A rare cause of neural foraminal widening

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ABSTRACT

The differential diagnosis for lesions causing neural foraminal widening is vast. The majority are solitary benign peripheral nerve sheath tumours, such as neurofibromas or schwannomas. We present a case of a rare cause of neural foraminal expansion secondary to a posterior thoracic extradural angiolipoma. We describe the presence of chemical shift artefact on post gadolinium T1-weighted imaging as indirect evidence of a fatty component. This potentially important diagnostic sign may raise the suspicion of angiolipoma, especially in an isointense or hypointense dumbbell lesion on T1-weighted imaging, and has not been described previously in this context. Accurate radiological diagnosis of an angiolipoma is important to reduce unexpected haemorrhagic complications from biopsy or resection of the lesion.

CASE REPORT

CASE REPORT

A 71 year old woman presented with compressive symptoms from a large multinodular goitre. The patient was listed for thyroidectomy. Anteroposterior (AP) and lateral chest radiographs, performed as part of anaesthetic assessment, revealed an incidental right-sided posterior mediastinal soft tissue mass projected over the T5/6 neural foramen (figure 1). Routine pre-thyroidectomy unenhanced neck CT confirmed a focal soft tissue density lesion of 40 Hounsfield units (HU) in the right paravertebral pleural region at the extreme inferior aspect of the scan field of view (figure 2). There was also evidence of a discrete peripheral fat-density (-100HU) within the lesion (figure 2).

The patient denied any neurological symptoms. No focal neurological signs could be elicited on clinical examination.

Contrast enhanced chest computed tomography (CT) confirmed hypodense fatty tissue within the soft tissue mass (figure 3A) and revealed heterogeneous increase in CT attenuation relative to the unenhanced scan consistent with a vascularised lesion (figure 3B).

Gadolinium enhanced magnetic resonance imaging (MRI) of the thoracic spine demonstrated that the mass returned heterogeneous signal on T2-weighted imaging and was isointense to muscle on T1-weighted sequences (figure 4A). It expanded, but did not destroy, the T5/6 neural foramen. There was a region of high T1-weighted signal within the mass which correlated anatomically to the hypoattenuating area demonstrated on CT (figure 5A). The lesion enhanced avidly on post-gadolinium T1-weighted MR images consistent with a copious blood supply (figure 4B). Coronal gadolinium enhanced T1-weighted MR images revealed chemical shift artefact at the boundary of the fatty component of the lesion (figure 5B and 5C), providing further indirect evidence of significant fatty component to the mass. In summary, the CT and MR findings demonstrated a well-vascularised soft tissue mass with a fatty component, expanding but not destroying the neural foramen of T5/6. These features are in keeping with a posterior thoracic extradural angiolipoma.

The patient underwent total excision of the lesion. Histopathological examination of the tumour revealed adipose tissue within which were very numerous thin-walled blood vessels of varying calibre. The appearances were typical of an angiolipoma, confirming the radiological diagnosis (figure 5).

DISCUSSION

A dumbbell solitary benign peripheral nerve sheath tumour is the cause of neural foraminal widening in the vast majority of cases [1]. These tumours are divided into two major groups: neurofibroma and schwannoma, with the former being slightly more common.

Benign peripheral nerve shealth tumours are characteristically isointense to muscle on T1-weighted MR images. Heterogeneous signal return is frequently observed on T2-weighted sequences, where high signal corresponds to areas of cystic degeneration and low signal to collagen fibres which may enhance following administration of gadolinium. Where there is a central fibrous component and peripheral cystic degeneration, this constitutes the target sign, which is seen more frequently in neurofibromas than schwannomas [2]. Schwannomas are typically eccentric to the parent nerve but within the epineurium, whereas neurofibroma can obliterate the nerve [2]. Both entities may demonstrate mild uptake of gallium-67 on scintigraphy and can demonstrate avidity for fluorodeoxyglucose 18F (FDG) on PET imaging.

