Asymptomatic lipomas of the conus medullaris


Lipoma of the conus medullaris is one of the most commonly described forms of occult spinal dysraphism and represents a considerable challenge in clinical decision making.6,9 There is an ongoing unresolved debate in the neurosurgical community about its optimal management: Should children diagnosed with this condition be offered prophylactic surgery? We have recently read with great interest the publication by Talamonti and colleagues.7 This potentially meaningful clinical study, apparently a prospective cohort study (although not explicitly stated), compares the outcome of surgical and conservative management of patients with an asymptomatic conus lipoma.

The authors should be commended for their thorough investigative work, methodological stringency, and elaborate statistical analysis. The children in the surgical and conservative groups had similar baseline demographic data, except for the remarkably younger age in the surgical group (mean 1.9 years vs 3.1 years in the conservative group). This disparity may reflect some selection bias of which the authors were aware and accounted for in the statistical analysis. The clinical characteristics and MRI types of the conus lipoma (according to Chapman’s classification)1 were also fairly comparable between the 2 groups. Still, some points of the paper are worthy of further consideration.

There is no clear indication as to why asymptomatic patients were brought to medical attention, leaving the reader to assume that it was because of suspicious cutaneous stigmata. The results are very impressive in the surgical group, with a 90% favorable outcome (except for the 3 patients with repeated urinary tract infections) and a cumulative 3.7% operative morbidity, consisting predominately of sphincter and motor deficits. This risk profile is remarkably lower than that reported in most of the commonly cited clinical papers, including the oft-cited large experience of the Paris group5–7 that favored conservative management. The surgical morbidity rate is only comparable to the work of Pang and colleagues,3,4 who structurally described a detailed surgical approach for maximum safe resection of the lipoma with extensive intraoperative monitoring. Although we expected that a similar detailed description of the operative technique would be included in the paper by Talamonti et al. in order to provide some insight on the factors that favored this outstanding outcome, the authors stated that radical resection was never attempted and did not report the operative data on the extent of lipoma resection, whether complete untethering was identified either directly or with surrogates (e.g., getting the edge of the lipoma, restoration of CSF flow inside the thecal sac, or visualization of freely flowing conus). With that being said, the group relied on postoperative MRI to provide data (cord/sac ratio and extent of resection) as surrogate indicators of adequate untethering. These measures are indeed very useful, but they can only serve for postoperative follow-up purposes and do not help with intraoperative guidance. Moreover, the follow-up was devoid of explicitly defined structured end points (although a protocol for follow-up was referred to) for the recognition of secondary tethered cord syndrome.

The study presents a patient cohort with clinical and radiological characteristics that are strikingly similar to those of the cohort of the UK group8 who offered a natural history perspective and a management algorithm for this condition. But the article by Talamonti et al. supersedes the British article with its prospective nature and its comparative data between the surgical and conservative groups, and in some of its operative technical details (e.g., filling the thecal sac with hyaluronic gel) and postoperative imaging data (favorable cord sac ratio < 0.5).

The authors have contributed significantly to the growing body of literature and reinforced certain notions. Among these is the complex nature and grave prognosis of the transitional type of conus lipomas as well as the expected age of deterioration (around 7–8 years). Of special note in this respect is that the study failed to show a statistically significant association between the risk of deterioration and type of lipoma, despite the fact that 5 of the 7 cases in which deterioration was observed in the conservative group and all of the complicated cases in the surgical group involved transitional lesions. This has many explanations; the simplest is that the small number
of cases underpowers the study’s ability to detect a difference. Given these results, the authors’ conclusion and subsequent change of management protocol to offer surgery to all patients with nontransitional lipomas remains without explanation.

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DISCLOSURE
Dr. Souweidane reports a consultant relationship with Aesculap.

References

Response
We sincerely thank Drs. Latif and Souweidane for their kind words about our article. As they correctly outlined, the only remarkable difference between the surgical and the conservative groups was the mean age (1.9 vs. 3.1 years). Our policy was to offer the choice between surgical and conservative management as soon as we encountered these patients. Accordingly, there was no voluntary bias. The older age in the conservative group was simply related to delayed observation of these patients. The situation of a newborn with a recently diagnosed lipoma is likely to be quite different from that of an older child with an asymptomatic lipoma. In the case of the newborn, the parents were usually more worried about the malformation than about the treatment. Most likely, they had not yet accepted the idea of a potentially dangerous malformation. Therefore, despite reassurance and information, they were probably more inclined to choose surgery, hoping this could be immediately and definitely decisive. Conversely, when we first met with parents of older, asymptomatic children, the parents had already personally verified that their child had remained normal despite the malformation. Furthermore, often they had previously met with other neurosurgeons with different opinions about treatment. Accordingly, they were probably more worried about the surgical risks than about the natural history, which had not seemed so negative up to that point.

Dr. Latif and Dr. Souweidane wondered about the cutaneous stigmata of our patients. In our experience, it is quite difficult to find a truly occult lipoma in a completely asymptomatic patient. This may occur only incidentally. In fact, in this series, there was no truly occult malformation, and all patients had cutaneous stigmata, which resulted in their being brought to medical attention when they were still asymptomatic: most patients presented true subcutaneous masses, others had gluteus asymmetry, and so on. However, all of these patients were asymptomatic when we met them for the first time.

