

# CASE REPORT

Nagoya J. Med. Sci. 77. 501 ~ 506, 2015

## Accessory scrotum with perineal lipoma diagnosed prenatally: case report and review of the literature

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### ABSTRACT

We report a case of accessory scrotum (AS) in the perineal region with peduncular lipoma, diagnosed prenatally. A male fetus of 31 weeks' gestation was referred to our department with a perineal mass. Prenatal ultrasonography and magnetic resonance imaging showed a mass of 1.0 × 1.2 cm located posterior to the scrotum. No other abnormalities were noted during pregnancy. The patient was delivered vaginally at 38 weeks of gestation. On physical examination, a soft peduncular mass with a rugged and pigmented swelling was located between the normally developed scrotum and the anus. There were no specific symptoms or any other associated congenital anomalies. We completely excised the mass at one month of age. A histological examination revealed lipoma, with tissue suggestive of scrotum, so a definite diagnosis of AS was made. AS is a rare congenital anomaly of the scrotum. We review the literature.

Key Words: congenital scrotal anomaly, accessory scrotum, perineal lipoma, neonate, prenatal diagnosis

### INTRODUCTION

Congenital scrotal anomalies are unusual and include penoscrotal transposition, bifid scrotum, ectopic scrotum, and accessory scrotum (AS). Among these, AS is the least frequent, with only 42 cases reported in the English literature.<sup>1-40</sup> AS is characterized by additional scrotal tissue lacking a testis, besides a normally developed scrotum. Various associated anomalies have been reported. In particular, contiguous subcutaneous tumor is the most frequently associated abnormality and is reported to be related to the etiology of AS.

Although prenatal screening techniques have advanced, most reported cases of congenital perineal mass have been identified after birth. We report a rare case of AS with peduncular lipoma diagnosed prenatally.

### CASE REPORT

A 28-year-old woman was referred to our hospital for the evaluation of a fetal perineal mass at a gestational age of 31 weeks. Prenatal ultrasonography and magnetic resonance imaging

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Received: January 20, 2015; accepted: February 18, 2015

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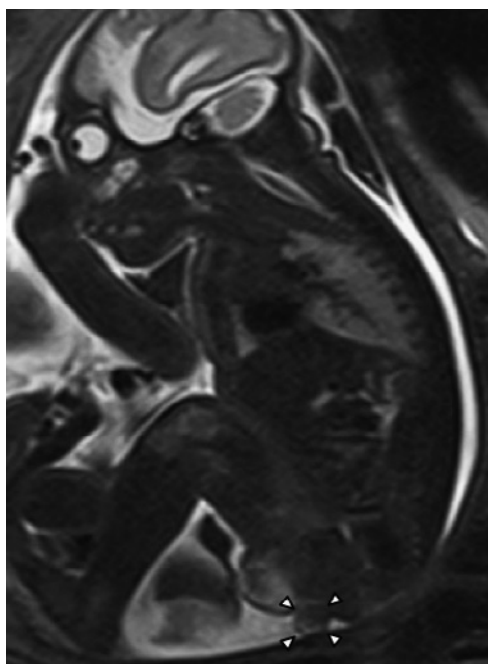
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(MRI) showed a mass of  $1.0 \times 1.2$  cm located posterior to the scrotum in a male fetus. (Fig. 1) The likely diagnosis was lipoma and the mass maintained a stable appearance until delivery. No other abnormalities were noted during the whole gestation. The male newborn was delivered vaginally at 38 weeks of gestation and his body weight was 2208 g. There were no specific symptoms after birth.

