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Paraganglioma of the cauda equina

Sir: Paragangliomas of the cauda equina region are indeed rare neoplasms, but are not as uncommon as Cole et al suggest. In addition to the 20 cases described and quoted in their recent letter,1 at least 44 others2–8 have been reported including four in the United Kingdom. In these 44 cases, the mean age at diagnosis was 49 years (age range 23–71 years) with a sex distribution of 25 males and 19 females, confirming the male predominance suggested by Cole et al, but to a far lesser extent. In addition to the clinical features reported recently,1,9 one case2 has presented with endocrine activity in the form of flushing attacks accompanied by increased urinary levels of noradrenaline. Several cases have been accompanied by spinal blockage with an unusually high level of protein in the CSF10–11 and it has been suggested that this finding may be useful in the differential diagnosis of intradural neoplasms in the cauda equina.11

These neoplasms resemble paragangliomas occurring in other sites in both their histological appearances and biological behaviour;12 occasional similarities to ependymomas can be resolved by electron microscopy and special staining techniques, some of which have been described by Anderson and Gullan.9 Positive staining for gamma (“neuron-specific”) enolase has been the most consistently observed immunocytochemical finding to date,1,6,9 but significant differences in immunoreactivity for glial fibrillary acidic protein and neurofilament protein have been reported.3,6,9 These probably relate to differences in the immunocytochemical techniques employed by the various authors. Recent experience in this laboratory suggests that immunoreactivity for PGP 9.512 is consistently present in these neoplasms (as in paragangliomas in other sites11), but not in ependymomas. Positive immunostaining for cytokeratin,3 somatostatin, and serotonin6 may also be useful in the differentiation from spinal ependymomas, but electron microscopy remains the most reliable investigation, even on formalin-fixed tissue, when the characteristic neurosecretory granules and fibrous bodies are diagnostic.3,6,9

In the largest published series,6 the authors recommended that paragangliomas in the cauda equina should not be biopsied, but treated by total resection whenever possible. Post-operative radiotherapy is recommended in cases where only incomplete resection is possible; combined modality treatment in such cases was generally successful, but tumour recurrence was recorded in one case.6 As with paragangliomas in other sites,12 it is not possible to predict the likely behaviour of these unusual neoplasms on histological grounds alone.

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References

Dermatomal somatosensory evoked potentials in lumbosacral root compression

Sir: Katifi and Sedgwick1 recently reported their experiences with dermatomal somatosensory evoked potentials (DSEP) in the evaluation of patients with lumbosacral root compression. Their conclusions regarding the clinical utility of DSEPs directly conflict with ours23 despite normative data virtually identical to our own previously-published values.24

Part of the difference between us may relate to the population of patients studied. We specifically excluded patients with evidence of bilateral disease or disease outside the L5 or S1 territory in order to simplify the comparison of clinical findings with neurophysiological data. We also confirmed the diagnosis by myelography, metrizamide CT scanning, needle electromyography, or operation, and two-thirds of our patients had unilateral involvement confined to either the L5 or S1 root.2 In contrast, a third of the patients reported by Katifi and Sedgwick had bilateral disease either clinically or at operation, and 80% had evidence of multilevel disease between L2 and S4. Thus, their patients had more widespread disease, which complicates interpretation.

Another difference between us was in the criteria used to define abnormality. We used three standard deviations from our normal latency values to define abnormality, whereas they used two. We both used the same amplitude criteria. In our study we had two criteria of abnormality (latency and amplitude) for each of two segments (L5 and S1) for each side, and one additional criterion of abnormality (interside latency difference) for each of the two segments (L5 and S1), giving a total of 10 comparisons per patient. Even if we were 100% confident that these two standard deviations encompassed 95% of the population of interest, and even though some of these comparisons are not completely independent of others, the chance of finding some “abnormality” in a patient without root compression (that is, the false-positive rate) would be
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unacceptably high if we had used two standard deviations for even a few of the 10 comparisons. In the study of Katifi and Sedgwick, four criteria of abnormality (amplitude, latency, inappropriate latency compared to the segment above and below, and interside latency difference), five segments (L2, L3, L4, L5, and S1) and two sides were used for a total of 35 comparisons per patient. In this setting, a limit of two standard deviations is even less acceptable than it would have been in ours.

Katifi and Sedgwick claim a high sensitivity for DSEPs in the evaluation of lumbar sacral root compression, suggesting that the technique “predicted root compression in 19 of 20 cases”. Less stringent criteria for abnormality always leads to an increased sensitivity, but only at the expense of reduced specificity. A careful reading of their table 1, fact, demonstrates this pitfall and the misleading conclusions which follow from it. They were able to detect correctly 74% (25/34) of the roots involved at surgery, but incorrectly identified 12 additional roots that were unaffected when explored. Moreover, in three instances (cases 2, 6 and 19) DSEPs only identified roots contralateral to the clinical signs and in one (case 2) contralateral to all of the symptoms as well. In another (case 10), the DSEP abnormality was contralateral to the surgically involved root. More troublesome from a clinical perspective, however, is the fact that in only four of the 21 patients (cases 1, 14, 18 and 20) did the DSEP accurately and completely predict the operative findings.

Some of these discrepancies could relate to surgically apparent disease or subclinical compression as a result of severe and widespread pathology of the lumbar sacral area. If such is the case, however, then patients with more restricted disease need to be studied (as we did) before the clinical utility of a new technique such as the recording of DSEPs can be accurately defined.

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References


Katifi and Sedgwick reply

We are grateful to Aminoff and Goodin for their interest and critique of our paper.1 Careful consideration of the points raised is given below but in the end we stand by our conclusion that the DSEP “...is a very accurate method of detecting abnormalities in root function”.

1. Normal Data

Our normal regression lines2 of latency on height and those of Aminoff and Goodin3 4 are plotted together in the fig. The regression lines of mean are almost parallel with ours but about 2 ms lower which seems a good correspondence between the two laboratories. The crucial difference lies in the standard error of estimate (SEE) which are 4.7 and 3.8 ms for L5 and S1 dermatomes respectively in the Aminoff and Goodin data3 4 but 2.90 and 2.95 in ours2 (table 3). This difference is further compounded by their choice of +3 SEE as the upper limit of normal which gives a value some 10 ms higher than ours; even taking +2 SEE gives a figure 5 ms higher. One reason contributing to a higher SEE may be the smaller sample size of Aminoff and Goodin, 32 compared with 54 subjects in our series.2

2. Definition of abnormality

Selection of +2 or +3 SEE as the upper limit of normality depends on two related principles. First, the nature of the abnormality in the diseased population; do the latencies of patients with unequivocal root compression lie far to the right of the tail of the normal distribution or do they overlap? They overlap, most delays lie between 2 and 3 SEE of normal. The figure shows our 20 abnormal delays plotted. Second, how dangerous or misleading is a false positive result? Only experience allows one to balance carefully these factors. We opted to use +2 SEE which includes 95% of the population in question, the chances of wrongly interpreting a normal result is only 2.5%. It is, after all, just as important not to have many false negatives as a normal result in a sensitive test is valuable in excluding root involvement. These criteria are safe as long as the referring doctors understand these implications.3

We routinely studied L5 and S1 and the other dermatomes only where clinically indicated, not all five roots as suggested. Interside differences of above 6-3 and 5-1 ms for L5 and S1 respectively were taken as abnormal by Aminoff and Goodin; our upper limits were 3-5 and 4-5 ms which represented 2SD.
Dermatomal somatosensory evoked potentials in lumbosacral root compression.
M J Aminoff and D S Goodin

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Updated information and services can be found at:
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