sasaguri Y. Congenital prepubic sinus. Pediatr Surg Int 1994;9:287–9.
[8] Daher P, Diab N, Moussa C, Korkmaz G. Congenital prepubic sinus. Eur J Pediatr Surg 1994;4:119–21.
[9] Chou TD, Chu CC, Diau GY, Chiang JH. Subpubic sinus: a remnant of cloaca. J Urol 1995;153:1671–2.
[10] Walther MM, Woodard JR. Subpubic fistula: a urethral duplication. J Urol 1996;155:1728–9.
[11] Green JS, Madden NP. Congenital prepubic sinus: a form of dorsal duplication? Br J Urol 1997;80:964.
[12] Ergun O, Sayan A, Arikan A. Congenital prepubic sinus: possible variant of dorsal urethral duplication. Eur J Pediatr Surg 1998;8:380–1.
[13] Nirasawa Y, Ito Y, Tanaka H, Seki N. Urachal cyst associated with a suprapubic sinus. Pediatr Surg Int 1999;15:275–6.
[14] Huang CC, Wu WH, Chai CY, Wu TC, Chuang JH. Congenital prepubic sinus: a variant of dorsal urethral duplication suggested by immunohistochemical analysis. J Urol 2001;166:1876–9.
[15] Soares-Oliveira M, Julia V, Aparicio LG, Morales L. Congenital prepubic sinus. J Pediatr Surg 2002;37:1225–7.
[16] Chao HM, Chuang CJ, Chen KC, Chu CC, Chou JM. Unusual epithelium in a subpubic sinus. Pediatr Surg Int 2002;18:494–5.
[17] Park WH, Choi SO. An unusual urachal sinus with external opening in the midline suprapubic area. J Pediatr Surg 2003;38:E18–20.
[18] Al-Wattar KM. Congenital prepubic sinus: an epispadiac variant of urethral duplication: case report and review of literature. J Pediatr Surg 2003;38(4): E10–2.
[19] Tsukamoto K, Yamataka A, Kuga T, Yanai T, Watanabe T, Miyano T. Congenital prepubic sinus: is it a residual cloacal membrane and umbilicophallic groove? Pediatr Surg Int 2004;20:47–50.
[20] Samujh R, Sinha A, Ojha S, Menon P, Rao KL. Congenital prepubic sinus. Indian J Pediatr 2004;71(5):477–8.
[21] Usami M, Hayashi Y, Kojima Y, Maruyama T, Tozawa K, Kohri K. Congenital prepubic sinus: a variant of dorsal urethral duplication (Stephens type 3). Int J Urol 2005;12(2):231–3.
[22] Hayase M, Kojima Y, Hayashi Y, Maruyama T, Sasaki S, Kohri K. Pseudoclitoromegaly associated with congenital prepubic sinus. Int J Urol 2006; 13(7):1031–2.
[23] Kim HY, Shin OR, Jang ED, Yoon BI, Chung MS, Chung YS, et al. Congenital prepubic sinus: a variant of urethral duplication. Korean J Urol 2007;48: 881–4.
[24] Sasaki Y, Deguchi E, Tsukada S, Ono S, Iwai N. Usefulness of magnetic resonance imaging for congenital prepubic fistula. J Pediatr Surg 2010;45:1734–6.
[25] Ozdemir E, Yildiz T, Kanbay M, Kanbay S. A case of congenital prepubic sinus. Eur J Pediatr Surg 2011;21(6):408–9.
[26] Nasir AA, Abdur-Rahman LO, Olaoye I, Oyinloye AO, Bamigbola KT, Adeniran JO. Congenital prepubic sinus: a variant of epispadiac dorsal urethral duplication. J Pediatr Urol 2013;9(1):e82–5.
[27] Yamada K, Kanamori Y, Tanaka H, Fujino A, Watanabe T, Takeda N, et al. Congenital prepubic sinus associated with a urachal remnant: report of a case. Surg Today 2013;43(11):1330–2.
[28] Shaw JS, Caldamone AA, Rardin CR. Congenital prepubic sinus in an adolescent female: a case report and review of the literature. Urology 2015;85(1):227–9.
[29] Kobayashi H, Nomura T, Haneda Y, Sawada N, Araki I, Takeda M. Congenital prepubic sinus accompanied by prevesical abscess. Clin Pract 2015;5(1):701.
[30] Celebi S, Kuzdan Ö, Sander S, Gündüz N, Özaydın S, Yavuz S. Congenital prepubic sinus with remnant tissue mimics corpus spongiosum: a rare case. Eur J Pediatr Surg Rep 2015;3(1):43–5.
[31] Moore KL. The developing human: clinically oriented embryology. 4th ed. Philadelphia: WB Saunders Co; 1988. p. 258–61.
[32] Stephens FD. Abnormal embryology-cloacal dysgenesis. In: Congenital malformation of the urinary tract. New York: Praeger Scientific; 1983. p. 15–52.