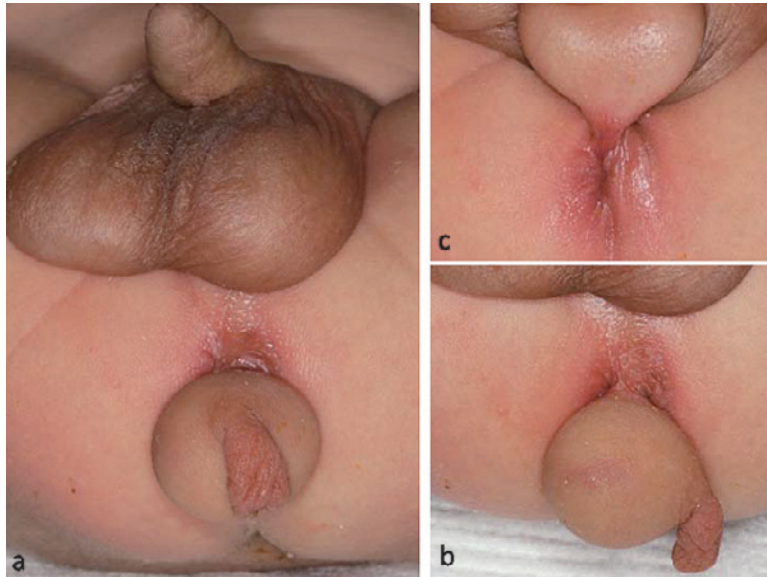
On physical examination, the soft peduncular mass, measuring 2.0 cm in diameter, was attached to a midperineal skin tag. There was also a rugged pigmented swelling on the mass, measuring 0.7 cm in diameter, which resembled the scrotum. (Fig. 2) The normal testes could be palpated in both the developed scrotal sacs. There were no additional abnormalities of the external genitalia. On MRI, the perineal mass showed high signal intensity on T1- and T2-weighted images and the signal intensity was suppressed on fat-suppressed T1-weighted images. MRI revealed no associated abnormalities of the intraabdominal organs, musculoskeletal system, or genitourinary system.

The preoperative diagnosis was AS with perineal lipoma, and we completely excised the mass under general anesthesia at one month of age. The postoperative course was uneventful, and the patient was discharged on the day after the operation. There was no recurrence or functional sequelae within a follow-up period of six months. A histological examination revealed that the peduncular mass consisted of mature adipose tissue. In this case, it was difficult to distinguish between lipoma and normal adipose tissue pathologically. However, our clinical diagnosis was lipoma because the peduncular mass was separated from normal perineal region by the skin tag. The rugged swelling on the peduncular mass showed smooth muscle fibers in the subcutaneous layer, which represented the tunica dartos. (Fig. 3) The swelling was definitively diagnosed as AS.

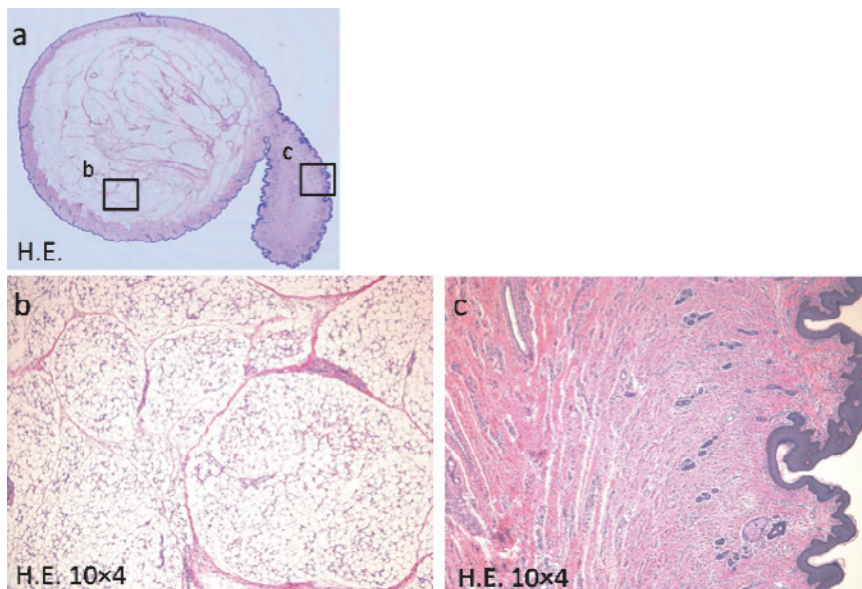


**Fig. 1** MRI at 31 weeks of gestation. Arrowheads indicate a  $1.0 \times 1.2$  cm mass located posterior to the scrotum.

## Accessory scrotum with perineal lipoma



**Fig. 2** a. A soft peduncular mass with a rugged and pigmented swelling is located posterior to the normally developed scrotum.  
 b. Lateral view.  
 c. The mass is attached at the midperineum with a skin tag.



