Clinics in Diagnostic Imaging (68)

S T Lalloo, A G Mayat, A T Landers, S S Nadvi

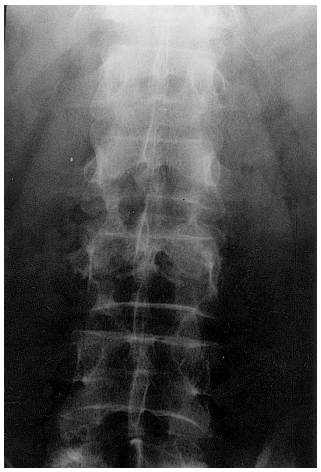

Fig. 1a Frontal radiograph of the lumbar spine.

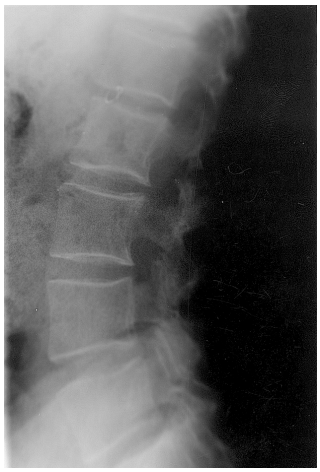

Fig. 1b Lateral radiograph of the lumbar spine.

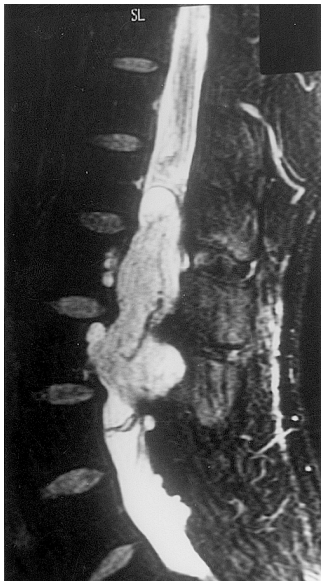

Fig. 2a Sagittal T2-W MR image of the lumbar spine.

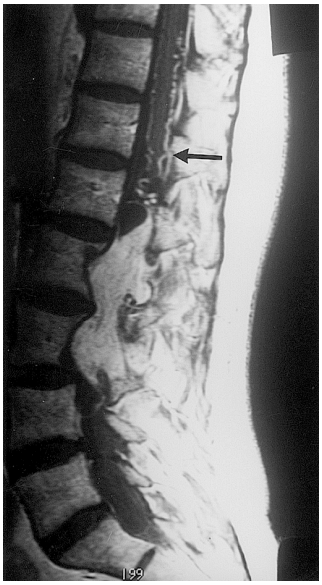

Fig. 2b Gadolinium - enhanced sagittal T1-W MR image of the lumbar spine.

CASE PRESENTATION

A 36-year-old man presented with a 14-year history of low backache. In addition, numbness and weakness in the right leg was noted during the preceding three years, with development of radicular pain involving the left leg in the previous six months. The patient admitted to having impotence for one year. Physical examination revealed saddle anaesthesia and reduced sensation in the L4, L5 and S1 dermatomes bilaterally. His blood pressure was 130/80 mmHg and his heart rate was 80/min. Radiographs of the lumbar spine were performed (Figs. 1a-b), followed by magnetic resonance (MR) scans (Figs. 2a-b). What do these show? What is the diagnosis?

