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Patient Report

Supernumerary nipple: Should we be alert?

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Supernumerary nipples (SN) are common minor congenital anomalies. SN pose no medical threat, however they can be associated with other medical conditions especially kidney and urinary tract malformations. We present here a female infant with SN with interesting associated malformations. In our patient, umbilical hernia, umbilical granuloma, spina bifida occulta and bilateral ovarian cyst accompanied the accessory nipple, instead of the expected renal or urinary system anomalies.

Case Report

A 53-day-old female infant presented with the complaint of a skin fold present in the lower back ever since birth. Her mother was 28 years old and gave birth to her vaginally at the 40th week of pregnancy following a healthy pregnancy. The infant was the mother's second pregnancy and second living offspring. Postnatal history was unremarkable. The family history was also unremarkable. The mother and father were non-consanguineous. Physical examination findings of the mother and father were normal. No family history of similar physical anomalies, malignancy, cardiovascular or renal diseases was present. On physical examination the infant's weight (6 kg; 75–90%), height (61 cm; 75–90%), and head circumference (43 cm; 75–90%) were normal; her vital findings were stable, her general condition was well and she was active and lively. Approximately 2 cm below the right nipple, an accessory nipple without areola was seen (Fig. 1). Umbilical hernia and umbilical granuloma were present. Gluteal cleft was in bifid form. On her right hip, pili asymmetry was observed. No atypical facial appearance, skeletal anomaly or mental–motor retardation findings were found. In laboratory investigations, hematological and biochemical abnormalities were normal. On radiological examination, in ultrasonographic screening of the whole abdomen, no anomaly was seen apart from ovarian cysts. In the ultrasonographic examination of the accessory nipple, it was observed to consist solely of the nipple, without breast tissue. According to the supernumerary nipple classification by Kajava,[1] the supernumerary nipple in our patient was classified as polythelia. Serial graphics were taken for a probable skeletal anomaly, but no anomalies were found. Echocardiography, taken to determine a possible cardiac anomaly (like mitral valve prolapse), was normal. Transfontanel ultrasonography was also normal. In a magnetic resonance imaging investigation of the sacral region, spina bifida occulta (Fig. 2), slight dysplasia in the right hip, two bilateral ovarian cysts on the left (11 and 8 mm) and one ovarian cyst on the right (9 mm) were established.

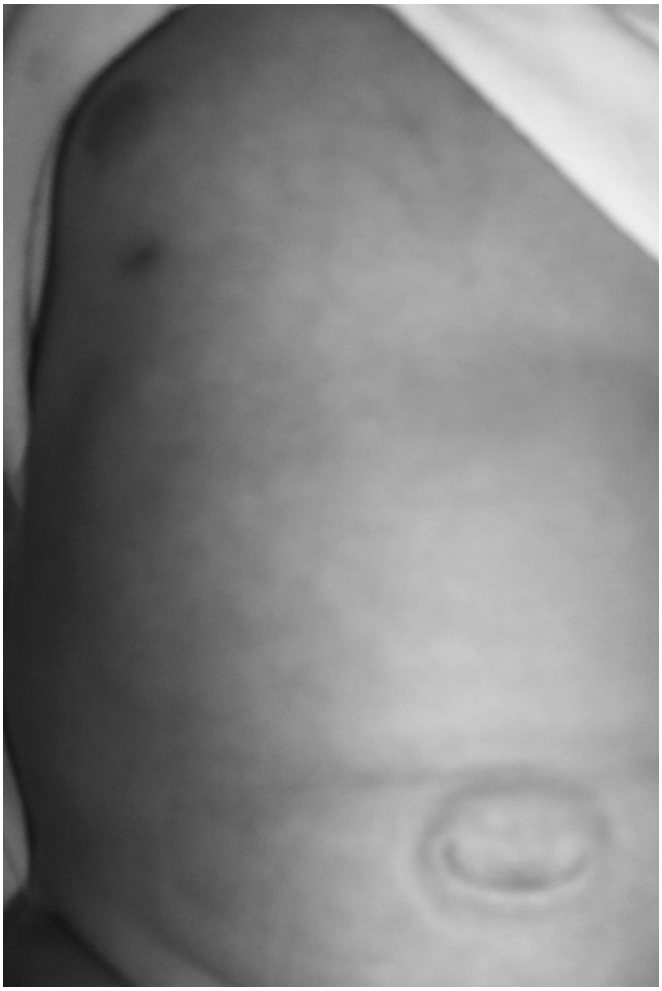

Fig. 1 Supernumerary nipple.

