PARAGANGLIOMA OF THE CAUDA EQUINA: CASE REPORT AND REVIEW OF THE MRI FEATURES

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We report a case of paraganglioma of the cauda equina and review the MRI features in 11 previously published cases with MRI description. On Tl-weighted images, paraganglioma is isointense relative to spinal cord and shows moderate to intense enhancement after Gd-DTPA application, usually with heterogeneous texture. On T2-weighted images, different findings were found. MRI features do not allow to distinguish paraganglioma of the cauda equina from majority of other tumors in this region.

INTRODUCTION

Paraganglioma is a tumor arising from the neuroectodermally derived cells[1,2]. These cells have neural features of biogenic amine- and peptide-containing secretory granules and potential endocrine function[3,4]. Tumors arising from these cells have distinctive names, but together are also called neuroendocrine tumors (NETs)[3]. Pheochromocytoma of the adrenal medulla is the most typical NET[2,4–7]. Most common extra-adrenal NETs are carotid body and glomus jugulare tumors[4]. Numerous other locations of NETs have been described including retroperitoneum, mediastinum, gastrointestinal and respiratory tracts, among others[2–4,6,7]. The first published, although unrecognized case of paraganglioma of the cauda equina region was reported in 1970[8] and since then more than 80 cases have been published. In recent cases also magnetic resonance imaging (MRI) findings were described[1,3,5–7,9–14]. Recognition of this tumor is important because of its biologic characteristics, which differ significantly from ependymoma, the tumor with which it is most commonly confused[9]. We report a new case and review the MRI features of the cases described in the literature.

CASE REPORT

A 46-year-old man presented with a three years history of low back pain and bilateral sciatica, more prominent on the left side. He denied any bladder or bowel symptoms. On the physical examination the patient was found to have bilateral straight-leg raising limited to 45° and Achilles tendons hyporeflexibility. The neurologic examination was otherwise unremarkable. Plain film and two unenhanced computed tomographic (CT) examinations of the lumbosacral spine performed during the last two years were reported as normal.

At the last presentation to University Hospital of Cattinara (Trieste, Italy), lumbar myelography was performed which showed an intradural irregular filling defect at the L2-L4 level and widening of the spinal canal below the L4 level. A consecutive CT myelography confirmed voluminous soft tissue mass occupying spinal canal (Fig. 1). MRI study, using a Gyroscan ACS II (Philips, Netherlands) 1.5 T unit, showed an intradural lesion below conus. On unenhanced Tl-weighted spin-echo (SE) images, the lesion's signal intensity was approximately equal to that of the conus (Fig. 2a). On turbo SE proton-density-weighted images, the signal of the lesion was only minimally more intense than that of both cerebrospinal fluid (CSF) and conus, and it was difficult to delineate the tumor borders. T2-weighted turbo SE images demonstrated the lesion well, its signal was heterogeneous, less intense than CSF and more intense relative to conus (Fig. 2b). Tl-weighted SE images after intravenous injection of Gd-DTPA (0.1 mmol/kg of Magnevist, Schering, Germany) showed moderate enhancement of the tumor with some inhomogeneities caused by a vascular or nervous structure. A flow void from prominent tortuous vessels above the tumor were seen in all sequences.

Right side hemilaminectomy from L2 to L4 was performed. After opening the dura, highly vascularized lesion compressing nerve roots was found. One nerve root was involved and infiltrated by the lesion and had to be cut to facilitate complete excision. One feeding artery entered the tumor along the course of involved nerve. Resection of the whole tumor was performed. Microscopic examination and immunohistochemical staining revealed features of a paraganglioma. Postoperative radiation therapy was given. A follow-up MRI 6 months after surgery has shown adequate postoperative findings with no residual mass and disappearance of prominent vessels (Fig. 3).

