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CASE REPORT

Magnetic Resonance Imaging Evaluation of Paraganglioma in the Cauda Equina

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ABSTRACT

Paragangliomas of the cauda equina, previously thought to be a rare disease affecting the spine, are being increasingly reported in the literature. This report is of a patient with paraganglioma arising in the cauda equina, and describes the magnetic resonance imaging features, with emphasis on the differential diagnosis of this tumour.

Key Words: *Cauda equina, Magnetic resonance imaging, Paraganglioma*

INTRODUCTION

Paragangliomas are collections of neural crest–derived, neurosecretory, and chemoreceptor cells of the autonomic nervous system, belonging to the amine precursor uptake decarboxylase (APUD) system.[1] Approximately 85% to 90% of paragangliomas arise in the adrenal gland.[2] Tumours of the carotid body and glomus jugulare constitute more than 90% of reported extra-adrenal paragangliomas.[1,3] Spinal paragangliomas are rare, and the majority develop in the lumbosacral region, especially the filum terminale.[1,4,5] These are most often intradural, extramedullary, encapsulated lesions.[3] Other potential sites in the central nervous system include the pineal or pituitary glands, the cerebellopontine angle, and the cauda equina.[1,5]

CASE REPORT

A 28-year-old woman presented with right sciatica and low back pain of 9 months duration. Physical examination showed features of cauda equina syndrome. Magnetic resonance imaging (MRI) revealed a well-demarcated, intradural, extramedullary tumour at the L3 to L4 level. The lesion was isointense to conus medullaris on T1-weighted images (Figure 1) and slightly hyperintense on T2-weighted images (Figure 2). In addition, there were serpiginous signal void structures suggestive of high velocity flow vessels around the tumour (Figure 2). Postcontrast scans showed marked but slightly inhomogeneous enhancement of the mass and the surrounding vessels (Figure 3). The patient subsequently underwent surgery with complete excision of the tumour.

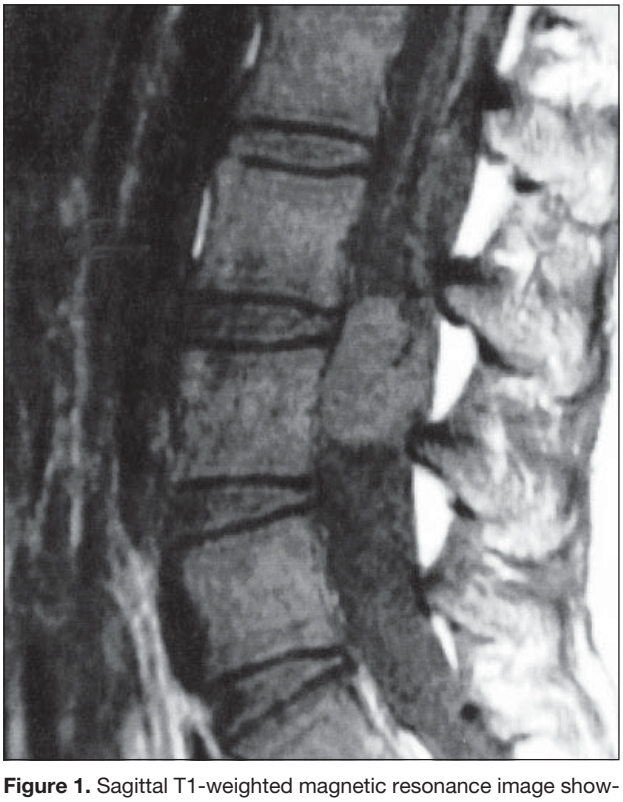

Figure 1. Sagittal T1-weighted magnetic resonance image showing a well demarcated intradural extramedullary mass isointense to conus medullaris at the L3 to L4 level.