Department of Radiology
Wentworth Hospital
Durban and University of Natal
Private Bag 7
Congella
4013
South Africa

S T Lalloo, MBChB, FCRad
Consultant Radiologist

A G Mayat, MBChB, FCRad
Consultant Radiologist

Department of Neurosurgery
Wentworth Hospital
Durban

A T Landers, MBChB, FCS
Consultant Neurosurgeon

S S Nadvi, MBChB, FCS, MD
Consultant Neurosurgeon

Correspondence to:
Dr Shivu Lalloo
Tel: (27 31) 260 4301
Fax: (27 31) 260 4408
Email: <kasper7@mweb.co.za

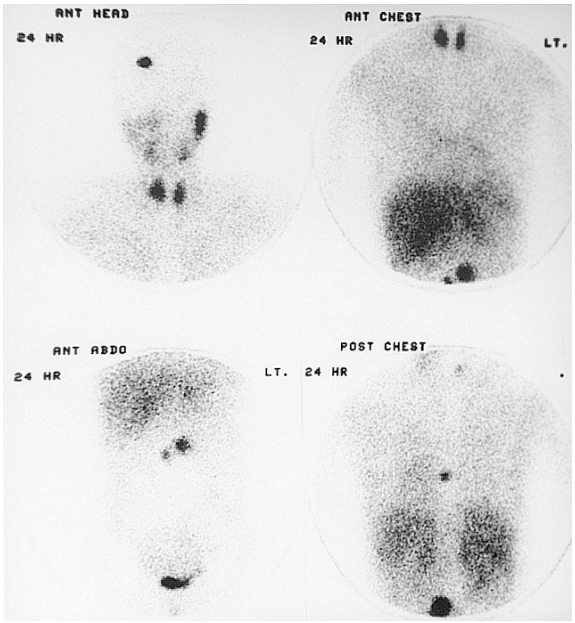

Fig. 3 33-year-old woman with histologically-proven metastatic paraganglioma. Planar MIBG scintiscans show foci of uptake in the calvarium, mandible, para-aortic area and mid-dorsal spine.

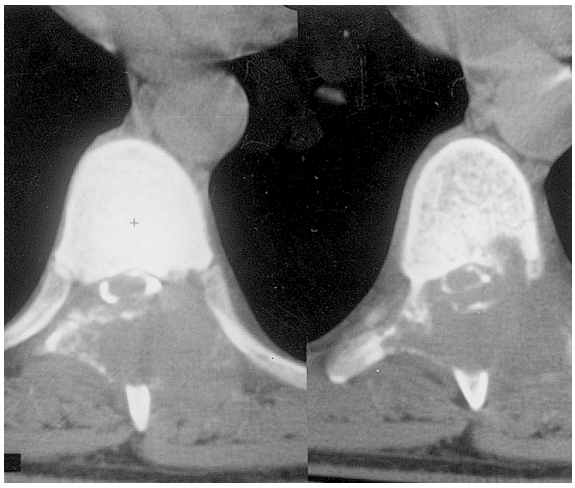

Fig. 4 Same patient as in Fig. 3. 33-year-old woman with metastatic paraganglioma. Axial CT scans taken following myelography show an expansile osteolytic lesion involving the posterior elements of T8 vertebra, with an associated extradural mass compressing the thecal sac. Emergency laminectomy and tumour excision were subsequently performed.

IMAGE INTERPRETATION

Frontal radiograph (Fig. 1a) shows asymmetrical flattening and splaying of the L2 and L3 pedicles due to spinal canal expansion. Lateral radiograph (Fig. 1b) shows posterior scalloping of the L2 and L3 vertebral bodies with widening of the L1-3 neural exit foramina. MR images (Figs. 2a-b) show a T2-hyperintense lobulated expansile tumour located inferior to the conus. There are intrinsic signal voids indicating areas of prominent intra-tumoral circulation. Intense tumour enhancement and hypertrophied pial vessels (arrowed) are present.

DIAGNOSIS

Intradural extramedullary spinal paraganglioma.

CLINICAL COURSE

At L2 - L4 laminectomies, an intradural extramedullary blue-greyish vascular tumour was found and debulked. No blood pressure fluctuation was observed intra-operatively. Pathological examination showed a paraganglioma. A second procedure was subsequently undertaken to remove the residual tumour. The post-operative course was complicated by wound sepsis and cerebrospinal fluid (CSF) leakage, which were managed by debridement, dural patching and appropriate antibiotics. Although right ankle weakness persisted, the patient remained ambulant and continent at follow-up.