Other rare lesions causing neural foraminal widening include spinal metastasis, solitary bone plasmacytoma, chordoma, malignant fibrous histiocytoma, osteoblastoma, chondrosarcoma, superior sulcus tumour, as well as a malignant peripheral nerve sheath tumour [1, 3]. In addition, a plethora of rare non-neoplastic lesions can widen the neural foramen, including tuberculous spondylitis, vertebral hydatid disease, aneurysmal bone cysts, infraforaminal synovial cysts, traumatic pseudomeningoceles, extradural arachnoid cysts and vertebral artery tortuosity [1, 3]. Our case confirms angiolipoma as an additional cause.

Spinal angiolipomas are rare, benign neoplasms composed of mature fatty and abnormal vascular elements, accounting for 0.04-1.2% of all spinal axis tumours, 3% of extradural spinal tumours and 24% of spinal lipomas [4]. The mean age at presentation is 44 +/- 5.9 years.

The etiology of angiolipomas is not fully understood. There are no known risk factors. It has been postulated that angiolipomas develop from pluripotential mesenchymal stem cells by divergent differentiation along both angioid and adipose tissue cell lines [5]. Hormonal influences are also suggested to play a role in the biology of angiolipoma and may account for the slight female preponderance (female : male = 1.5 : 1) [6]. Pregnancy has been documented as an exacerbating factor in patients with angiolipomas [6]. Furthermore, termination of pregnancy may result in symptom regression [4]. Whether the hormonal effects of pregnancy are responsible or the associated increased adiposity is not certain. The latter theory is supported by the fact that people who gain weight following steroid treatment may also experience increased symptoms [7].

Angiolipomas are thought to represent an intermediate entity in a spectrum with lipomas and haemangiomas at either end [8]. Distinguishing an angiolipoma from soft tissue lipoma or haemangioma may be difficult. Owing to their fat context, lipomas are hyperechoic on ultrasound. They exhibit low CT attenuation coefficients consistent with fat. Due to their fat content, lipomas are hyperintense on T1-weighted and hyper/isointense on T2-weighted sequences with free diffusion characteristics. They are photopenic on scintigraphy and are not FDG avid. The lack of discernible enhancement helps distinguish a lipoma from an angiolipoma. Haemangiomas can have complex ultrasound appearances and may demonstrate low arterial resistance signal consistent with internal blood flow. They tend to contain less discrete fat than angiolipomas and have a CT attenuation similar to muscle. On MR, they are hypo/isointense on T1-weighted sequences but hyperintense on T2-weighted sequences and do not demonstrate free diffusion. Owing to their vascular contents, they may exhibit prominent flow voids on MRI and consequently they enhance avidly. They may demonstrate early uptake on Thallium-201 scintigraphy but do not show avidity for FDG on PET imaging [9]. Another differential to consider is liposarcoma. These lesions manifest as inhomogeneous soft tissue density with varying degrees of fat on ultrasound and CT. On MR, they tend to be hyperintense on T1-weighted and isointense on T2-weighted sequences with free diffusion. They exhibit heterogeneous post contrast enhancement. Distinguishing features include the fact that they usually present as a predominant soft tissue mass in the retroperitoneum. They may also demonstrate uptake of Gallium-67 on scintigraphy and can be FGD avid on PET.

A recent review of all 123 published cases of spinal angiolipoma describes erosion of the walls of the adjacent vertebrae as the most frequent bony manifestation [6]. Other documented osseous sequelae of angiolipomas include enlargement of the interpeduncular distances primarily in infiltrating angiolipomas [10]. Further bony associations have been described including scoliosis [11], osteoporosis [12] and Klippel-Feil [13]. Even though neurofibromas and schwannomas are far more frequent causes of neural foraminal expansion, our case demonstrates that angiolipomas need to be considered in the differential of a dumbbell lesion expanding the neural foramen because of the peri-operative bleeding risk associated with these vascular tumours. Indeed, tumour embolisation, as a precaution, is sometimes performed in cases of angiolipomas prior to surgical resection [7], but not for neurofibromas or schwannomas.