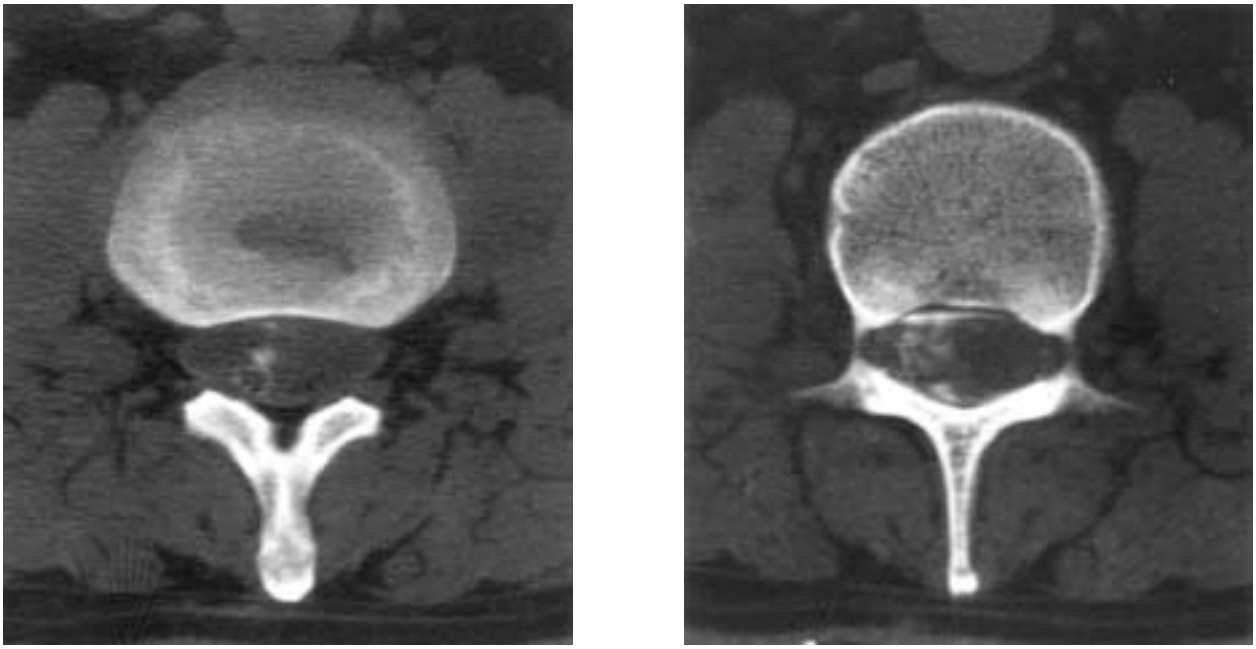

Fig. 1. a), b) CT-myelography scans obtained at L2–L3 (**a**) and L3 (**b**) levels. Soft tissue mass is completely filling spinal canal and allows only minimal amout of contrast material to pass through the obstruction.