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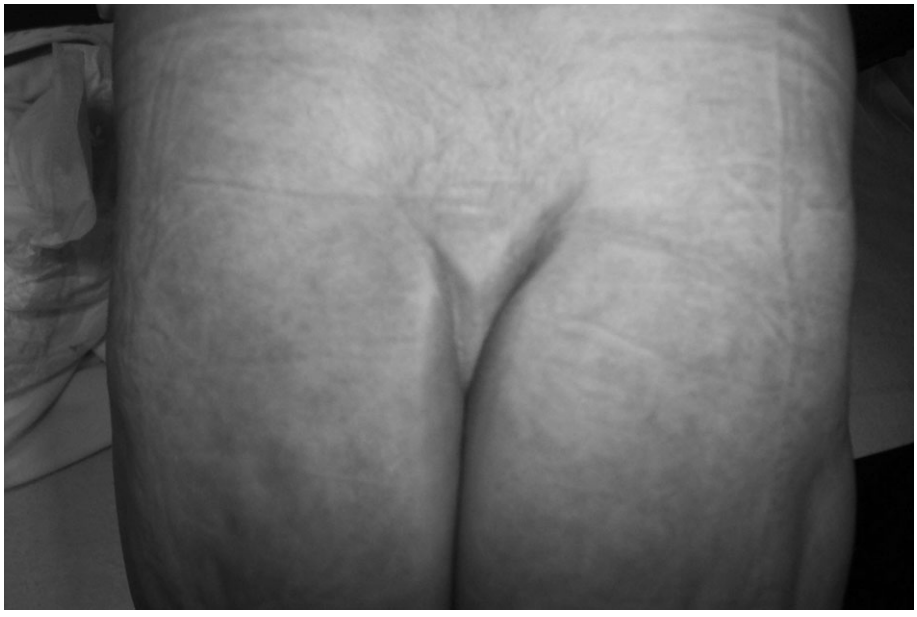

Fig. 2 Gluteal skin fold with underlying spina bifida.

The pediatric surgery department was consulted regarding the umbilical granuloma and hernia. The granuloma was cauterized with silver nitrate and the patient was followed up for hernia and ovarian cysts.

The orthopedics department was consulted regarding the hip dysplasia and a pelvic bandage was applied.

Discussion

Supernumerary nipples (solitary or multiple) usually arise within the embryonic milk lines but they can also occur in locations such as the back, thigh, vulva and neck.[1–5] The frequency of this condition ranges from 0.2% to 5.6% depending on various factors.[6] Although much has been written on the association between SN and other conditions, this still remains as a controversial and speculative area. The major association of supernumerary nipples is with urinary tract anomalies. Apart from characteristic patterns of well-defined syndromes, the only specific association found was the higher prevalence of supernumerary nipples in patients with isolated (and hidden) urinary tract abnormalities of various types (9.4% vs 1.8% in controls; $P < 0.005$).[7] Kidney malformations, urinary tract malformations (renal agenesis, polycystic kidney disease, supernumerary kidney, renal adenocarcinoma, megaureter ureteropelvic junction obstruction, etc.)[4] and malignancies (testicular cancer)[5] are the most frequent associations with SN. Other medical conditions sporadically associated with SN include vertebral anomalies, cardiovascular system disorders, central nervous system problems, chromosomal abnormalities, genetic syndromes, gastrointestinal disease, skeletal disease (spina bifida, diastematomyelia, scoliosis, etc.)[8] and cutaneous disorders.[4] Genetic disorders associated with SN include Simpson–Golabi–Behmel syndrome, Char syndrome, Hailey–Hailey disease, Bartsocas–Papas syndrome, trisomy 2p and some spondylocostal dysostosis syndromes with different patterns of inheritance.[8] Our patient demonstrated supernumerary nipple associated with spina bifida, hip dislocation, umbilical hernia with umbilical granuloma and bilateral ovarian cysts. Until more conclusive data is identified linking SN with other medical conditions, a high index of suspicion should be maintained in these patients. With this case report we hope to encourage dermatologists, pediatricians and family physicians to screen for hidden anomalies that could be associated with SN and to work together to form an algorithm for the management of patients with SN.

References

1 Brown J, Schwartz RA. Supernumerary nipples. An overview. *Pediatr. Dermatol.* 2003; **71**: 344–6.
2 Leung AK, Robson WL. Renal anomalies in familial polythelia. *Am. J. Dis. Child* 1990; **144**: 619–20.
3 Meggyessy V, Mehes K. Association of supernumerary nipples with renal anomalies. *J. Pediatr.* 1987; **111**: 412–13.
4 Lewis EJ, Crutchfield CE, Prawer SE. Accessory nipples and associated conditions. *Pediatr. Dermatol.* 1997; **14**: 333–4.
5 Goedert JJ, Mckeen EA, Javadpour N *et al*. Polythelia and testicular cancer. *Ann. Intern. Med.* 1984; **101**: 646–7.
6 Ferrara P, Giorgio V, Vitelli O *et al*. Polythelia: Still a marker of urinary tract anomalies in children? *Scand. J. Urol. Nephrol.* 2009; **43**: 47–50.
7 Méhes K, Pintér A. Minor morphological aberrations in children with isolated urinary tract malformations. *Eur. J. Pediatr.* 1990; **149**: 399–402.
8 Panigrahi I, Saxena A, Marwaha RK. Congenital scoliosis, supernumerary nipples and spina bifida occulta. *Clin. Dysmorphol.* 2008; **17**: 215–18.