Making the radiological diagnosis of angiolipoma may be difficult. As our case demonstrates, angiolipomas can contain discrete low attenuation material on CT consistent with fat. Despite the high lipid content of angiolipomas, their T1-weighted MRI appearances can vary from hyperintense, through isointense, to hypointense, depending on their associated vascular component [7]. We describe the presence of chemical shift artefact on post gadolinium T1-weighted

imaging as indirect evidence of a fatty component (figure 4C). This is potentially an important diagnostic sign which may raise the suspicion of angiolipoma, especially in an isointense or hypointense dumbbell lesion on T1-weighted imaging. To the best of our knowledge, this diagnostic feature has not previously been reported amongst angiolipomas. In addition, angilipomas tend to be hyperintense on T2-weighted sequences with free diffusion. They enhance following intravenous contrast administration and may demonstrate avidity for FGD on PET imaging.

Accurate radiological diagnosis of an angiolipoma should reduce unexpected haemorrhagic complications from biopsy or resection of the lesion. Conventional angiography can be performed to define the lesion's blood supply and enable pre-operative tumour embolization in appropriate cases. Most patients improve symptomatically following operation. Poor outcomes tend to relate to surgical complications rather than angiolipoma subtypes.

TEACHING POINT

Angiolipomas are a rare cause of neural foraminal widening. Careful scrutiny of CT and MR investigations, looking for evidence of discrete fatty components within a vascularised soft tissue density, can help make the diagnosis.

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FIGURES

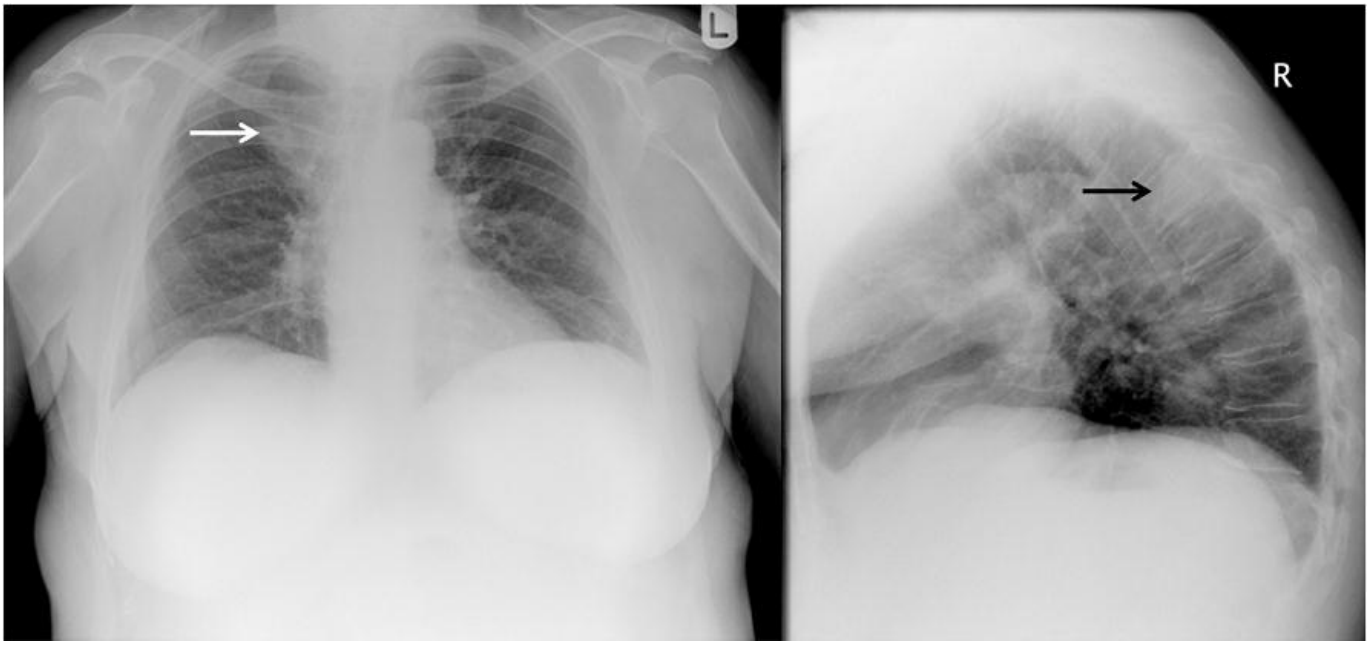

Figure 1. Chest radiograph appearances of an angiolipoma in a 71 year old female. Routine pre-thyroidectomy anteroposterior and lateral chest radiographs reveal an incidental right-sided posterior mediastinal soft tissue mass projected over the T5/6 neural foramen (white and black arrows).

