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Fig. 1 Anteroposterior radiograph of the abdomen.

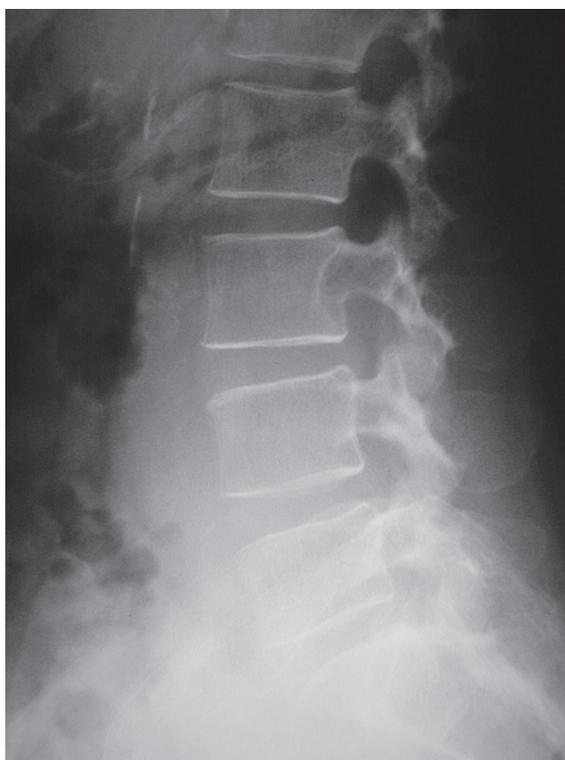


Fig. 2 Lateral radiograph of the lumbar spine.

CASE PRESENTATION

A 62-year-old woman presented with weight loss and anorexia. She had lost 10 kg over the preceding two months. On further questioning, she also complained of low back pain. Her medical history included hypertension and chronic renal failure for which she

was receiving treatment. Physical examination revealed a large smooth abdominal mass on the left side. What do radiographs of the abdomen (Fig. 1) and lumbar spine (Fig. 2) show? What is the diagnosis and what further investigations would be useful in confirming the diagnosis?

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IMAGE INTERPRETATION

Frontal radiograph of the abdomen (Fig. 1) shows a large soft tissue mass on the left side of the abdomen. It is oval-shaped with well-defined borders. There is also erosion of the lower portion

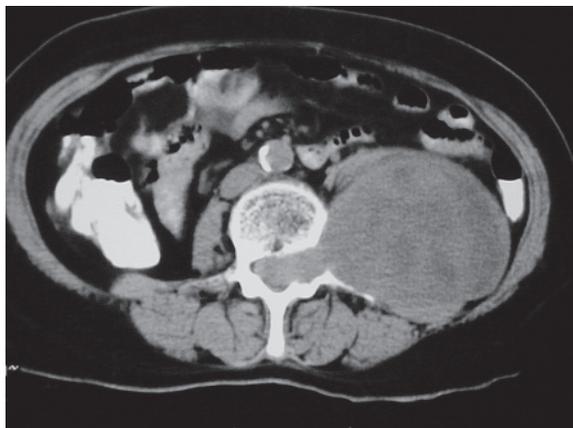


Fig. 3 Axial CT scan of the abdomen shows a dumbbell tumour with large extradural component in the left paravertebral region. The tumour also occupies the spinal canal with erosion of the left posterior L3 vertebral body, left pedicle and lamina.

of the left pedicle of L3 vertebra. Lateral radiograph of the lumbar spine (Fig. 2) shows well-defined posterior scalloping of L3 vertebral body. As these radiographic findings are highly suggestive of a left L3 neurogenic tumour with an extradural component, the next investigation should be a cross-sectional imaging study, such as computed tomography (CT) or magnetic resonance (MR) scan.

CT scan (Fig. 3) shows a dumbbell tumour arising from the left L3 intervertebral foramen. It has a large heterogeneous extradural component extending into the left paravertebral region and retroperitoneal space. Erosions of the left posterior L3 vertebral body, left pedicle and lamina are seen. MR images (Figs. 4a-e) confirm the presence of a large well-margined dumbbell tumour. The tumour shows marked contrast enhancement (Figs. 4b, e). The exact extent of the intradural component is well demonstrated on both axial and sagittal images. The tumour is markedly narrowing the spinal canal.

