Child's Nervous System (2019) 35:1207–1211
https://doi.org/10.1007/s00381-019-04148-6

ORIGINAL ARTICLE

Filar cysts in rare cases may progress in size, particularly when associated with filar lipoma

Kyoichi Seo[1] · Hirofumi Oguma[1] · Rieko Furukawa[2] · Akira Gomi[3]

Received: 24 September 2018 / Accepted: 2 April 2019 / Published online: 10 April 2019

Abstract

Purpose Filar cysts (FCs) are detected incidentally on ultrasonography (US) of the neonatal spine. Their clinical significance has not been widely discussed in the literature because FCs are usually asymptomatic. This study aimed to investigate the clinical features of FCs and distinguish FCs that warrant attention.

Methods We retrospectively analyzed 396 patients with lumbosacral skin stigmata. Patients who were younger than 1 month old at reference underwent US initially, and those older than 1 month of age underwent magnetic resonance imaging (MRI) at the age of 5–12 months. Patients with an FC in the US underwent subsequent MRI at the age of 5–12 months. Patients with an FC were followed clinically for at least 3 years.

Results FCs were identified in 56 (14.1%) patients. Of the 195 children who underwent US initially, FCs were detected in 49 (25.1%) children. FCs were detected in seven children who underwent MRI initially. Of the 50 children with FCs who underwent MRI at the age of 5–12 months, FCs in 20 patients (40%) showed natural regression and FCs in 30 patients (60%) remained. Two of these 30 patients showed progression in size of the FC, and in both cases, the FCs were associated with a filar lipoma; however, the resected cysts were not neoplastic and did not have obvious clinical significance.

Conclusions Our study characterizes clinical features of filar cysts. Two-thirds of FCs remained in late infancy. The best sequence of MRI to follow-up FCs is heavily T2-weighted images.

Keywords Filar cyst · Ventriculus terminalis · Ultrasonography · Natural history · CISS image

Introduction

Filar cysts (FCs) are sometimes found on lumbar spine sonography as isolated, anechoic, cyst-like structures bounded by echogenic septations that exist within the midline or lateral of the filum terminale just below the conus medullaris [1–7]. An FC is similar to a terminal ventricle (TV) and is sometimes confused with a TV [5, 10]. A TV is a small, ependymal-lined cavity within the conus medullaris. The terminal part of the human spinal cord develops through three main steps: (1) secondary neurulation, (2) canalization, and (3) retrogressive differentiation. A TV is thought to be a remnant of this canal, caused by unsuccessful retrogressive differentiation [4, 5, 7]. However, the etiology of FCs is unknown [1, 3]. A normal filum terminale is a long, thin, fibrous filament and a product of complete retrogressive differentiation. It is therefore considered that either excessive canalization or incomplete retrogressive differentiation may be responsible for FC formation [3, 5]. And, an FC may represent a remnant of neural tissue from the caudal cell mass undergoing normal apoptosis [3].

Because FCs are usually asymptomatic and often show natural regression, FCs are considered normal variants [1, 3, 7], and their clinical significance and natural history have not been widely discussed in the literature. Spinal ultrasonography (US) has become more widely used and has improved in accuracy during recent years [1, 2]. As a result, cystic lesions of the filum terminale are more frequently being detected. This has led to a need for guidelines of clinical assessment

✉ Akira Gomi
gomi@jichi.ac.jp

[1] Department of Neurosurgery, Jichi Medical University, 3311-1 Yakushiji, Shimotsuke, Tochigi, Japan

[2] Department of Pediatric Medical Imaging, Jichi Children's Medical Center Tochigi, Jichi Medical University, 3311-1 Yakushiji, Shimotsuke, Tochigi, Japan

[3] Department of Pediatric Neurosurgery, Jichi Children's Medical Center Tochigi, Jichi Medical University, 3311-1 Yakushiji, Shimotsuke, Tochigi, Japan

for neonates and infants with an FC. Therefore, this study aimed to obtain information on clinical features and natural history of FCs, and to determine characteristics of FCs that require long-term follow-up.

Patients and methods

This study was approved by the Office of Research Integrity Review Board of Jichi Medical University. We retrospectively analyzed 473 patients with skin stigmata in the lumbosacral coccygeal area who were referred to our institution between 2006 and 2014. Among these patients, 396 underwent US and/or magnetic resonance imaging (MRI). The patients' ages ranged from the first day of life to 66 months. Patients who were aged younger than 1 month at the corrected age (the chronological age minus the number of weeks born premature) at reference underwent US initially because the spinous processes were not yet ossified [1]. Patients with an FC in the US underwent subsequent MRI at the age of 5–12 months. Patients who were aged older than 1 month at reference underwent MRI initially at the age of 5–12 months. We recommended subsequent MRI 6 to 12 months later to the patients with an FC in the first MRI. Patients with an FC were followed for at least 3 years until 2018. Clinical features of the patients were evaluated using a computer record search.