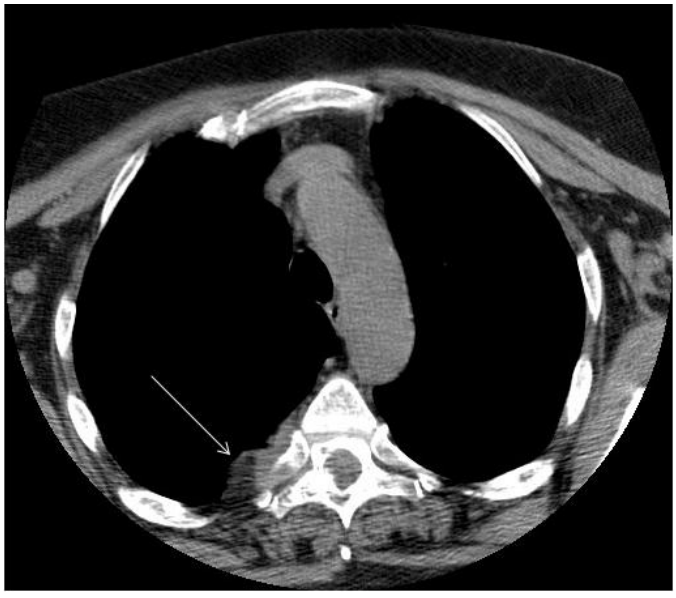

Figure 2. Unenhanced CT appearance of an agiolipoma in a 71 year old female. The inferior most image from a routine pre-thyroidectomy unenhanced axial image from a CT of the neck. It demonstrates a focal soft tissue density lesion in the right paravertebral pleural region with evidence of a discrete peripheral fat-density (white arrow). (Protocol: 120 kV, 45mAs, 2.5mm slice thickness).