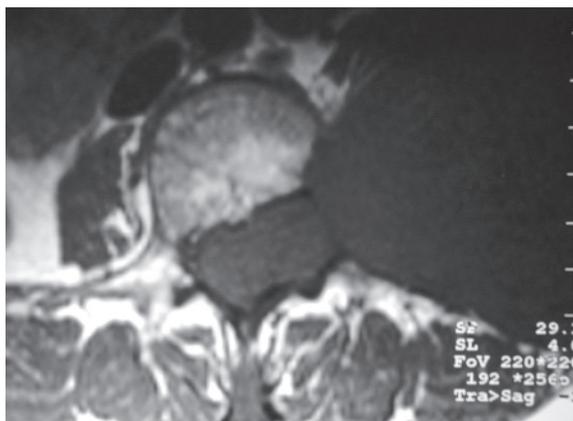


Fig. 4a Axial T1-W MR image of L3 vertebra shows an isointense mass expanding the spinal canal. The dumbbell tumour extends through the L3/4 foramen to form a large left paravertebral mass.

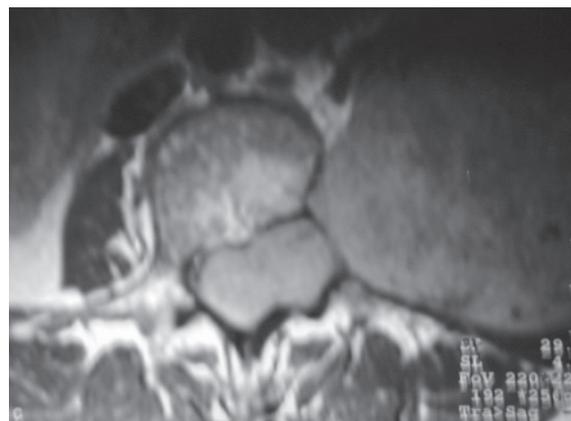


Fig. 4b Enhanced axial T1-W MR image taken at the same level as Fig. 4a shows intense heterogeneous contrast enhancement of both components of the lesion.



Fig. 4c-e Sagittal (c) T1-W, (d) T2-W and (e) enhanced T1-W MR images of lumbosacral spine show a well-demarcated oval intradural component of the tumour. The mass is T1- isointense, T2- hyperintense and displays heterogeneous contrast enhancement.