**Fig. 3** Histological examination.  
 a. Macroscopic image  
 b. The peduncular mass consists of mature adipose tissue.  
 c. Smooth muscle fibers in the subcutaneous layer of the rugged swelling represent the tunica dartos.

## DISCUSSION

The scrotum is formed by the fusion of the labioscrotal swellings. These swellings appear at four weeks of gestation and migrate to the caudal portion after 12 weeks of gestation. Abnormal migration or early division of the labioscrotal swellings is possibly related to the etiology of congenital scrotal anomalies.<sup>9)</sup> The least frequent congenital scrotal anomaly is AS,<sup>25)</sup> characterized by additional scrotal tissue without a testis, besides a normally developed scrotum. Forty-three cases have been reported, including ours.<sup>1-40)</sup> (Table)

Various anomalies associated with AS have been reported. In particular, contiguous subcutaneous tumor has a high incidence (72.5%) of association with AS. Histologically, one case of subcutaneous tumor was lipoblastoma,<sup>13)</sup> three cases were hamartoma,<sup>1,12,21)</sup> and the others were consistent with lipoma. (Table) It is assumed that the contiguous subcutaneous tumor is related to the etiology of AS. Sule hypothesized that AS develops when intervening mesenchymal tissue (i.e., the developing subcutaneous tumor) disrupts the continuity of the developing caudal labioscrotal swelling.<sup>26)</sup> However, the complete etiology of AS is not explained by this hypothesis because AS can occur with no contiguous tumor. Takayasu hypothesized that AS develops from the early division and teratoid growth of pluripotential labioscrotal tissue elements.<sup>39)</sup> Most ASs including ours were located in the perineal region. However, two cases associated with skeletal abnormalities were located in the pubic area,<sup>5, 10)</sup> and one case was located on the distal penile shaft.<sup>23)</sup>

Only three cases, including ours, have been detected prenatally. Our case was detected at a gestational age of 31 weeks, and the other two reported cases were detected at 24 weeks<sup>11)</sup> and 32 weeks<sup>8)</sup> of gestation. A congenital perineal mass is unusual in itself, and most reported cases have been diagnosed after birth.<sup>41,42)</sup> However, detection is possible with careful prenatal

**Table** Characteristics of accessory scrotum

		n = 43 (1949–2014)
Sex (male)		100% (43/43)
Age at operation	median (range)	9 months (4 days–46 years)
Prenatal diagnosis		7.0% (3/34)
Associated anomalies		
	Contiguous subcutaneous tumor	72.5% (29/40)
	Hamartoma	10.3% (3/29)
	Lipoblastoma	3.4% (1/29)
	Anorectal malformation	18.6% (8/43)
	Other scrotal anomalies	16.3% (7/43)
	Hypospadias	9.3% (4/43)
	Pseudodiphallia	4.7% (2/43)
	Skeletal abnormalities	4.7% (2/43)
	Renal dysplasia	4.7% (2/43)
	Spina bifida	4.7% (2/43)
	Retrocerebellar arachnoid cyst	2.3% (1/43)
	Vertebral abnormality	2.3% (1/43)

screening. The differential diagnosis of a fetal perineal mass includes lipoma, lipoblastoma, infantile hemangioma, hamartoma, and choristoma.<sup>41)</sup> If a fetal perineal mass is detected during the antenatal period, it is important to look for any associated congenital anomalies.<sup>42)</sup>

The prognoses of surgically treated patients are good, and only one has died, from an associated anomaly before surgery.<sup>22)</sup> The reported ages at surgery range from four days to 46 years (median, nine months), and three adult cases are recorded in the literature. Our patient was operated upon in the neonatal period because the mass was considered to be excisable without complications and the associated subcutaneous tumor had a low probability of malignancy.<sup>13)</sup>

## CONCLUSION

We experienced a rare case of AS with peduncular lipoma. Although a fetal perineal mass is difficult to diagnose, it can be detected with careful prenatal screening. Many ASs are associated with contiguous subcutaneous tumors, which are assumed to be related to the etiology of AS.

## CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest.

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