US imaging was performed using an Acuson Antares Premium Edition machine (Siemens, Tokyo) by two expert radiologists. Images were obtained in the prone position with no sedation. An FC was defined as an anechoic structure within the filum terminale demonstrating a fusiform shape in the longitudinal plane and a circular shape in the transverse plane. MRI was performed with a Magnetom Avanto (1.5 T, Siemens, Tokyo) in the supine position. Sequences included sagittal T1-weighted, axial T1-weighted, sagittal T2-weighted, axial T2-weighted, and heavily T2-weighted (CISS: constructive interference in the steady state) images without contrast material. We calculated the product of the longitudinal distance and the transverse distance of the maximum plane, and defined "regression" as completely disappeared, "decrease" as a > 50% decrease in size, "no change" as < 50% decrease to < 25% increase, and "enlargement" as a > 25% increase in size.

Results

Of the 195 children younger than 1 month old at their corrected age who underwent US as the first investigation, FCs were detected in 49 children (25.1%), and the mean age of those with FCs was 8 days old (range 1–55 days) (Fig. 1). Of the 49 patients with an FC, 43 patients underwent subsequent MRI at the age of 5–12 months (mean 5.8 months). Three patients dropped out because

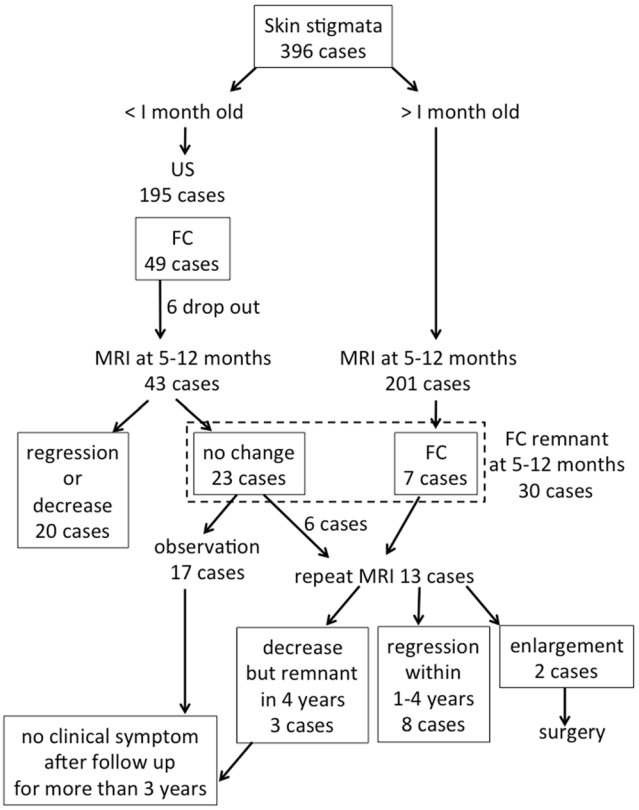

Fig. 1 Incidence of patients with filar cysts (FC) from magnetic resonance imaging (MRI) results of patients with skin stigmata, and evaluation and clinical results of patients with filar cysts. US ultrasonography

of transfer to another hospital, and three patients were excluded because of predisposing conditions. FCs disappeared or decreased in 20 patients (46.5%; Fig. 2), and remained the same size in 23 of 43 patients (53.5%; Fig. 3).

Of the 201 children with a skin stigmata who were aged older than 1 month at reference and underwent MRI as an initial investigation at the age of 5–12 months, FCs were detected in seven children (3.5%), and their mean age was 6.9 months (range 5–12 months).

In total, 56 of 396 patients (14.1%) with a skin stigmata had an FC. Of the 56 patients with an FC, 39 were boys and 17 were girls. Skin stigmata at the lumbosacral area of 56 patients with an FC appeared as a dimple in 54 patients and an abnormal gluteal crease in five patients (and both in three patients).