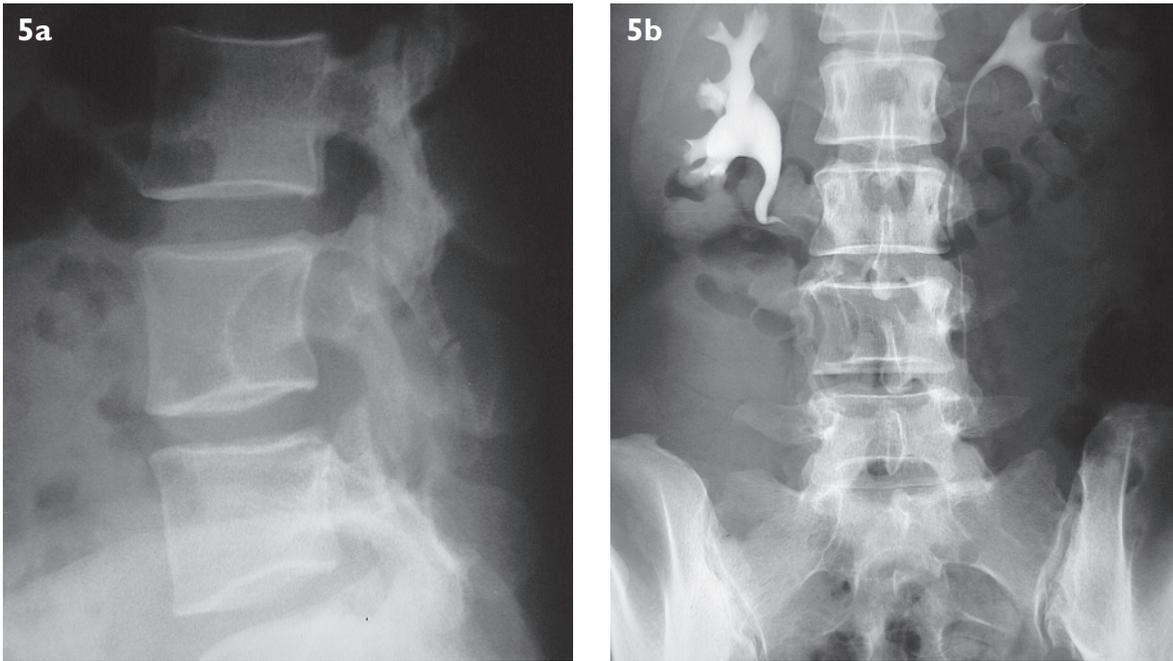


Fig. 5 39-year-old woman with pathologically-proven schwannoma. (a) Lateral radiograph of the lumbar spine shows marked enlargement of the L4/5 intervertebral foramen with a well-defined sclerotic rim. (b) Intravenous urogram shows medial displacement of the right ureter by a large soft tissue mass. The right L4 transverse process and pedicle are eroded. There is a well-defined area of scalloping in the right L4 vertebral body.

DIAGNOSIS

Left extradural-intradural lumbar neurofibroma.

CLINICAL COURSE

Excision of both the extra- and intradural components of the tumour was performed using anterior and posterior approaches. The final histopathological diagnosis was neurofibroma. The patient made an uneventful post-operative recovery. Latest CT scan performed 5.5 years following her presentation shows no evidence of tumour recurrence. At clinical follow-up around the time of the most recent CT scan, there were no symptoms or signs relating to the neurofibroma.

DISCUSSION

Nerve sheath tumours are one of the most common intraspinal tumours and together with meningiomas, constitute 90% of all intradural-extramedullary tumours. Schwannoma, neurilemmoma, neuroma and neurofibroma are all grouped under nerve sheath tumours. There are two main categories of nerve sheath tumours, namely, schwannomas and neurofibromas. Neurofibroma is a separate entity while schwannoma, neurilemmoma and neuroma are synonyms. Neurofibroma is derived from both schwann cells and fibroblasts, forming a fusiform mass that encases the dorsal sensory nerve roots. They may be multiple, and are seen in association with neurofibromatosis type I. In contrast, schwannomas are composed solely of schwann cells that arise eccentrically from the dorsal sensory nerve roots. The

tumour displaces the normal nerve fibres, and tends to appear as a lobulated rather than fusiform tumour^(1,2). Krandsdorf and Murphey found MR imaging differentiation between schwannoma and neurofibroma to be difficult, except in large tumours⁽³⁾. From our own experience, we are in agreement and therefore prefer to refer to these tumours as “neurogenic tumours” (rather than a specific histological type) on MR imaging.

Schwannoma is usually solitary but may be multiple in neurofibromatosis type II. Schwannomas are most commonly seen in the fourth decade of life. They occur in both sexes with about equal frequency, and are distributed uniformly among the cervical, thoracic and lumbar regions. The majority of lesions are intradural-extramedullary (70%-75%), followed by extradural (15%), and dumbbell-shaped with both extra- and intradural components (15%). Intramedullary schwannoma is rare, accounting for less than 1% of these tumours^(4,5). The clinical symptoms and presentation are usually indistinguishable from those associated with disc herniation. Back pain, radiculopathy, paraesthesia and limb weakness are the most common presenting symptoms. Myelopathic symptoms may occur if the tumour compresses the spinal cord⁽⁴⁾.

The radiographic findings in schwannomas, if present, are characteristic. Scalloped bony erosion of the posterior vertebral bodies, erosion of the pedicles and widening of neural foramina are the most common radiographic findings (Fig. 5a). In the presence of a dumbbell tumour, a paraspinal soft tissue mass may be detected radiographically (Fig. 5b). Traditionally,

