Lumbosacral lipomyelomeningocele case series

2016

A retrospective review of all infants with lumbosacral lipoma seen at BCCH between 1997 and 2013 was carried out. The study population was stratified on the presence of a congenital, non-progressive deficit and subdivided on treatment approach. The subsequent developments of deficits resulting in untethering procedures were recorded.

Of the 44 infants in this study, 24 patients had no neurologic deficit while 20 patients had a fixed, non-progressive deficit evident at birth. Ten of 24 patients without a neurological deficit at birth underwent a prophylactic untethering with 3 eventually requiring repeat untethering after, on average, 62.7 months. Eleven of 14 asymptomatic, monitored patients required untethering for clinical deterioration. Two required a second untethering procedure after 48.7 months. Ten of 20 infants with congenital deficits present at birth underwent prophylactic untethering, and 4 required further surgery after 124 months. Ten patients underwent observation with 8 eventually requiring surgery. Two required repeat untethered after 154 months. The complication rates and operative burden for patients are similar whether prophylactic or delayed surgery is performed.

The presence of congenital neurologic deficit does not affect the likelihood of deterioration in patients managed expectantly; prophylactic detethering of these patients did not prevent delayed neurologic deterioration. Comparing the need for repeat surgery in prophylactically untethered patients with initial untethering of patients operated upon at the time of deterioration, prophylactic untethering may confer a benefit with respect to subsequent symptomatic tethering if complication rates are low. However, in a setting with multidisciplinary follow-up, a period of observation for patients and intervention when patients become symptomatic is an acceptable approach for patients with or without congenital deficits [1)].

A retrospective review of all patients with lumbosacral lipomas between 1997 and 2013 and who were managed without prophylactic surgery was performed. The clinical history and imaging results for each patient were reviewed in detail and then correlated to the pattern of and age at clinical deterioration.

Twenty-four patients were identified. Nine worsened within the first 18 months of life (early deterioration), and 15 patients deteriorated or remained stable after 30 months (late deterioration/stable). No patients worsened between 18 and 30 months of age. Patients who deteriorated early were more likely to have large intradural lipomas that filled the canal, increased during the 1st year of life, and compressed neurological structures. Some had a syrinx extending above the neural-lipoma interface. Syrinxes in patients with early deterioration were large and expanded in infancy. Patients with early deterioration had motor deficits at the time of deterioration, whereas patients with late deterioration developed mixed urological and motor dysfunction.

Patients with large lipomas displacing the cord and an enlarging syrinx have a propensity for early clinical deterioration. Given this, their families may be counseled that 1) the risk of deterioration in infancy may be higher than in infants without these features, and 2) they require more diligent

observation. Intervention before deterioration in these infants should also be considered. Patients without these features may be safely observed to a lesser extent [2].

2012

Over the past decade, children presenting with complex LSL to a single clinician at Great Ormond Street Hospital (GOSH), London, UK have undergone a thorough assessment focusing on neurological and urological evaluation and MRI of the lumbosacral spine. For children deemed to be asymptomatic, conservative management has been adopted with close periodic surveillance of neurological and urological function, thus avoiding untethering surgery unless symptomatic deterioration occurs. A retrospective review identified this cohort of children asymptomatic of their LSL and their progress closely recorded.

This study suggests that the natural history of this subgroup of dysraphic patients may be more benign than hitherto considered. Conservative management with adoption of a novel surveillance policy and timely intervention only in the presence of symptomatic deterioration resulted in 71% of this series remaining clinically asymptomatic at mean follow up period of 5.9 years (range, 1.0-19.3 years). At 10 years, the cumulative risk of deterioration determined by the Kaplan-Meier method was 40%. Children aged<2 years, female, with presence of a transitional type of LSL and associated syrinx were independently associated with a higher risk of deterioration [3].

2006

42 consecutive cases of lumbosacral lipoma in patients aged 2 months to 15 years who underwent untethering operation during the period from 1986 to 1997. All of them underwent preoperative and postoperative urodynamic studies (UDS). At the last follow-up (mean duration of follow-up 108 months, range 44 to 176 months), 26 cases maintained social continence with or without intermittent catheterization. Young age (< or =12 months) at operation, preoperative absence of urologic symptoms, and absence of neurologic abnormalities were significantly correlated with favorable urologic outcome.

UDS in cases with lumbosacral lipomas is a valuable tool for detecting neurourologic abnormalities as well as for monitoring the postoperative course and guiding management. History taking and neurologic examination are also proved to be important aspects in the evaluation of children with lumbosacral lipomas, predictive of urologic outcome. Better urologic results are anticipated if surgery is performed when the child is 12 months old or younger [4].

In 32 patients with LMMC (21 female and 11 male patients). The majority of patients had their primary tethered cord release (TCR) by ≤1 year of age (59 %), with 22 and 19 % having primary TCR at ages 1-15 and >15 years, respectively. Fifteen patients had at least one repeat TCR, with ten of these having more than one repeat TCR. A significant relationship was noted between low back/radicular pain and repeat TCR ($p < 0.001$). Ten patients (31%) had a limb length discrepancy of >2.5 cm, and 53 % of patients had asymmetric involvement. Nine patients (28 %) had scoliosis of whom only one required operative treatment. Fifteen patients had foot deformities. Thirteen patients (41 %) had two or more orthopaedic procedures in addition

The presenting musculoskeletal clinical signs and symptoms in patients with LMMC are uniquely different in terms of both pattern and frequency compared to myelomeningocele and other forms of spinal dysraphism.

Its a high prevalence of asymmetrical involvement, a high operative burden, and a high rate of repeat symptomatic tethered cord syndrome requiring TCR. As previously noted by others, TCR in LMMC does not prevent long-term functional deterioration. These findings may be important to our colleagues providing counsel to their patients with LMMC and to their families [5)].

1)
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