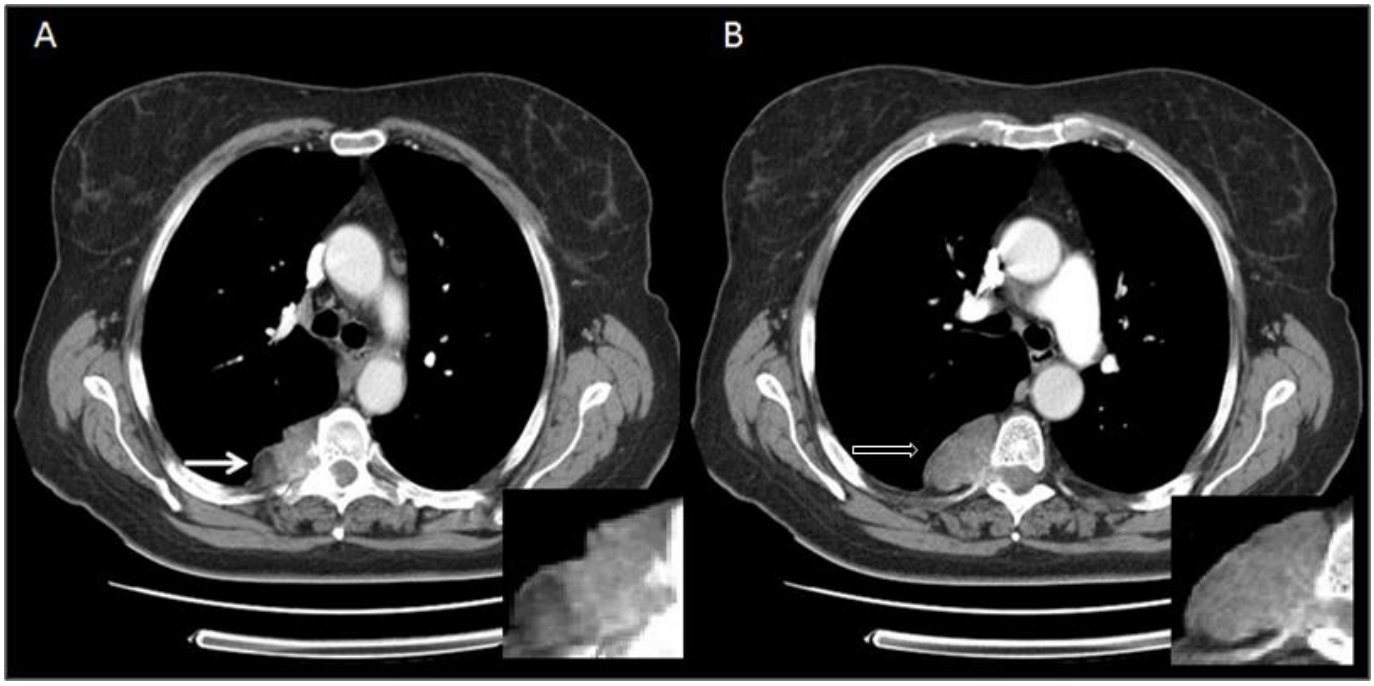

Figure 3. Post contrast CT appearances of an angiolipoma in a 71 year old female. Axial images from post contrast arterial phase CT scan of the chest. 3A) This demonstrates a focal fat density region within the soft tissue mass (solid white arrow). A magnification of the area of interest is demonstrated in the lower right corner. 3B) This demonstrates a heterogeneously enhancing lesion expanding the T5/6 neural foramen (hollow white arrow). A magnification of the area of interest is demonstrated in the lower right corner. (Protocol: 120 kV, 45 mAs, 4mm slice thickness, 100ml Optiray 300 contrast medium).

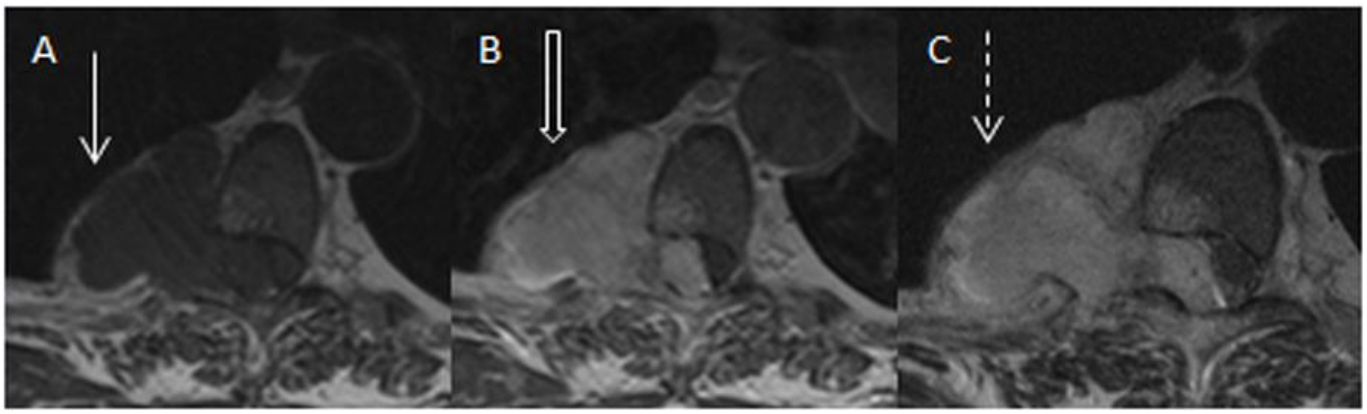

Figure 4. MR appearances of an angiolipoma in a 71 year old female. 4A) Axial image from T1-weighted spin echo sequences (TR 591, TE 17, 4mm slice thickness, FoV 512 x 512) through the thoracic spine revealing a dumbbell shaped lesion which is isointense to muscle and expands the neural foramen (solid white arrow). 4B) Axial image from post- gadolinium T1-weighted spin echo sequence (TR 591, TE 17, 4mm slice thickness, FoV 512 x 512, 15ml Dotarem contrast medium) revealing avid but heterogeneous enhancement (hollow white arrow). 4C) Axial image from T2-weighted turbo spin echo sequences revealing the tumour is of intermediate intensity (TR 5060, TE 99, 4mm slice thickness, FoV 408 x 512) (dotted white arrow).