FCs in 20 of 50 patients (40%) showed natural regression within 6 months, and FCs in 30 of 50 patients (60%) showed a remnant at the age of 5–12 months. Of those with residual FCs, 13 consented to undergo a repeat MRI, and 17 patients were clinically followed up without repeat MRI. Enlargement of the FC in two patients was observed with MRI after 7 months in one patient and 1 year in the other. Both patients were cases with coexistence of an FC and a filar lipoma (Fig. 4). Both cases had no other congenital anomalies. We performed surgical resection of the FC and lipoma for these two patients to confirm the diagnosis. There were no

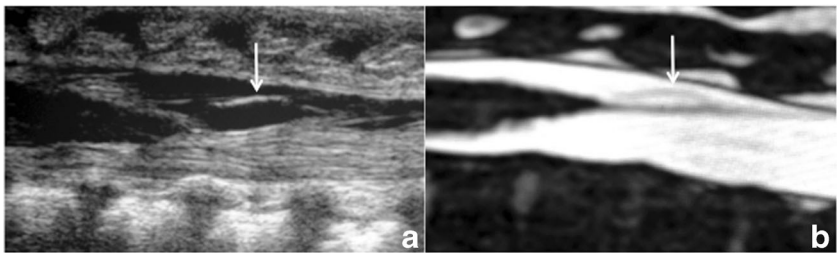

Fig. 2 An example case demonstrating a decrease of the filar cyst. **a** Longitudinal spinal ultrasonography showing a fusiform-shaped filar cyst just below the conus medullaris (arrow) at 2 days of age (corrected). **b** Heavily T2-weighted sagittal image showing shrinkage of the cyst (arrow) at 6 months of age (corrected)

surgical complications. The resected cysts showed no neoplastic findings. Of the remaining 11 patients who underwent subsequent repeat MRI, yearly MRI showed regression of the FC in 1–4 years in eight patients, and a decrease in size but residual FC in 4 years in three patients. Clinical evaluation after follow-up for more than 3 years showed no progressive clinical symptoms in 20 cases with a remaining cyst.

Coexistence of an FC and a filar lipoma was observed in eight cases. A comparison of FC without filar lipoma and FC with filar lipoma is shown in Table 1. The incidences of FCs for patients aged 5–7 months with or without a filar lipoma were not statistically different with Mann–Whitney's *U* test. However, the results show that the rate of cyst regression may be slower in the cases with a filar lipoma. Enlargement of FCs was found only in the cases with lipoma; however, there was no statistically significant difference as there were few cases without filar lipoma who underwent follow-up MRI.

Discussion

Few studies have focused on the epidemiology and natural history of an FC. Irani et al. have reported that FCs are found by US in up to 12% of young infants who are referred for spinal US [3]. The authors of the study described that the detection rate of FCs by US decreases with each month of age, with a detection rate of zero after 6 months of age. Their finding indicates a detection limit by US because evaluation of the spinal cord with US becomes difficult as the ossification of the spinous processes progresses. Choi et al. have reported that the incidence of FCs in patients with a sacral dimple who underwent an initial US within 6 months after birth is 24.8% [2]. In our series, 14% of infants with skin stigmata in the lumbosacral coccygeal area had an FC, and 25% of neonates had an FC. Furthermore, 60% of FCs remained at 5–12 months of age, although many FCs were predicted to shrink or disappear in the early infant period [6].

What additional evaluation should be done if an FC is found by US? An international survey by Ponger et al. showed that 84% of experts recommend MRI [6]. However, Irani et al. described that no cases who underwent follow-up MRI showed remnant FCs and that the difficulty of visualization on MRI was caused by limitations of the modality [3]. The authors did not mention the sequence of MRI and probably did not use the heavily T2-weighted imaging necessary for detection of FCs. In ordinary T1- or T2-weighted images, FCs often could not be clearly displayed (Fig. 4). Regarding the detection rate of FCs, heavily T2-weighted images exceed ordinary T1- or T2-weighted images. The best sequence of MRI to detect FCs appears to be heavily T2 (CISS)-weighted images [7].

To the best of our knowledge, our study is the largest reported series of MRI assessment of infants with FCs; to date, no reports are available on follow-up MRI studies for so many cases of FCs. We were able to perform follow-up MRI in most of the patients of our series. Although FCs have been postulated to regress in early infancy, only one third of FCs disappeared in the present study. Over several years, some FCs may shrink, but some FCs may remain.