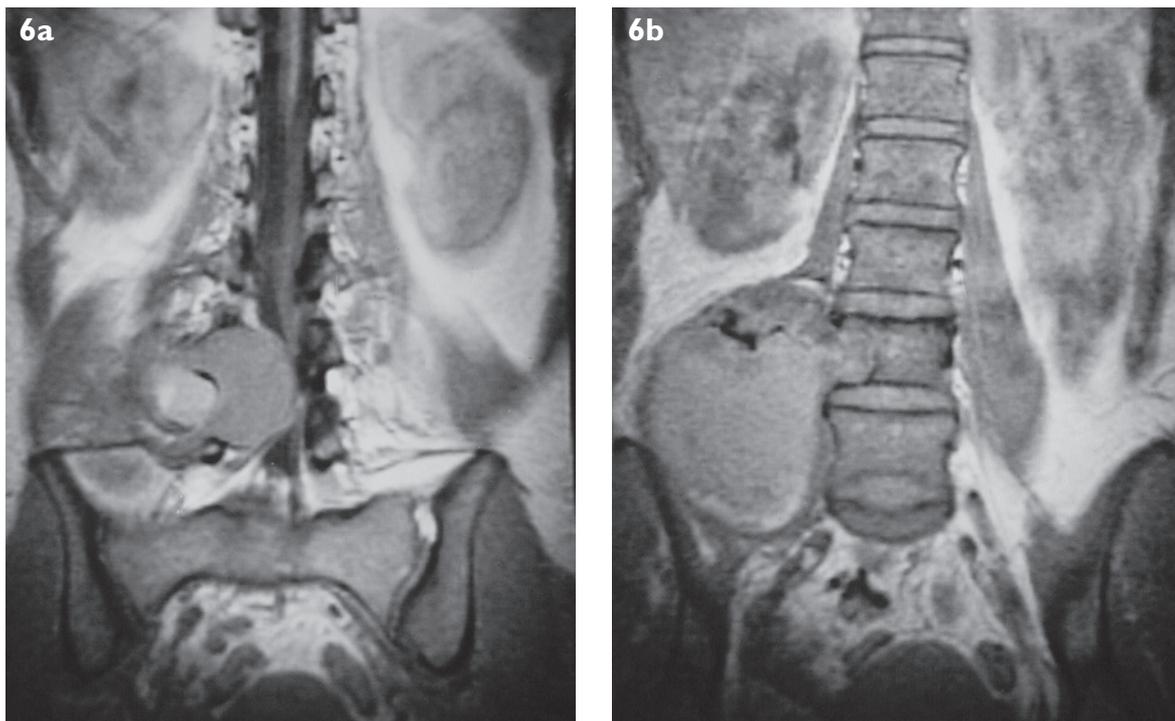


Fig. 6 Same patient as Fig. 5. Coronal T1-W MR images. (a) More posterior image shows the intradural component of the tumour which has eroded the right L4 pedicle and displaced the cauda equina. There is a prominent central hyperintense area, representing subacute haemorrhagic blood products. (b) More anterior image shows the large extradural component of the dumbbell tumour which has extended into the right paravertebral and retroperitoneal area.

myelography was the imaging modality of choice for spinal tumours. Schwannomas are well shown on myelogram and appear as an intradural-extradural lesion with displacement of the spinal cord. The subarachnoid space is widened above and below the tumour ipsilaterally, and there is a sharp interface between the injected contrast agent and undersurface of the tumour.

On CT, schwannomas appear either as an intraspinal mass or intraspinal lesion with an extradural and/or paraspinal component. Bony erosion of the vertebral bodies is well seen on CT. Their CT attenuation varies from hypodense to hyperdense, relative to the spinal cord. Marked homogeneous enhancement is typically seen after administration of intravenous contrast agent. Although the tumour may be well seen on CT, the distinction of the tumour from the spinal cord is sometimes difficult unless a CT myelogram is performed.

MR imaging is presently the imaging modality of choice for all forms of spinal tumours. The majority of schwannomas (75%) are isointense relative to the spinal cord on T1-weighted images, while 25% are hypointense. These tumours appear hyperintense relative to spinal cord on T2-weighted images. Homogeneous contrast enhancement is commonly seen. Cystic degeneration or necrosis within the tumour is not unusual especially in schwannoma. These features are more common in schwannomas than in neurofibromas, and do not necessarily

imply malignant degeneration⁽¹⁻⁶⁾. Haemorrhage is infrequently seen, and is common in large lesions (Fig. 6). Bony erosion of the vertebral bodies and widening of intervertebral foramina are also well seen on MR imaging (Fig. 7).

The prognosis of schwannomas is excellent. The risk of recurrence is very low except with subtotal resection. Complete resection of a dumbbell lesion is possible without neurological deficits by the combined laminectomy and thoracoscopic resection^(7,8) Malignant degeneration in nerve sheath tumours is uncommon, with the reported incidence estimated to be between 1% to 12%. It arises either from pre-existing nerve sheath tumours or de novo. These tumours have a variety of names including malignant schwannoma, neurofibrosarcoma and malignant neurinoma. It is preferable to group all under the category of malignant nerve sheath tumours. Malignant degeneration in association with neurofibromatosis tends to occur at a younger age and to have a worse prognosis^(1,2).