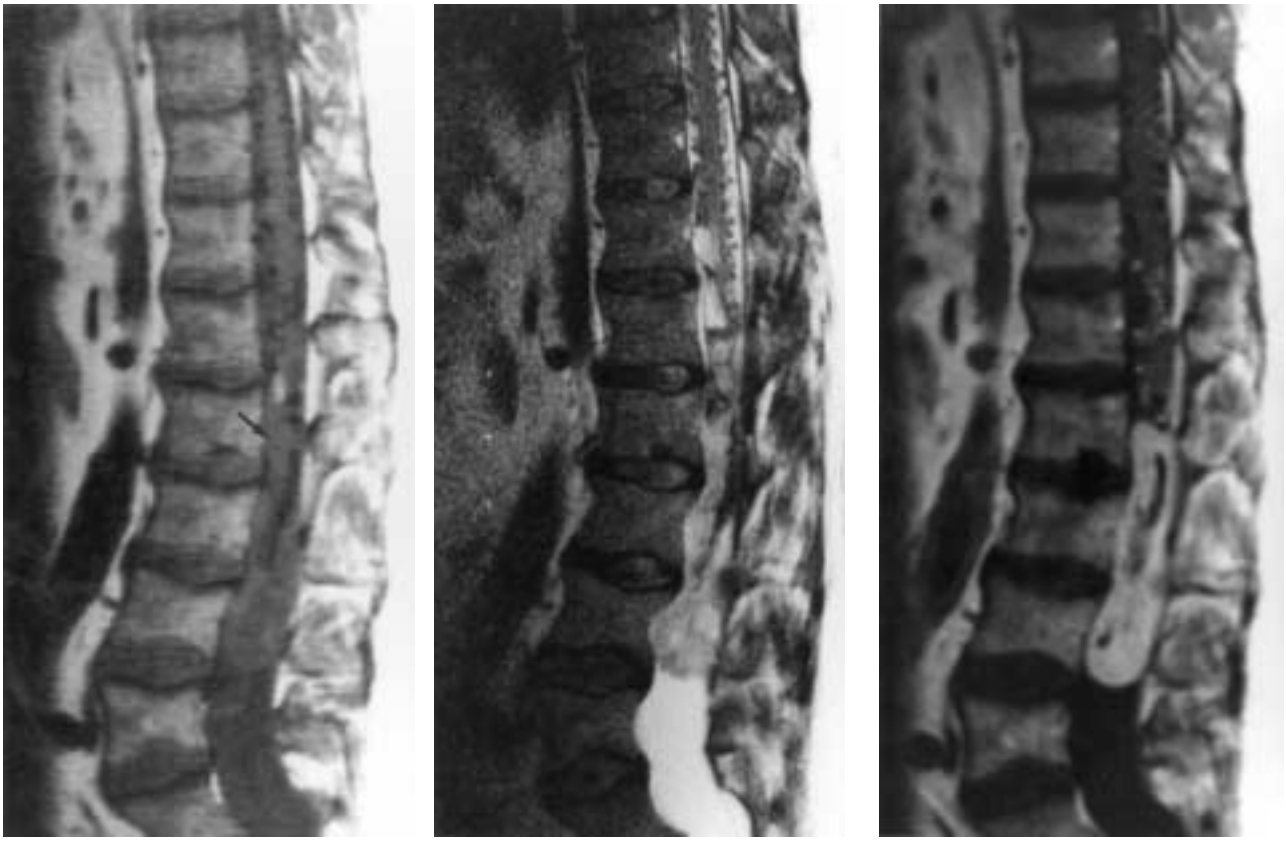

Fig. 2. **a)** Nonenhanced sagittal Tl-weighted spin-echo (SE) image (450/19/2, TR/TE/excitations) reveals a large intradural tumor at the L2-L4 level (arrows) that is almost isointense to the spinal cord and slightly heterogeneous.
b) T2-weighted turbo SE image (2000/150/6) shows tumor signal to be higher relative to spinal cord. Heterogeneity of the lesion is more pronounced.
c) Tl-weighted SE image (450/19/2) after intravenous administration of Gd-DTPA (0.1 mmol/kg) shows well-defined, enhancing lesion. A linear hypointense structure is seen within the mass. At surgery a nerve root inside the tumor was found together with a feeding artery. Prominent epidural vessels above the tumor and ectatic dural sac below obstruction are seen in all sequences.

Fig. 3. Sagittal Tl-weighted SE image (359/19/2) after intravenous administration of Gd-DTPA (0.1 mmol/kg) obtained 6 months after surgery demonstrates no residual mass and disappearance of prominent vessels (compare to Fig. 2). Higher signal from vertebral bodies T12-L5 and sacrum is due to postoperative radiotherapy.

DISCUSSION

We found 11 reports describing MRI features of the paraganglioma of the cauda equina[1,3,5–7,9–14]. They are presented (including our case) in Table 1. When summarize: on T1-weighted unenhanced images, paraganglioma appears isointense relative to the spinal cord, usually with homogeneous texture. After Gd-DTPA application it shows moderate to intense enhancement which is heterogeneous or less commonly homogeneous (two out of 11 cases; smaller tumor in both cases). Various features are found on T2-weighted images (with different types of sequences used). Most commonly the tumor is heterogeneous, iso- or hypointense relative to CSF; iso- or hypointense relative to spinal cord.

Various tumors must be included in the differential diagnosis of intradural mass of the cauda equina region including ependymoma, neurinoma, hemangioblastoma, meningioma, lipoma, epidermoid, metastasis, and rarely paraganglioma[1,3,6,13–18]. Out of them, ependymoma is the most common[3,13]. Epidermoid and lipoma can be ruled out by MRI, because they have typical features. Although some characteristic MRI findings of remaining tumors have been described, an accurate prediction of tumor histology is impossible[15,16]. Some authors believe that paraganglioma has typical MRI features which reflect encapsulation of the tumor[6,13] and should be diagnosed preoperatively by this technique[6]. However, our review showed that this tumor has no specific MRI findings. Unfortunately, the same is true for its clinical presentation[4,10,12] which is similar to other cauda equina lesions. Catecholamine secretion of paraganglioma of the cauda

Table 1. MRI features of paraganglioma of thecauda equina – review of the literature

Case	Age/Sex	MRI unit (Tesla)	T1 – weighted (intensity relative to conus)	T2 – weighted (intensity relative to CSF)	Enhanced T1 – weighted
Hayes[9] 1989	34/M*	1.5	isointense	heterogeneous, slightly more intense	
Pigott[12] 1990	36/M	0.15	homogenous		
Raftopoulos[14] 1990	33/M*	1.5		moderately hyperintense	
Iliya[11] 1991	35/M	1.5	hyperintense	isointense (on protondensity weighted)	
Mylonas[10] 1992	62/M	0.5	isointense	hypointense	moderate, homogeneous enhancement
Aggarwal[3] 1993	44/W		isointense with hypointense area	iso-, hypointense, heterogeneous	heterogeneous intense enhancement
Araki[6] 1993	48/M	1.5	isointense, outlined by band of low signal	isointense, outlined by band of low signal	intense homogeneous enhancement
Boukobza[5] 1993	61/W		isointense, homogeneous		heterogenous, intense enhancement
Levy[13] 1993	54/M	1.5	isointense	heterogeneous with hypo- and isointensities and hypointense margin (fast SE sequence)	heterogeneous enhancement
Toyota[7] 1993	53/M				heterogenous, more peripheral enhancement
Wester[1] 1993	61/W		isointense		heterogeneous enhancement
Own case	46/M	1.5	isointense, heterogeneous	heterogeneous, hypointense (turgo SE sequence)	moderate enhancement with linear inhomogeneities
* Recidive of the tumor 16 years (Hayes[9]) and 8 years (Raftopoulos[14]) after initial resection					

equina is rare[7,8]. Clinicopathological features were described in detail by Sonneland et al.[4]. Adequate treatment requires total surgical removal[4].

In conclusion, neither clinical nor MRI (and other radiological) features allow to distinguish paraganglioma of the cauda equina from majority and most common other tumors in this region. Although rare, paraganglioma should be considered in the differential diagnosis of an intradural tumor in this location.

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