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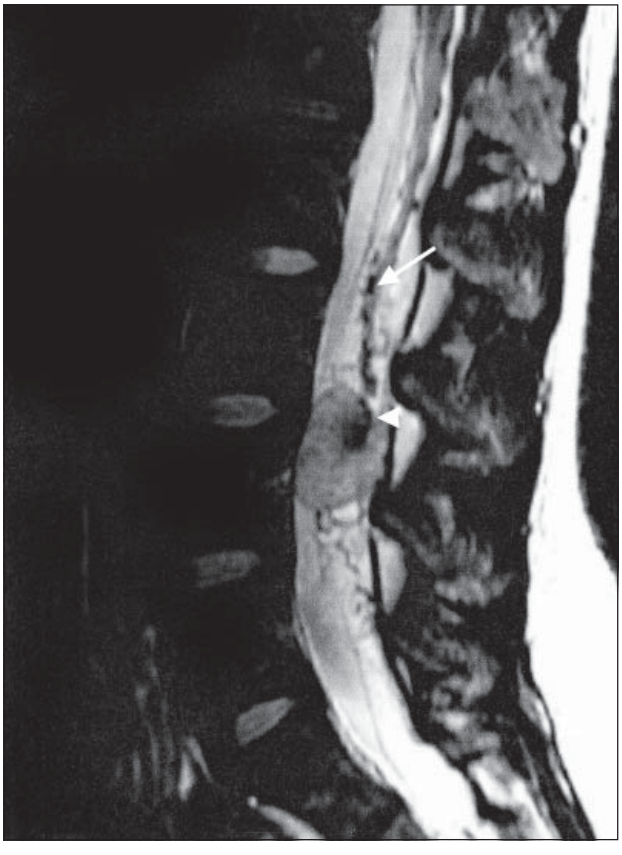

Figure 2. Sagittal T2-weighted magnetic resonance image demonstrating a slightly hyperintense tumour (arrowhead) with serpiginous signal void structures suggestive of high velocity flow vessels around the tumour (arrow).

Figure 3. Sagittal T1-weighted magnetic resonance image after intravenous gadolinium injection, showing marked but slightly inhomogeneous en-hancement of the mass (arrowhead) and the surrounding vessels (arrow).

Histological examination of the tumour showed typical findings of paraganglioma, with tumour cells arranged in nests and lobules (zellballen), admixed with a small number of spindle cells with dendritic processes. The zellballen were surrounded by a delicate capillary network. Immunohistochemistry staining of tumour cells showed positivity for chromogranin, synaptophysin, neurone specific enolase, and CD56, while scattered dendritic cells showed positivity for S100.

The patient made an uneventful recovery. A follow-up MRI scan a year later showed postoperative changes, with no evidence of tumour recurrence.

DISCUSSION

The first case of paraganglioma of the cauda equina was reported in 1970.[1,3,6] Since then, more than 100 cases have been reported.[7] Nevertheless, paraganglioma is still an easily missed differential diagnosis for tumours of the cauda equina. The mean age of occurrence is approximately 50 years,[1,3,5,6] with a male predominance.[1,3,5,6,8] The most common presenting symptoms are low back pain and sciatica, as noted with this patient. Occasional findings of motor or sensory deficits in the lower extremities, or urinary and faecal incontinence have been reported.[1,3,5,7,8] The cerebrospinal fluid (CSF) protein level is usually markedly raised in these patients.[3,7,8]

Differential diagnoses for a tumour in the cauda equina region include ependymoma, neurinoma, meningioma, haemangioblastoma, solitary drop metastasis, lipoma, and dermoid tumours.[1,3,5-9] Previous preoperative evaluation by plain radiography and myelography has been surpassed by MRI. The latter is excellent for defining the extent of the lesion and its relationship to the nerve roots. The tumour is usually hypo- or isointense to the conus medullaris on T1-weighted images, and hyperintense on T2-weighted images. It can occasionally be inhomogeneous and sometimes shows cystic areas.[1] Multiple punctate and serpiginous structures of signal void, demonstrating heterogeneous and intense enhancement, may be detected around and within the tumours on all pulse sequences.[1,5,10]