Detection of malignant degeneration on MR imaging is challenging. At present, MR imaging cannot reliably distinguish malignant from benign nerve sheath tumours, although areas of marked heterogeneity, infiltrating margins and irregular bone destruction are more commonly seen in malignant nerve sheath tumours. However, the presence of the target sign may be helpful in distinguishing benign from malignant tumours as the target sign

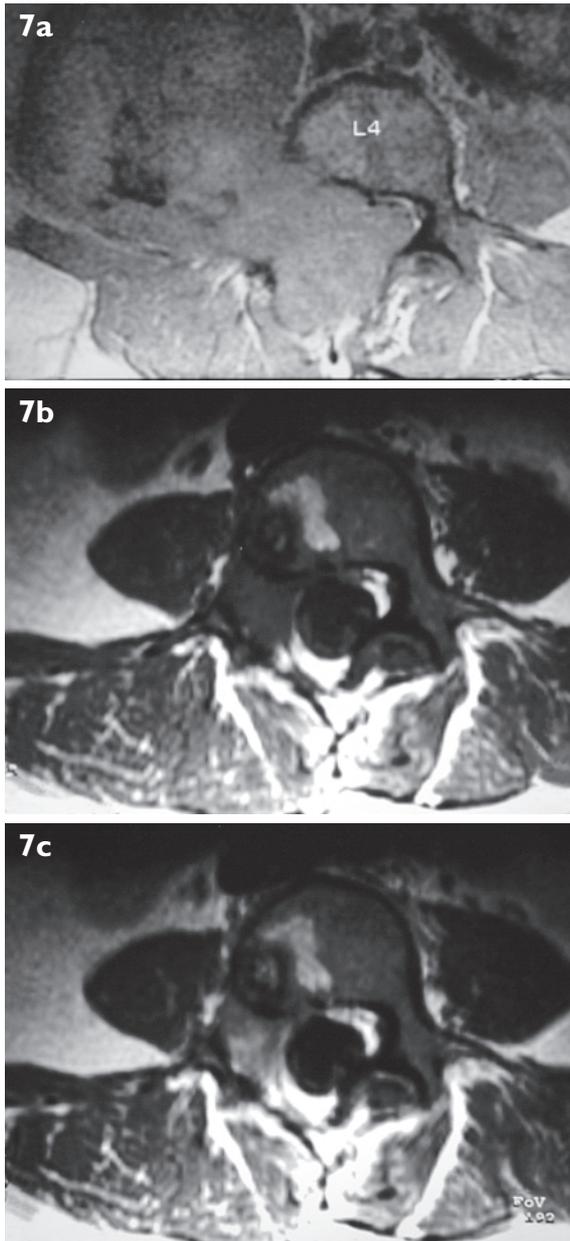


Fig. 7 Same patient as Figs. 5 and 6. (a) Axial T1-W image shows a large dumbbell tumour extending through the expanded right L4/5 intervertebral foramen. The lesion is well demarcated and its signal intensity is mildly hyperintense relative to muscle. Hypointense areas are also present within the lesion compatible with cystic degeneration or necrosis. The right pedicle and posterolateral L4 vertebral body are eroded. The thecal sac is markedly compressed and displaced to the left side by the tumour. Follow-up axial (b) T1-W and (c) enhanced T1-W images, taken six years post-excision, show mildly-enhancing soft-tissue in the right L4/5 intervertebral foramen, consistent with scarring. The thecal sac has a normal outline. There is no evidence of tumour recurrence.

has not been reported in malignant nerve sheath tumours^(9,10). A target sign consisting of a hypointense centre and a hyperintense periphery on T2-weighted images has been described in nerve sheath tumours. The low signal intensity in the central part of the lesion is attributed to dense collagen tissue and Schwann cells. The sign was originally described in neurofibroma, and was subsequently also found to occur in schwannoma^(1,2).

Table I. MR imaging findings of intraspinal-extramedullary tumours.