DISCUSSION

Paragangliomas are tumours of the paraganglia. Paraganglias are accessory organs consisting of collections of specialised cells of neural crest origin found in association with the peripheral nervous system. Adrenal phaechromocytoma is the commonest variety, and the sites of extra-adrenal paragangliomas include the carotid body, glomus jugulare, glomus tympanicum and para-aortic organ of Zuckerkandl. Although generally benign, the tumour may demonstrate locally-aggressive behaviour and metastasise to lymph nodes, bone, liver, brain and the lung.

Clinically-apparent spinal lesions are uncommon and include metastatic paraganglioma[1-3] (Fig. 3) and less commonly, primary spinal paraganglioma, as seen in our patient. Although several reports of the latter have appeared in the literature, it remains a rare lesion in the differential diagnosis of spinal tumours. The most common locations are the intradural extramedullary cauda equina and filum terminale. Vertebral, extradural and intramedullary sites of origin are much less common[4-7].

Intradural extramedullary (IDEM) lesions occur within the dura but outside the spinal cord, and by convention include cauda equina masses. Clinical signs and symptoms are non-specific. The so-called "meniscus-sign" describing the silhouette of the lesion against the opacified CSF at conventional myelography, is typical of IDEM lesions compressing the cord. With larger lesions, a "complete myelographic-block" may occur. Although MR imaging is the modality of choice, considerable overlap exists in the appearance of individual lesions. The various IDEM lesions are listed in the Table.

Nerve sheath tumours (NST) are the commonest IDEM lesion and together with meningiomas, account for 80%-90% of cases. With neurofibromatosis,

Table. Causes of Intradural extramedullary lesions.

A. Neoplasms		
Primary	- Nerve sheath tumours: schwannoma (neurinoma, neurilemmoma), neurofibroma, ganglioneuroma, neurofibrosarcoma	
	- Meningioma	
	- Paraganglioma	
	- Non-Hodgkin's lymphoma	
	- Other rare dural tumours e.g. melanoma, solitary fibrous tumour, dural angioma	
Metastases	- Dural (e.g. breast, prostate, lung malignancy)	
	- Leptomeningeal:	
	CNS primary	- glioblastoma multiforme, anaplastic astrocytoma, ependymoma, medulloblastoma (PNET-MB), germinoma, pinealoblastoma, choroid plexus papilloma and carcinoma, clear-cell meningioma
	non-CNS primary	- lung, breast, melanoma, lymphoma, leukaemia
B. Tumour-like masses and cysts		
Developmental - epidermoid cyst, dermoid cyst, lipoma, neurenteric cyst		
Arachnoid cyst		
Parasitic cyst e.g. cysticercosis, echinococcosis		
Granulomatous lesions e.g. TB, schistosomiasis, sarcoidosis		
Subdural empyema		
Hypertrophic neuropathies		

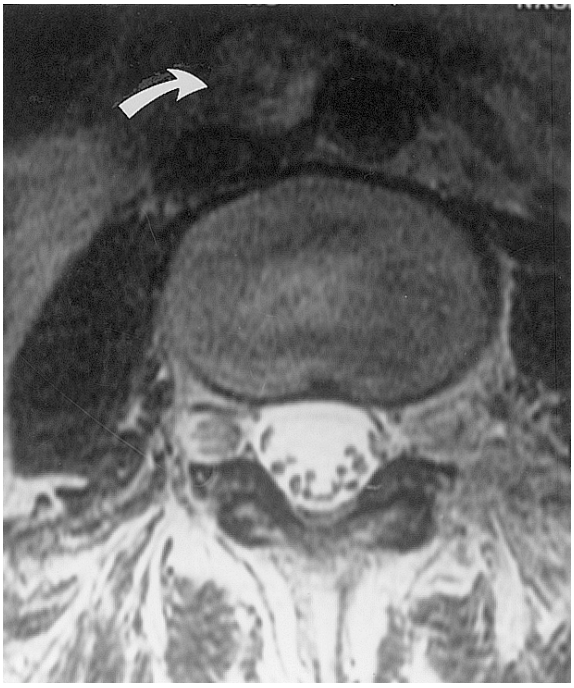

Fig. 5a Same patient as in Figs. 3 & 4. Axial T2-W MR image shows a well-circumscribed heterogeneous inter-aortocaval nodule at the level of the inferior mesenteric artery origin, corresponding to the organ of Zuckerkandl (curved arrow).

