Matters arising

sode of meningis with tetraplegia that was attributed to rebleeding. He developed hydrocephalus and died with a nosocomial infection. The anatomical diagnosis was hemangioblastoma.

Spinal intramedullary haemangioblastoma and similar tumours present diagnosis problems clinically, radiologically and pathologically. The spinal haemangioblastomas are certainly a rare cause of spontaneous subarachnoid haemorrhage and their classical presentation is a progressive syndrome of spinal cord compression or a syringomyelic syndrome. However, a sudden clinical episode compatible with arteriovenous malformation does not exclude a haemangioblastoma and a patient with subarachnoid haemorrhage and a normal cranial CT should be investigated by myelography, spinal CT and arteriography in order to exclude a vascular tumour in the spinal canal.

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References


Computed tomographic findings of brain and skull in myotonic dystrophy

Sir: With great interest we read the article by Avrahami et al about computed tomographic findings of brain and skull in myotonic dystrophy. However, I do not agree with their opinion that bones in the base of the skull and others of the body seem not to be involved. We have previously reported two myotonic dystrophy patients with ossification of the posterior longitudinal ligament causing transverse myelopathy, in that one patient without calvarian hyperostosis but another with not only calvarian hyperostosis but also abnormal ossification of the clivus, which was confirmed by necropsy. To our knowledge, ossification of the posterior longitudinal ligament has not been recognised in patients with myotonic dystrophy outside of Japan, of which there have been at least eight reports. Ossification of the posterior longitudinal ligament is a common condition in Japan, frequently incidentally identified, but sometimes causing myelopathy. Although the precise cause of ossification of the posterior longitudinal ligament remains unknown, it is suspected that it may be a part of manifestations of the generalised hyperostotic potential in patients, probably genetically transmitted, because of high incidence of association with ossification of the other ligaments of the spine and concurrence in the same families on national surveys of ossification of the posterior longitudinal ligament.

Avrahami et al hypothesise that the cause of hyperostosis of the calvarium is secondary to microcephaly because the base of the skull is not involved. However, in our two patients with ossification of the posterior longitudinal ligament, one had even hyperostosis of a part of the base of the skull. Jequier has suggested that cranial hyperostosis might have been genetically determined in patients with myotonic dystrophy. In Japan, there has been a case of concurrence of myotonic dystrophy and ossification of ligaments of the spine in two siblings and additionally calvarian hyperostosis in one reported. Therefore, it is suspected that association with ossification of the posterior longitudinal ligament and hyperostosis of the clivus in myotonic dystrophy may be more than fortuitous. Further investigations are certainly needed to clarify whether patients with myotonic dystrophy have generalised hyperostotic potential.

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References


Korczyn and Avrahami reply:

We are grateful to Dr Kawamura for drawing our attention to his work, demonstrating ossification of the posterior longitudinal ligament in some Japanese patients with myotonic dystrophy. As Dr Kawamura notes, this phenomenon is common among Japanese, and the relationship to myotonic dystrophy in the cases described by him may therefore be fortuitous. Otherwise, this heterotopic calcium deposit is more likely to be related to basal ganglia calcification, described by us than to the thickening of the calvarian bones. Clearly, however, further studies on calcium and bone metabolism in myotonic dystrophy are needed.

Notice

The World Federation of Societies of Biological Psychiatry

A Regional Congress will be held 2-7 April 1989 in Jerusalem. Information may be obtained from Professor RH Belmaker, Chairman, POB 983, Jerusalem 91009, Israel.