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Case Report

Hemorrhagic Paraganglioma of the Cauda Equina: Case Report and Review of the Magnetic Resonance Imaging Features

Woo Yat Ming Peter[1]*, Hung Sze Lok[1], Wong Kai Sing Alain[1], Iu Po Ping[2], Chan Kwong Yau[1] and Kwok Ching Kwong John[1]

Abstract

Cauda equina paragangliomas are rare tumors and are known to be highly vascular. Most patients are middle-aged adult males that present with non-specific symptoms of chronic lumbar back pain. We report a patient with acute cauda equina syndrome caused by a hemorrhagic cauda equina paraganglioma. Intraoperatively a hypervascular intradural extramedullary tumor was encountered and completely excised by first performing circumferential microsurgical devascularization. Although there are no pathognomonic radiological signs, several features supportive of a paraganglioma were identified on magnetic resonance imaging. We advocate meticulous radiological assessment and introduce the polar sign as an additional feature. Establishing a preoperative diagnosis is challenging, but is important in order to alert the surgeon of the operative risks.

Keywords

Paraganglioma; Cauda equina; Magnetic resonance imaging

Introduction

Cauda equina paragangliomas (CEP) are rare, accounting for 3.5% to 5% of all intradural spinal tumors of this region [1]. Although more than 210 cases have been documented only a few reports devoted to their radiological features exist [2-10]. Regardless of their location paragangliomas are highly vascular [11]. We report a patient with a hemorrhagic paraganglioma that presented with acute cauda equina syndrome, review the magnetic resonance imaging (MRI) characteristics and suggest a surgical strategy for excision. A radiological feature, the polar sign, is introduced to aid in the preoperative diagnosis and alerts the neurosurgeon of the risk of encountering such vascular spinal tumors.

Case Report

A 60 year-old male, with a three-month history of chronic lumbar back pain, experienced acute lower limb weakness and retention of urine. Physical examination confirmed cauda equina syndrome characterized by paraplegia, lower limb hyporeflexia and lax anal tone. Pinprick sensation over the L4 to S1 dermatomes and

*Corresponding author: Woo Yat Ming Peter, Department of Neurosurgery, Kwong Wah Hospital, 25 Waterloo Road, Yau Ma Tei, Hong Kong, China, Tel: (+852) 3517 5052; Fax: (+852) 3517 5260; E-mail: peterymwoo@gmail.com

proprioception were diminished bilaterally. Spinal MRI scans revealed a heterogeneously contrast-enhancing intradural extramedullary mass spanning from the T12 to L2 levels (Figure 1). On T2-weighted (T2W) imaging the lesion had a salt-and-pepper appearance with intralesional flow voids and a hypointense rim at its inferior border. In addition T2W hyperintense and T1-isointense signals within the superior and inferior poles of the lesion were noted. The lesion filled the spinal canal with no infiltration into the intervertebral foraminae. Pronounced serpentine perimedullary vascular flow void signals extending from the lumbar to cervical regions were detected. Emergency spinal catheter angiography neither showed evidence of a vascular malformation nor of a tumor capillary blush. The preoperative diagnosis was a conus medullaris ependymoma or a hemorrhagic schwannoma.