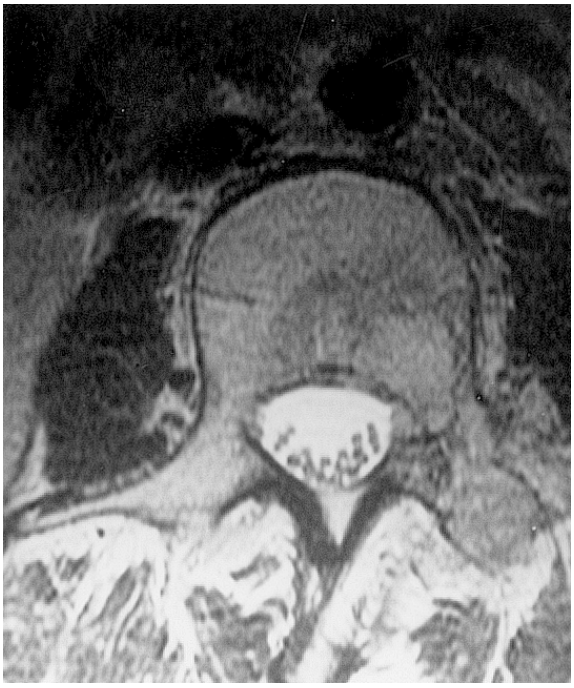

Fig. 5b Same patient as in Figs 3 & 4. Axial T2-W MR image shows an expansile deposit involving the left L3 pedicle and transverse process.

multiple NST and skeletal dysplasia (e.g. kyphoscoliosis, ribbon-ribs) may be seen. Neural foraminal expansion occurs with "dumb-bell" lesions i.e. partly intra- and extradural tumours. A variable degree of Gd-enhancement is seen in virtually all NST. Neurofibromas often demonstrate a "target" appearance on T2-W and enhanced T1-W images, reflecting central fibrocollagenous and peripheral myxomatous tissue. 40% of schwannomas demonstrate cystic components. Meningiomas classically occur in middle-aged females, particularly in the dorsal spine. Enhancement is usually moderate and uniform, and may include a "dural-tail". Multiple meningiomas are a feature of NF-type 2. Rare benign dural tumours (e.g. solitary fibrous tumour, melanoma, capillary haemangioma) and dural metastases may mimic meningiomas. Leptomenigeal metastases may be the result of haematogenous dissemination of non-CNS primary malignancy or

CSF seeding of a primary neuraxis tumour. One-third of patients with medulloblastoma have spinal metastases at the time of initial presentation or at their first recurrence.

Developmental tumours/cysts are congenital and usually associated with spinal dysraphism or segmentation anomalies. Forty percent of epidermoid cysts are acquired as a complication of lumbar puncture, particularly with non-stylet needles. Epidermoid cysts are typically isointense to CSF on most sequences. Dermoid cysts may be intra- or extramedullary, and usually of fat signal intensity. Neurenteric cysts occur anteriorly in the thoracic spine. Lipomas may arise in the filum terminale, or juxtamedullary and subpial in the midline. Eighty percent of the intradural arachnoid cysts occur along the posterior surface of the thoracic cord and most opacify at conventional myelography. Parasitic cysts (e.g. "Racemose" neuricysticercosis, echinococcosis) although uncommon in the IDEM compartment, should always be considered in the differential diagnosis of a cyst in this location, particularly if the patient is from an endemic area. Granulomatous lesions (e.g. tuberculosis, schistosomiasis, sarcoidosis) usually present as intramedullary lesions or diffuse spinal leptomeningeal involvement. However, nodular tumour-like IDEM lesions have been described. Hypertrophic neuropathies (e.g. Charcot-Marie-Tooth and Dejerine-sottas disease) may produce "onion-bulb" nodular thickening of the cauda equina simulating neurofibromatosis.