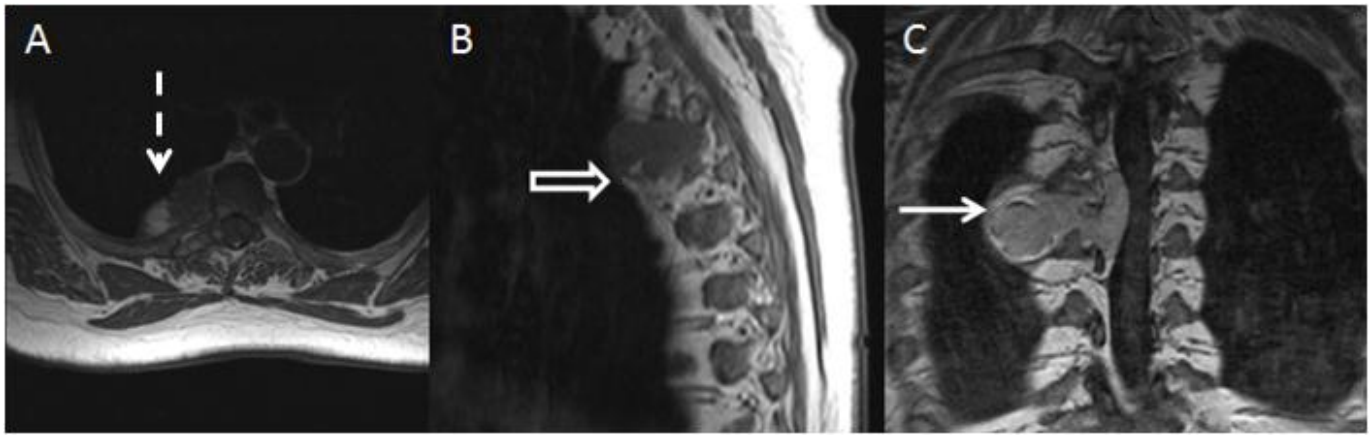

Figure 5. MR appearances of an angiolipoma in a 71 year old female. 5A) Axial image from T1-weighted spin echo sequences (TR 591, TE 17, 4mm slice thickness, FoV 512 x 512) demonstrating a focus of high signal intensity consistent with fat (dotted white arrow). 5B) Sagittal image from T1-weighted turbo spin echo sequences (TR 655, TE 13, 3mm slice thickness, FoV 512 x 512) revealing a rim of hyperintensity around the isointense lesion, in keeping with a fatty capsule (open white arrow). 5C) Coronal image from post-gadolinium T1-weighted spin echo sequences (TR 359, TE 15, 3mm slice thickness, FoV 320 x 240, 15ml Dotarem 15ml contrast medium) demonstrating avid enhancement and a curvilinear region of signal void (white arrow) consistent with chemical shift artefact.

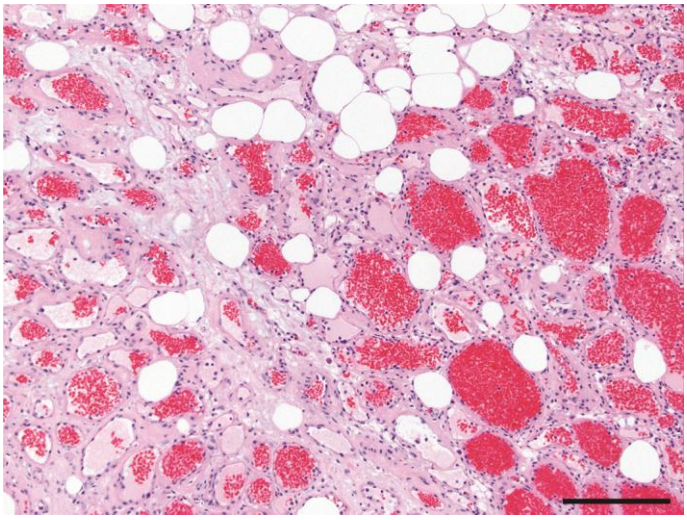

Figure 6 (left). Histology of an angiolipoma in a 71 year old female. Haematoxylin- and eosin-stained section through the tumour composed of adipose tissue interspersed with numerous thin-walled blood vessels of varying calibre, an appearance diagnostic of an angiolipoma. Scale bar = 200µm.