MRI features of paraganglioma were previously considered non-specific, but with more reported cases,

2 signs have been recognised that assist diagnosis. The first is the detection of serpentine vessels around the lesion.[1,8] This is well demonstrated in the current patient. The second is the complete encapsulation of the tumour by a margin that is hypointense on T2-weighted, proton-weighted, and gradient echo imaging.[1,8] This latter sign occurs in approximately 75% of patients[5] and probably indicates haemosiderin or ferritin from previous haemorrhage.[1,8] Ependymoma, neurinoma, meningioma, and solitary metastasis seldom demonstrate these signs. Serpentine vessels can also be seen around haemangioblastomas but the encapsulation sign has not been reported. Differentiation of lipoma and dermoid tumours is relatively easy, as their fat content can be identified as hyperintensity on T1-weighted images.

The ultrastructural and immunohistochemical appearance of paragangliomas are unique.[1,6,8,9] The zellballen pattern of cells, surrounded by a capillary network is typical. Ultrastructurally, the granules in the cytoplasm of these cells are shown to contain dopamine, adrenaline, noradrenaline, or 5-hydroxytryptamine (5-HT).[1,6] Despite this fact, most of the tumours are endocrinologically non-functional.[1,3,6]

Surgical resection is the mainstay of treatment. Preoperative embolisation of feeding vessels can be undertaken.[1] Adjuvant radiotherapy is given for non-encapsulated lesions or in the case of incomplete resection.[1] Patient outcome is usually excellent. Only a few patients with extradural extension have been reported,[3,5,6] and only rarely has aggressive behaviour, with CSF metastases, been observed.[4,11] Recurrence is unlikely when surgical excision is complete, but can be late and has been reported to occur a decade after initial surgery.[3,8]

CONCLUSION

Paraganglioma of the cauda equina is an uncommon tumour but should be considered if associated serpentine vessels are demonstrated on preoperative evaluation. MRI is the modality of choice in the detection and diagnosis of this lesion.

REFERENCES

1. Aghakhani N, George B, Parker F. Paraganglioma of the cauda equina region — report of two cases and review of the literature. Acta Neurochir 1999;141:81-87.
2. Burton SS, Ros PR. Adrenal glands. Stark D, William G, Bradley Jr, editors. Magnetic resonance imaging. St Louis: Mosby; 1999: 503-516.
3. Sonneland PRL, Scheithauer BW, Lechago J, Crawford BG, Onofrio BM. Paraganglioma of the cauda equina region — clinicopathological study of 31 cases with special reference to immunocytology and ultrastructure. Cancer 1986;58: 1720-1735.
4. Sundgren P, Annertz M, Englund E, Stromblad LG, Holtas S. Paragangliomas of the spinal canal. Neuroradiology 1999;41: 788-794.
5. Levy RA. Paraganglioma of the filum terminale: MR findings. AJR Am J Roentgenol 1993;160:851-852.
6. Mylonas C. Case report: paraganglioma of the cauda equina. Clin Radiol 1992;46:139-141.
7. Haslbeck KM, Eberhardt KEW, Nissen U, et al. Intracranial hypertension as a clinical manifestation of cauda equina paraganglioma. Neurology 1999;52:1297-1298.
8. Boncoeur-Martel MP, Lesort A, Moreau JJ, et al. MRI of paraganglioma of the filum terminale. J Comput Assist Tomogr 1996; 20:162-165.
9. Wester DJ, Falcone S, Green BA, Camp A, Quencer RM. Paraganglioma of the filum: MR appearance. J Comput Assist Tomogr 1993;17:967-969.
10. Shin JY, Lee SM, Hwang MY,Sohn CH, Suh SJ. MR findings of the spinal paraganglioma: report of three cases. J Korean Med Sci 2001;16:522-526.
11. Roche PH, Figarella-Branger D, Regis J, Peragut JC. Cauda equina paraganglioma with subsequent intracranial and intraspinal metastases. Acta Neurochir 1996;138: 475-479.