Drs. Latif and Souweidane commented that our rate of good surgical results (90%) is comparable only with the rate of favorable outcome reported by Pang et al.10,11 We do not agree with this statement. As they correctly emphasized, worse results have been reported by the Parisian authors5,12,15,16 and also by others,1 but the incidence of spinal cord retethering following lipoma resection has been repeatedly found to be between 10% and 20%.2,3,4,11 Apart from the cited papers by Pang et al.,10,11 there are also papers by other authors reporting results quite comparable with ours. For instance, Wu et al.14 demonstrated that 84% of their 43 patients maintained stable urodynamical and neurological functions after surgery. The same success rate was obtained by Morimoto et al.15 in 56 patients with a mean follow-up of 7.9 years. Kaney et al.4 reported that 92.1% of their 38 asymptomatic patients had no neurological deficits or bladder dysfunction at long-term postoperative follow-up (range 1–21 years, mean 6.2 years). Finally, in the wide experience of McLone and colleagues,6,7 71 asymptomatic children with lipomas of the medullary conus underwent prophylactic surgery; 5 of these patients experienced inexorable deterioration, while 66 (93%) remained asymptomatic throughout the follow-up period (mean 6.2 years).

We did not detail our surgical techniques only because we did not use a particular personal technique. In the article, we clearly declared that we adopted the classical techniques, namely those carefully described by masters such as McLone, Oakes, and especially Pang7,9,10. However, we introduced some small personal modifications (indicated in the text): for instance, the use of hyaluronic gel or lipoma removal following the instructions of Pang et al.10 but without attempting a truly radical removal. Pang and colleagues are so skilled that they were able to completely
remove lipomas with impunity. Instead, in our series, all cases of both transitory and permanent morbidity occurred in patients treated with more than 90% removal, while problems never occurred in patients with less extended removal. Of course, we always try to remove as much of the lipoma as possible, but when we are operating close to the neural plaque and the ventral roots, we balance the risk of morbidity against the potential benefit of further resection. If the local anatomy is not completely clear and/or the intraoperative monitoring is alarming, but we have already gained enough space for adequate neurulation and sac reconstruction, we prefer to settle for the removal already obtained, rather than to run useless risks. The lesson we learned is that it is more important to fashion a good neurulation and a wide dural sac (thus obtaining an adequate cord/sac ratio), than to insist on pursuing a radical resection that may be risky and perhaps not so useful.

Drs. Latif and Souweidane also reported that we did not detail the extent of lipoma resection. They may have missed the report of postoperative results on the fourth page of our article. In spite of our aforementioned prudent attitude, in 19 (59.4%) of our patients, we achieved subtotal resection (more than 90%, but never 100%; examples of such cases are shown in Figs. 1, 3, and 5 of our article); in 12 patients (37.5%), we performed partial removal (less than 90%, but more than 70%; as shown in Figs. 2 and 4), and in 1 patient (3%), limited removal (about 60%) was performed. In Table 1 of the article, we compared the cases with more than 90% removal (called “extended”) versus cases with less than 90% removal (called “not extended”) and found no statistically significant effect on the risk of delayed tethering ($p = 0.098$). Conversely, the patients with a cord/sac ratio of less than 50% had significantly less probability of developing delayed tethering ($p = 0.017$).

Dr. Latif and Dr. Souweidane criticized our providing only postoperative (MRI) data to assess the completeness of detethering and nothing helping with intraoperative guidance. Probably, we could say that the detethering is adequate only when an adequate neurulation can be obtained. In general, we are satisfied about the detethering when the terminal end of the neural plaque is completely detached from the surrounding fibroadipose tissue, when a good neural tube can be reconstructed with a smooth and rounded tubular shape, and when the exposed, sticky, cut surface of the fatty tissue is concealed within the neural tube. In the series we reported in our paper, we were quite satisfied in all cases but one. The exception involved a patient with a chaotic lipoma, the only one who underwent 60% lipoma removal; in spite of the limited removal, the detachment appeared intraoperatively complete even in this case, and the neurulation could be fashioned anyway, but the local anatomy was so distorted that we did not feel entirely satisfied. Of course, sometimes short neural roots may play a role in the position of the conus. Pang et al. reported that the ascension of the detached neural tube is sometimes possible, but we do not accept this as evidence that the detethering is adequate, since we are not addressing lipomas of the filum terminale. We consider the detethering adequate when the reconstructed neural tube has no more attachments and is relatively “free-floating” inside the expanded dural sac.

Finally, Dr. Latif and Dr. Souweidane expressed concern about our change of management protocol to offer surgery to all patients with nontransitional lipomas. We admit that their perplexity is quite understandable. Indeed, we clearly specified that our series was small and that our results required confirmation with larger numbers and longer follow-up periods. However, the only cases of nontransitional lipomas with unsatisfactory outcomes in our series were those that were conservatively managed; the patients with nontransitional lipomas who underwent surgery experienced neither surgical morbidity nor delayed tethering. Furthermore, in the surgical group, delayed tethering occurred only in patients with transitional lipomas, and we must also take into account that the nontransitional lipomas are well known to be easier to treat surgically. We do realize that our numbers were small. However, even though our results did not reach statistical significance, we do not believe that we should continue to propose conservative management to patients with relatively simple lesions. Undoubtedly, statistical demonstration remains the only reliable scientific method to show whether something is correct, but one can at times accept that something is useful even without a study showing statistical significance: No double-blind randomized study has been conducted on the use of a parachute, but nobody would recommend jumping out of a plane without one.

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References


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