A T12 to L2 multilevel laminectomy for tumor excision was performed. Intraoperatively the dura was highly vascular with engorged epidural veins. After durotomy, a vascular intradural extramedullary tan-colored tumor, with several arterial feeders and arterialized draining veins, was encountered (Figure 2). Tumor biopsy resulted in brisk tumor bleeding therefore the initial intention of central tumor debulking was changed to first identifying the tumor's arterial feeders and conducting microsurgical circumferential devascularization. The lesion was subsequently resected and the adherent tumor draining veins were divided. With this surgical strategy gross total excision was performed with a blood loss of 160 ml. Histological examination revealed polygonal tumor cells with round nuclei arranged in compact nests, *zellballen,* surrounded by a capillary network and spindle-shaped sustentacular cells (Figure 3). Immunohistochemistry showed the tumor cells were reactive to epithelial membrane antigen (EMA), neuron specific enolase (NSE) and synaptophysin. There was no expression of glial fibrillary acidic protein (GFAP). The sustentacular cells were positive for S-100 protein staining. The diagnosis of CEP was confirmed. Postoperatively
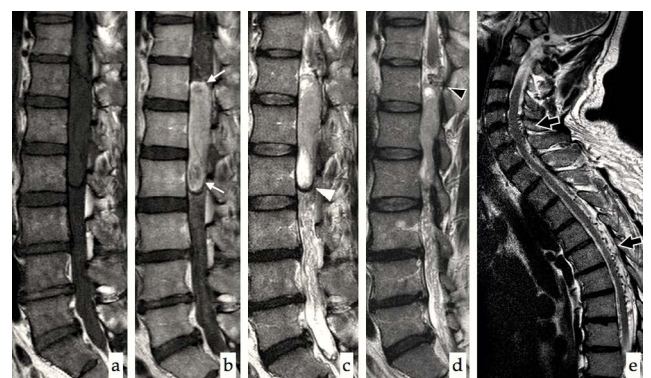

Figure 1: Sagittal MRI of the thoraco-lumbar spine showing an isointense intradural extramedullary spinal lesion extending from the T12 to L2 levels (a, T1W). Intense heterogeneous gadolinium contrast enhancement with intralesional superior and inferior polar hypointense signals, the polar sign (b, contrast T1W, white arrows). T2W imaging revealing corresponding hyperintense polar signal changes with a hypointense rim at the inferior aspect of the lesion, the cap sign (c, white arrowhead). Ectactic serpentine vascular flow voids rostral to the lesion involving the entire spinal cord (d, T2W, black arrowhead; e, T2W, black arrows).
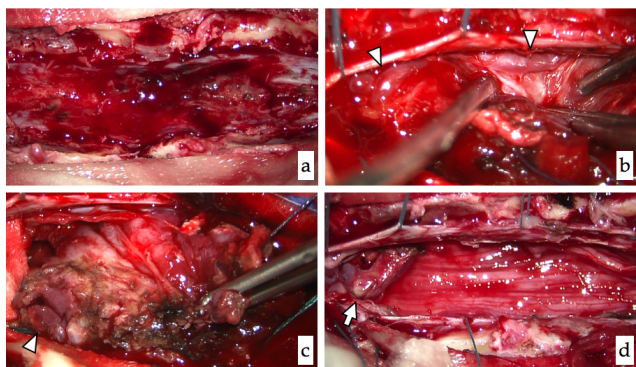
Figure 2: Intraoperative photographs. Vascularized dura exposed laminectomy (a). Hypervascular intradural extramedullary tan-colored tumor with an intralesional organized clot corresponding to the MRI polar sign, several arterial feeders and arterialized draining veins (b and c, white arrowheads) were identified. Complete excision achieved after microsurgical circumferential coagulation of the arterial feeders with preservation of the cauda equina nerve roots. Residual tumor draining vein was divided without clinical sequelae (d, white arrow).
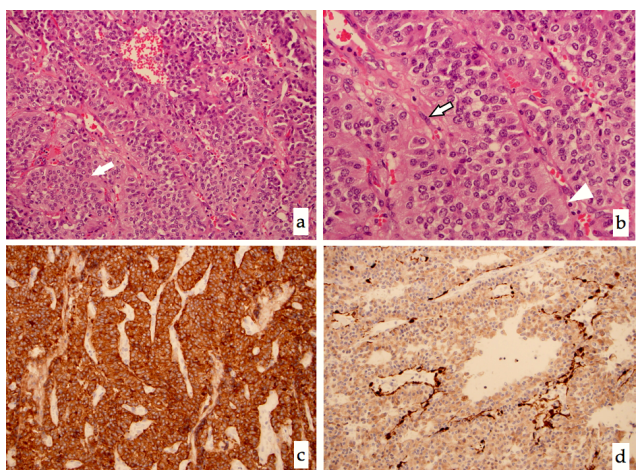