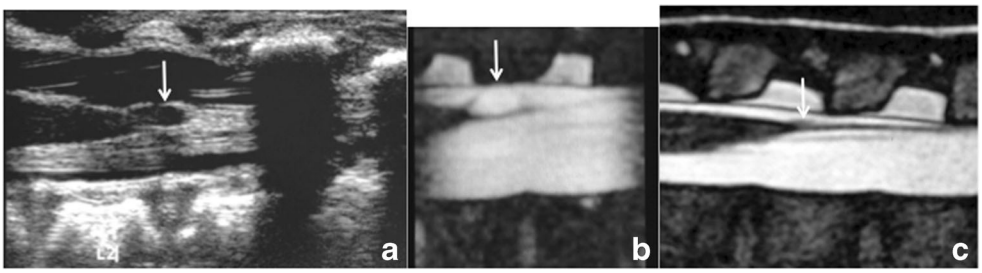

Fig. 3 An example case demonstrating a cyst with late regression. **a** Longitudinal spinal ultrasonography showing a fusiform-shaped filar cyst just below the conus medullaris (arrow) at 7 days of age. **b** Heavily T2-weighted sagittal image at 12 months of age showing no change of the cyst size (arrow). **c** Heavily T2-weighted sagittal image at 4 years of age showing regression of the cyst (arrow)

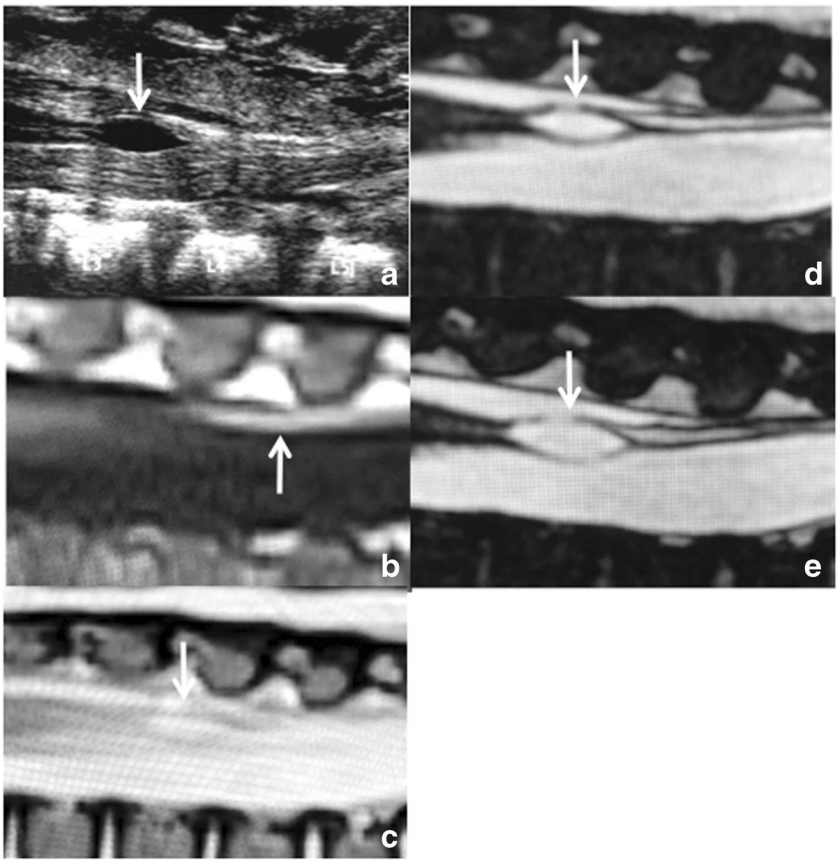

Fig. 4 An example case demonstrating cyst enlargement with a filar lipoma. **a** Longitudinal spinal ultrasonography showing a fusiform-shaped filar cyst just below the conus medullaris (arrow) at 7 days of age. **b–d** Magnetic resonance imaging performed at 7 months of age. **b** T1-weighted sagittal image showing filar lipoma (arrow). **c** T2-weighted sagittal image vaguely showing an FC (arrow). **d** Heavily T2-weighted sagittal image showing no change of the cyst size (arrow). **e** Heavily T2-weighted sagittal image at 1.5 years of age showing apparent enlargement of the cyst (arrow). Subsequently, we removed the cyst and cut the filum terminale to avoid tethered cord syndrome

Enlargement of an FC was observed in 2 (3.4%) of 59 patients in our series. Both infants had an FC associated with a filar lipoma. The identification of expanding cysts requires a differential diagnosis from other cystic or neoplastic diseases such as dermoids. We performed surgical resection of the cysts in two patients who had an expanding cyst to verify the pathological diagnosis. Both resected cysts were non-neoplastic simple cysts. These results suggest that surgical intervention is not necessary even if FCs of infant patients expand. Both expanding FCs existed in the midline of the filum terminale and appeared to be continuous to the central spinal canal (Fig. 4d, e). We suppose that the cause of FC enlargement was due to spinal cord tethering by a co-existing filar lipoma and continuity to the central spinal canal. However, in this study, we could not find any evidence of clinical repercussions, even in cases associated with lipomas.