The clinical presentation of patients with spinal paragangliomas is largely non-specific. Systemic hypertension secondary to catecholamine production is rare with primary spinal paragangliomas but may occur with metastatic functional paragangliomas (phaeochromocytoma) of adrenal, retroperitoneal and mediastinal origin. The functional primary focus may be the organ of Zuckerkandl (Fig. 5a). Radiological findings often mimic other more frequently-encountered spinal tumours such as vertebral metastases (Fig. 5b), ependymoma and schwannoma. The fleshy blue-grey appearance of the tumour at surgery, together with flow voids and intense enhancement on MR imaging (Fig. 2a-b) reflects the vascular nature of these lesions[8]. MIBG scintiscans may reveal other occult paragangliomas (i.e. lesion multiplicity) or metastatic paraganglioma[9] (Fig. 3).

In conclusion, although the clinical presentation and imaging appearances of paraganglioma involving the spine are non-specific, there have been a sufficient number of reports to warrant its inclusion in a more comprehensive differential diagnosis of spine tumour. The presence of previously-undiagnosed systemic hypertension should prompt catecholamine assays. This should be followed by paraganglia MIBG scintigraphy and adrenal imaging, prior to surgery.

ABSTRACT

Paragangliomas rarely present as spine tumours. The correct diagnosis is generally not suspected pre-operatively and initial imaging is often non-specific. A 36-year-old man with low back pain, and progressive leg numbness and weakness, was found to have an expansile intradural extramedullary spinal tumour on radiographs and magnetic resonance imaging. Surgery revealed a paraganglioma. The features of spinal paraganglioma and differential diagnosis of intradural extramedullary tumours are discussed.

Keywords: Paraganglioma, spine tumour, intradural extramedullary lesion, magnetic resonance (MR) imaging, radiography.

Singapore Med J 2001 Vol 42(12):592-595

REFERENCES

1. Chakeres D, Howieson J. Sclerotic thoracic vertebral compression with metastatic phaeochromocytoma. AJNR 1981; 2:477-8.
2. North CA, Zinreich ES, Christensen WN, North RB. Multiple spinal metastases from paraganglioma.Cancer 1990; 66:2224-8.
3. Brodkey JA, Brodkey JS, Watridge CB. Metastatic paraganglioma causing spinal cord compression.Spine 1995; 20:367-72.
4. Sundgren P, Annertz M, Englund E, Stromblad LG, Holtas S. Paraganglioma of the spinal canal.Neuroradiology 1999; 41:788-94.
5. Robinson JC, Kilpatrick SE, Kelly DL Jr. Intraosseous glomus tumor of the spine. Case report and review of the literature. J Neurosurg 1996; 85:344-7.
6. Bessho Y, Kataoka O, Sho T, Kitazawa S, Okada S. Intraosseous glomus tumor in the upper thoracic spine complicating compression myelopathy. A case report. Spine 1991; 16:988-90.
7. Coles CP, Alexander DI, Gross M, Holness RO, Covert AA, Murray SK. Intraosseous paraganglioma of the sacrum: a case report. Can J Surg 2000; 43:137-9.
8. Levy RA. Paraganglioma of the filum terminale: MR findings. Am J Roentgenol 1993; 160:851-2.
9. Razakaboay M, Maillefert JF, Wendling D, Juvin R, Toussirot E, Tavernier C, Phelip X. Bone metastases from a paraganglioma. A review of five cases. Rev Rhum Engl Ed 1999; 66:86-91.