Figure 3: Photomicrographs of CEP. Tumor parenchymal cells arranged in compact clusters (*zellballen*, a, H&E, x200, white arrow) and surrounded by a delicate capillary network. Polygonal parenchymal cells with round nuclei (b, H&E, x400, arrowhead) with supporting spindle-shaped sustentacular cells were observed (white arrow). Immunohistochemical staining showed strong parenchymal cell reactivity to synaptophysin (c) and sustentacular cell positivity to S-100 protein (d).

the patient experienced full neurological recovery and by six months he could walk unaided without any sphincter dysfunction symptoms.

Discussion

The overall incidence of spinal paragangliomas in the general population is estimated to be 0.07% per 100 000 [10]. With improved MRI resolution and precise immunohistochemical techniques this pathology is increasingly recognized [1]. It remains unknown as to why the cauda equina is the most frequent central nervous system site. It has been postulated that aberrant migration of paraganglion cells along the neural tube occurs during embryogenesis, but such cells have yet to be identified in this region [12]. Alternatively, the discovery of ectopic neuroblasts within the adult filum terminale and intradural nerve roots have prompted speculation that paragangliomas arise from dormant precursor cells [1].

Cauda equina paragangliomas affect adults of an age ranging from 12 to 71 years (mean, 48 years) with the majority being male (male: female, 1.54: 1) [1]. They are slow-growing benign lesions that manifest with non-specific chronic back pain associated with sciatica [1,10]. Due to the difficulty in clinically distinguishing from degenerative lumbar spondylosis, the diagnosis is often delayed until substantial tumor growth occurs [13]. Occasionally patients present with cauda equina syndrome due to intratumoral hemorrhage necessitating early operative decompression [1]. Like other extra-adrenal paragangliomas, CEPs are essentially non-functional with only two reported patients exhibiting vasomotor amine syndrome [14,15].

The investigation of choice is MRI. Although there are no pathognomonic radiological signs, several features when considered in combination can be helpful (Table 1). The differential diagnoses for cauda equina region tumors include myxopapillary ependymomas (constituting 90% of lesions), followed by schwannomas and uncommonly meningiomas, hemangioblastomas, metastases, epidermoid tumors and lipomas [16]. The latter two pathologies are readily excluded by virtue of their conspicuous MRI appearance. Epidermoid tumors and lipomas are hypointense and hyperintense respectively on T1W imaging displaying no contrast enhancement [16].

On MRI T1W sequences CEPs are predominantly isointense, centrally located and well-circumscribed. Differentiating characteristics such as cystic degeneration are more frequently observed in schwannomas and hemangioblastomas whereas calcifications are typical of ependymomas. The clinician should appreciate that paragangliomas are vascular tumors and identification of features reflecting this quality is crucial [2-10,11]. These include their invariably intense heterogeneous gadolinium contrast enhancement relative to ependymomas [2]. In comparison, schwannomas are classcially homogeneously enhancing. On T2W sequences intratumoral flow-voids and a salt-and-pepper appearance also indicate hypervascularity [1,8,16]. The presence of a hypointense rim, known as the cap sign, a result of the paramagnetic effects of hemosiderin deposition is another hallmark [2-10,17]. Congested serpentine spinal vessels, distinctively involving the entire spinal cord in our patient, are redolent of paragangliomas although hemangioblastomas also share this feature to a lesser extent [7,8,10,16].

To increase preoperative diagnostic accuracy we introduce the polar sign as an additional radiological feature in differentiating other hemorrhagic cauda equina lesions such as ependymomas, hemangioblastomas, metastatic melanomas and high-grade gliomas. The sign comprises of hypointense signals on contrast T1W sequences and hyper intensities on T2W imaging within the lesion's superior and/ or inferior poles. They represent subacute to chronic intratumoral hematomas at its leading points of growth and were confirmed by operative findings. Upon a retrospective review of MR images in previously published case reports the polar sign was evident, but was not described [1,8,12,13,17-19,].