1. Schwannoma	<ul style="list-style-type: none"> • solitary; multiple in neurofibromatosis • iso- to hypointense relative to spinal cord on T1-W images • markedly hyperintense on T2-W images • intense homogeneous contrast enhancement • cystic degeneration is common
2. Neurofibroma	<ul style="list-style-type: none"> • may be multiple • signal intensity is similar to schwannoma • central hypointensity on T2-W images is more common than in schwannoma
3. Meningioma	<ul style="list-style-type: none"> • hypo- to isointense relative to spinal cord on T1-W images • slightly hyperintense relative to spinal cord T2-W images • intense homogeneous contrast enhancement • frequently posterolateral in location except in the cervical region where they are more likely to be anterior • solitary
4. Metastases	<ul style="list-style-type: none"> • subarachnoid space in lumbosacral region • usually multiple
5. Paraganglioma	<ul style="list-style-type: none"> • most common location: cauda equina/filum terminale • heterogeneous signal due to haemorrhage • intense contrast enhancement
6. Ganglioneuroma	<ul style="list-style-type: none"> • young adults and children • paraspinal mass

The major differential diagnoses of intraspinal-extramedullary tumours include meningioma and drop metastases. Other tumours such as paraganglioma and ganglioglioma are rarer. The MR imaging findings of intraspinal-extramedullary tumours are summarised in Table I.

Meningiomas are slightly less common than nerve sheath tumours, and account for 25% of all spinal tumours. Meningiomas tend to occur in the thoracic region in women over 40 years of age. The location is somewhat different from that of nerve sheath tumours. Meningiomas in the thoracic and lumbar region (except cervical region) are usually located in the posterolateral part of the spinal canal in contrast to nerve sheath tumours that are more commonly anterior in location⁽¹¹⁾. Although the radiographic findings in meningiomas are similar to nerve sheath tumours, and include bony erosion in the vertebral bodies, pedicles and widening of neural foramina, they are less frequently found in meningiomas compared to nerve sheath tumours.

On CT, meningiomas are well-circumscribed, and may be slightly hyperdense or isodense relative to the spinal cord, with intense homogeneous enhancement following the administration of contrast agent. In general, the size of meningiomas is smaller than nerve sheath tumours. Calcifications may be detected

on CT although they are rare. On MR imaging, meningiomas are isointense relative to the spinal cord on T1-weighted images and slightly hyperintense on T2-weighted images. Intense homogeneous enhancement is typically seen after the administration of contrast agent. As with nerve sheath tumours, the cerebrospinal fluid space is widened on the side of the tumour and the spinal cord is displaced contralaterally by the tumour; a characteristic finding in an intradural-extramedullary lesion. Unlike nerve sheath tumours, meningiomas rarely have extradural components or the dumbbell appearance^(4,6,11). Metastasis is another differential diagnosis of an intradural-extramedullary lesion.

Intradural metastatic lesions may arise from primary tumours of the central nervous system (CNS) or from outside the CNS. The most common primary CNS tumour is germinoma, pineal tumour, ependymoma and medulloblastoma. Carcinomas of the breast, lung, melanoma, lymphoma and leukaemia are the most frequent non-CNS sources of intradural metastases. Metastases may be differentiated from nerve sheath tumours and meningiomas based on their location, appearance and number. The lumbosacral subarachnoid space is the most frequent site, and metastases are usually multiple. Furthermore, the presence of a dumbbell appearance would be against metastases^(3,4,11).

Other intradural-extramedullary tumours are rare. Paragangliomas may present as intraspinal tumours that are most commonly located in the cauda equina and filum terminale. These lesions are isointense on T1-weighted and hyperintense on T2-weighted MR images. Heterogeneous signal intensity may occur due to haemorrhage. Intense enhancement is usually present after the administration of contrast agent. Ganglioneuromas are tumours composed of mature ganglion cells and frequently arise from the paraspinal region, mimicking dumbbell tumours. Scalloped bony erosion of the vertebral bodies and extension into the intervertebral foramina are frequently present. However, the age of the patient may help suggest the correct diagnosis, as these tumours are most typically found in children and young adults^(4,11).

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ABSTRACT

A 62-year-old woman presented with weight loss, anoexia and back pain. She was found to have a palpable left abdominal mass. Radiographs, CT and MR imaging revealed a large left 3rd lumbar neurogenic tumour with both intra- and extradural components. A neurofibroma was excised and the diagnosis was confirmed histopathologically. The patient has no recurrence at six years follow-up. The pathological classifications, clinical and imaging features of neurogenic tumours are discussed. With the knowledge of characteristic imaging features, these tumours can be differentiated from other types of intradural-extramedullary tumours.

Keywords: Nerve sheath tumours, Schwannomas, MR imaging, Dumbbell spinal tumours

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