Matters arising

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*¹ 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*,¹ but to a far lesser extent. In addition to the clinical features reported recently,¹⁹ one case² has presented with endocrine activity in the form of flushing attacks accompanied by increased urinary levels of nor-epinephrine. 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 ependyomas 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,^{13,6} 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 9.5¹³ is consistently present in these neoplasms (as in paragangliomas in other sites¹⁴), but not in ependyomas. Positive immunostaining for cytokeratins,³ somatostatin, and serotonin⁶ may also be useful in the differentiation from spinal ependyomas, 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 pos-

sible. 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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Dermatome somatosensory evoked potentials in lumbosacral root compression

Sir: Katifi and Sedgwick¹ recently reported their experiences with dermatome somatosensory evoked potentials (DSEP) in the evaluation of patients with lumbosacral root compression. Their conclusions regarding the clinical utility of DSEPs directly conflict with ours^{2,3} despite normative data virtually identical to our own previously-published values.^{2,4}

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 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 95%.