Although intradural cauda equina metastatic malignancies constitute only 5% of spinal metastases they should be considered as a possible differential diagnosis. From a case series of metastatic cauda equina tumors, more than half of patients had a history of malignancy [20]. The majority being hematogeneously spread from bronchogenic, colorectal carcinomas, melanomas and lymphomas

Table 1: MRI features of intradural-extramedullary tumors of the cauda equina region.

MRI features	Myxopapillary ependymoma	Schwannoma	Paraganglioma	Hemangioblastoma	Meningioma	Metastasis
T1	Iso- to hyperintense	Isointense	Isointense	Hypo- to isointense	Isointense	Isointense
T2	Hyperintense	Hyperintense	Hyperintense	Iso- to hyperintense	Iso-to hyperintense	Iso- to hyperintense
Calcifications	Yes	No	Uncommon	No	Yes	No
Hemorrhage	Yes	Occasional	Yes	Yes	No	No (unless high grade glioma or melanoma)
Salt-and- pepper appearance	No	No	Yes	No	No	No
Intralesional flow voids	No	No	Yes	Yes	No	No
Cap sign	Occasionally	No	Yes	Yes	No	No
Polar sign	No	No	Yes	No	No	No
Ectactic-serpentine spinal vessels	Occasionally	No	Yes	Yes	No	No
Cystic	Occasional	Occasional	No	Common	No	No
Syringomyelia	No	No	No	Common	No	No
Contrast enhancement	Homogeneous	Homogeneous (multifocal if NF II)	Heterogeneous	Heterogeneous	Homogeneous (multifocal if NF II)	Usually heterogeneous (can be multifocal/ CSF disseimnation)
Most common spinal location	Cauda equina or filum terminale	Cervical	Cauda equina or filum terminale	Thoracic	Thoracic	Lumbosacral
Morphology	Sausage-shape	Ovoid, sausage or dumbbell- shape	Ovoid or sausage-shape	Nodular or ovoid	Ovoid or sausage-shape	Nodular
Associated clinical presentation	None	Uncommon: NF II	None	Common: VHL	Uncommon: NF II	Medical history of tumor of the same histopathology

N.B. NF II, neurofibromatosis type 2; CSF, cerebrospinal fluid; VHL, von Hippel-Lindau disease.

[16,20]. Cerebrospinal fluid drop metastases from medulloblastomas, germinomas and high grade gliomas can also occur [20]. For patients without a typical medical history, rapid clinical deterioration is suggestive, but not the rule. Radiologically, metastatic tumors tend to be nodular, heterogeneously contrast enhancing and multifocal with evidence of leptomeningeal sugar-coating, *zuckerguss* [16,20]. They can be easily differentiated from paragangliomas as they seldom grow to an extensive size and rarely develop signs of chronic mass effect such as syringomyelia, ectatic spinal vessels and scalloping of the vertebral bodies (Table 1).

For intradural spinal lesions with serpentine ectatic vessels, catheter angiography can be considered for preoperative particle embolisation to reduce intraoperative blood loss [21]. The procedure is readily available at our center and was performed for our patient soon after admission, but no tumor vascular pedicle could be identified. We also propose methodical microsurgical circumferential devascularization of CEPs, as one would approach intradural perimedullary arteriovenous vascular malformations. This is to prevent retrograde intratumoral venous bleeding before core debulking of these often-large lesions. This strategy should facilitate tumor resection with minimal perioperative morbidity.

Conclusion

Cauda equina paragangliomas are rare benign tumors with an excellent prognosis after complete resection. MRI evaluation for the polar sign, cap sign and serpentine spinal vessels are important clues in deriving a preoperative radiological diagnosis. Awareness of their hypervascularity will aid the neurosurgeon in minimizing the risk of operative adverse events by adopting a surgical strategy of circumferential microsurgical devascularization before proceeding to tumor resection.

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Author Affiliation   **Top**

[1]Department of Neurosurgery, Kwong Wah Hospital, Hong Kong, China

[2]Department of Radiology, Kwong Wah Hospital, Hong Kong, China