CASE REPORT

Accessory scrotum with perineal lipoma/hamartoma mimicking penoscrotal pseudoduplication

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ABSTRACT

Congenital anomalies of the scrotum are thought to be caused by an abnormal migration of the labioscrotal swelling during the embryological process. We report the case of an infant in whom suspected penoscrotal pseudoduplication was apparent at delivery. Imaging using ultrasound scan and magnetic resonance imaging allowed anatomical delineation of the abnormality and the appropriate surgical intervention to be determined. Full surgical correction was done at 4 weeks of age with good outcome. We discuss the differential diagnosis and the management of congenital scrotal abnormalities.

KEYWORDS:
Perineal lipoma/hamartoma; Penoscrotal pseudoduplication; Sacrococcygeal teratoma; Diagnostic dilemma.

INTRODUCTION

Congenital anomalies of the scrotum are uncommon, and are thought to be caused by an abnormal migration of the labioscrotal swelling, though the exact embryological process remains unclear [1]. Four categories of abnormalities are described: penoscrotal transposition, bifid scrotum, ectopic scrotum, and accessory scrotum; the latter two anomalies being extremely rare [1]. Perineal lipomas are known to be associated with an accessory scrotum in >80% of cases [1]. This paper describes a case of accessory scrotum with perineal lipoma or hamartoma, and the associated challenges in management.

CASE REPORT

A male infant born at 34 weeks gestation with a birth weight 1.96 kg was admitted to the Special Care Baby Unit for supporting his feeds and temperature regulation. There were no maternal
risk factors for infection or family history of any congenital abnormality. Antenatal ultrasound scan at 20 weeks gestational age had been unremarkable.

Perineal examination showed suspected penoscrotal duplication located in the midline, posterior to anatomically normal male genitalia and anus (Figure 1). There were no other abnormalities found on neurological or abdominal examination, and the baby was otherwise well.

Ultrasound examination (Figure 2) showed normal renal tract with homogenous hyperechoic tissue identified within the accessory scrotum; no communication or connection was shown between the accessory scrotum and the anus or bladder. Testes were identified within the anterior (normally situated) scrotum. He was discharged home at 2 weeks of age with plan for further investigations and surgical management of the anomaly.

Pelvic magnetic resonance imaging (MRI) scan performed at 3 weeks showed a predominantly external lesion demonstrating high T1 and T2 signal with complete loss of signal on fat saturation images. The mass was confined to the intra-gluteal region, contained no solid or cystic components, and with no post-contrast enhancement (Figure 3). The remainder of the pelvic structures were normal.

At 4 weeks, the lesion was resected with the coccyx completely en bloc through an elliptical perineal wound which was closed in the midline.

Figure 1. Appearance immediately after birth.

Figure 2. Longitudinal ultrasound scan showing homogenous hyperechogenicity consistent with fatty tissue.
natal cleft. The postoperative period was uneventful. Histology demonstrated the mass to be comprised of mature adipose tissue with some intersecting fibrous septa, vessels and nerve trunks; no immature, teratomatous, or malignant tissue was seen, and the resection margins were clear. The histopathological diagnosis was fibrolipoma/lipoma with the presence of some sort of organisation showing mature adipose tissue organised somehow by intersecting fibrous septa, the lesion is also representing a perineal hamartoma forming an accessory scrotum (Figure 4). He was reported to be well at a clinic review 8 weeks later.

**DISCUSSION**

This case demonstrates the challenges of managing congenital abnormalities of the scrotum. Our patient was initially considered to have penoscrotal duplication as the appearance of the abnormality closely resembled that

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**Figure 3.** Pelvic MRI scan (A) sagittal and (B) coronal post-contrast fat saturated T1 weighted images showing homogenously low (saturated) signal suggestive of fat. No solid or enhancing elements.

**Figure 4.** Histology showing features of perineal fibrolipoma/lipoma. (A) Haematoxylin and eosin (H+E) stain×2, (B) H+E stain×10.
of an incompletely formed duplication of male genitalia. Penoscrotal duplication is an extremely rare anomaly affecting approximately one in every 5 million births, in which there is duplication of the phallus frequently associated with variable abnormalities of the scrotum, bladder and other ano-rectal anomalies [2-5].

Accessory scrotum is characterised by additional scrotal tissue, containing no testes, which is located beside a normally developed scrotum [1]. Absence of associated anomalies, combined with ultrasonography and MRI findings, indicated that lipoma or sacrococcygeal teratoma were diagnostic possibilities in our case, with the former being confirmed on histology following complete surgical excision.

Congenital perineal lipoma/hamartoma is considered to be extremely rare though there is no published incidence data [6]. The terms lipoma and hamartoma, once there is no central core of hyaline cartilage, are difficult to distinguish histologically; the differentiation between perineal lipoma and hamartoma in this context has no clinical relevance [7]. As the appearance may mimic that of external male genitalia, when congenital perineal lipoma/hamartoma occurs in a female infant, the condition may be mistaken for ambiguous genitalia [8]. A previous case report with both perineal lipoma and accessory scrotum, confirmed by histological examination was described [9]. Fathaddin [10] reported a rare case of accessory scrotum with congenital perineal lipoma in association with type 2 congenital pulmonary airway malformation. Most published reports of congenital perineal lipoma describe cases that have been diagnosed after delivery, though antenatal diagnosis on ultrasonography has also been described [11,12].

Differential diagnosis of a congenital perineal mass in the newborn includes hamartoma, sacrococcygeal teratoma, haemangioma, partial or complete genital duplication, lipoma, lipoblastoma, and incomplete conjoined twins [1,13,14]. Radiological imaging using ultrasound and MRI scan should be undertaken to delineate the internal urogenital and ano-rectal anatomy and to identify other associated abnormalities [13]. In cases where perineal lipoma is suspected, complete excision is recommended; the lesion is usually benign and does not recur [13].

CONCLUSION

Congenital perineal lipoma/hamartoma with accessory scrotum can cause diagnostic dilemma before specialist referral. There should be early discussion between relevant specialists, and radiological imaging to determine the precise anatomy and likely diagnosis, and to exclude any associated anomalies. The outcome of perineal lipoma/hamartoma (without additional abnormalities) after surgical excision is usually good.

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CONFLICT OF INTERESTS

The authors declare no conflict of interests.

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ETHICAL APPROVALS

Signed informed consent for participation and publication of medical details was obtained from the parents of this child. Confidentiality of patient’s data was ensured at all stages. The authors declare that ethics committee approval was not required for this case report.

REFERENCES