Table 1 Comparison of clinical results of filar cysts with and without filar lipomas

FC			
	w/o lipoma $n = 42$	w/ lipoma $n = 8$	
FC aged 5–12 months	22 (52.4%)	7 (87.5%)	$p = 0.068$
MRI follow-up	6/22 (27.3%)	7/8 (87.5%)	$p = 0.003$
Enlargement	0/6 (0%)	2/7 (28.6%)	ns
Remnant	1/6 (16.7%)	1/7 (14.3%)	ns
Regression	5/6 (83.3%)	4/7 (57.1%)	ns

FC filar cyst, *MRI* magnetic resonance imaging, *ns* not significant

The potential for late complications from FCs remains unclear. Irani et al. have compared children with FCs with a control population and have described that FCs have minimal clinical significance as there is no difference in motor development [3]. However, their follow-up periods were short. The present study confirmed and expanded their findings, showing that 20 cases with residual FCs had no clinical symptoms in follow-up for more than 3 years. Recently, adult cases with symptomatic TV have been reported [8, 9]. It remains unknown if remnant or expanding cysts become symptomatic in adulthood, as in the adult cases with TV. This should be studied in future research.

Limitations

Our study was a retrospective review of a single institute. As the cases were limited to children with a dimple, the prevalence may be overestimated. Some elements, such as indication of additional MRI, were not defined. A multicenter, prospective study and a longer follow-up period are required to establish the clinical significance of FCs.

Conclusions

Our study showed that FCs were present in 14% of infants with skin stigmata in the lumbosacral coccygeal area and the disappearance rate in early infancy was one third. The best sequence of MRI to follow up FCs was heavily T2-weighted images. FCs may progress in size, particularly when associated with filar lipoma, but do not have obvious clinical significance. These findings provide clinical insight into the incidence and course of FCs in infants, and provide a foundation for future studies investigating the clinical significance of FCs.

Acknowledgments We thank Lesley McCollum, PhD, from Edanz Group (www.edanzediting.com/ac) for editing a draft of this manuscript.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there are no conflicts of interest.

Ethical approval This study was approved by the Office of Research Integrity Review Board of Jichi Medical University.

References

1. Brian DC, Marilyn JS (2011) Spinal ultrasonography. In: Siegel MJ (ed) Pediatric sonography, 4th edn. Lippincott Williams & Wilkins, Philadelphia, pp 647–674
2. Choi JH, Lee T, Kwon HH, You SK, Kang WK (2018) Outcome of ultrasonographic imaging in infants with sacral dimple. Korean J Pediatr 61:194–199
3. Irani N, Goud AR, Lowe LH (2006) Isolated filar cyst on lumbar spine sonography in infants: a case-control study. Pediatr Radiol 36: 1283–1288
4. Kriss VM, Kriss TC, Babcock DS (1995) The ventriculus terminalis of the spinal cord in the neonate: a normal variant on sonography. AJR 165:1491–1493
5. Lowe LH, Johanek AJ, Moore CW (2007) Sonography of the neonatal spine: part I, normal anatomy imaging pitfalls, and variations that may simulate disorders. AJR 188:733–738
6. Ponger P, Ben-Sira L, Ben-Adani L, Steinbok P, Constantini S (2010) International survey on the management of skin stigmata and suspected tethered cord. Childs Nerv Syst 26:1719–1725
7. Schwartz ES, Barkovich AJ (2019) Congenital anomalies of the spine. In: Barkovich AJ, Raybaud C (eds) Pediatric neuroimaging, 6th edn. Wolters Kluwer, Philadelphia, pp 973–1042
8. Severino R, Severino P (2017) Surgery or not? A case of ventriculus terminalis in an adult patient. J Spine Surg 3:475–480
9. Suh SH, Chung TS, Lee SK, Cho YE, Kim KS (2012) Ventriculus terminalis in adults: unusual magnetic resonance imaging features and review of the literature. Korean J Radiol 13:557–563
10. Unsinn KM, Geley T, Freund MC, Gassner I (2000) US of the spinal cord in newborns: spectrum of normal findings, variants, congenital anomalies, and acquired diseases. RadioGraphics 20: 923–938