Etiology	Benign tumour composed of mature fatty and abnormal vascular elements caused by divergent differentiation of pluripotential mesenchymal stem cells along both angioid and adipose tissue cell lines.
Incidence	0.14 – 1.2% of all spine tumours 3% of extradural spinal tumours 24% of spinal lipomas
Gender ratio	Female: Male = 1.5 : 1. Hormonal influences have been postulated for the slight female preponderance.
Age predilection	Mean age = 44.03 +/- 5.9 years (range: 1.5 – 85 years)
Risk factors	No known risk factors
Treatment	Total surgical excision +/- pre-operative tumour embolisation to reduce haemorrhagic complications
Prognosis	Most patients improve postoperatively. Poor outcome tends to relate to surgical complications rather than angiolipoma.
Findings on imaging	X-ray – often no bone abnormality CT – soft tissue mass with heterogeneous enhancement, may contain discrete fatty component MR – T1-weighted signal varies depending on degree of fat / vascular elements in the tumour, often hyperintense on T2-weighted sequences, variable enhancement with gadolinium.

Table 1: Summary table of spinal angiolipoma

	Angiolipoma	**Lipoma**	**Soft tissue Haemangioma**	**Liposarcoma**	**Neurofibroma**	**Schwannoma**
X-ray	Soft tissue density	Fat density	Soft tissue density	Soft tissue density	Soft tissue density	Soft tissue density
US	Well defined, fatty component may appear as hyperechoic	Well defined, hyperechoic	Complex, occasional low resistance arterial signal	Soft tissue mass	Well defined, hypoechoic, Obliterating nerve	Well defined, hypoechoic, Eccentric to nerve
CT	Soft tissue mass, may contain discrete low attenuation region (i.e. fat)	Well defined, homogeneous low attenuation	Attenuation similar to muscle +/- fat density	Inhomogeneous soft tissue density with varying degree of fat density	Soft tissue mass, Neural foramina expansion	Soft tissue mass, Neural foraminal expansion
MRI – T1	Hypo / iso / hyperintense, depending on the degree of fat / vascular elements.	Hyperintense	Hypo / isointense, hyperintense fatty areas, may contain flow voids	Hyperintense	Isointense, Obliterate parent nerve.	Isointense, Eccentric to the parent nerve.
MRI – T2	Usually hyperintense	Hyperintense	Hyper-intense, may contain flow voids	Hyper / isointense	Target sign = hypointense centrally & hyperintense peripherally	Heterogeneous intensity
MRI – DWI	Free diffusion	Free diffusion	Free diffusion	Free diffusion	Free diffusion	Free diffusion
Pattern of contrast enhancement	Enhance, degree and pattern of enhancement depend on vascularity	None	Avid	Heterogeneous enhancement	Enhancement of central portion	Heterogeneous enhancement
Scintigraphy	Not documented findings in the literature	Usually photopenic	Early uptake of TI^{201} but delayed washout.	May demonstrate uptake of Ga^{67}citrate	Mild uptake of Ga^{67}citrate	Mild uptake of Ga^{67}citrate
PET	May show FDG avidity	Not FDG avid	Not FDG avid	May show FDG avidity	May show FDG avidity	May show FDG avidity

Table 2: Differential table for spinal angiolipoma

ABBREVIATIONS

AP = Anteroposterior
CT = Computed Tomography
FDG = Fluorodeoxyglucose 18F
HU = Hounsfield Units
MRI = Magnetic Resonance Imaging

KEYWORDS

Angiolipoma; Neural foraminal widening