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Case Report

Infiltrating spinal angiolipoma with paraplegia and hydrocephalus: A rare case report

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الملخص

يعتبر الورم الوعائي نوعا فرعيا نادرا من الأورام الشحمية التي تحتوي على كل من الخلايا الشحمية التامة النمو ومركب الأوعية الدموية الغنية. يسبب ارتشاح الورم الوعائي الشوكي تحديا جراحيا مع احتمال وقوع إصابات على طاولة العمليات للأعضاء المجاورة. نقدم حالة نادرة لارتشاح الورم الوعائي الشوكي عند سيدة يبلغ عمرها ٧٦عاما تسبب لها الورم بشلل نصفي واستسقاء بالرأس. في وقت وصول الحالة، كانت المريضة طريحة الفراش لمدة ثلاثة أشهر بسبب فقدان كامل لقوة الحركة. وقد أظهر فحص التصوير ضررا في الفقرة الصدرية السادسة مع اعتلال النخاع الضاغط. خضعت المريضة لتخفيف الضغط على العمود الفقري واستئصال الورم، الذي تبين أنه ارتشاح ورم وعائي شوكي بالفحص النسيجي. هذه حالة فريدة لأن الورم الوعائي الشوكي غير شائع للغاية وهذه الأورام عموما لا ترشح. يُبرز تقرير هذه الحالة وجود وأهمية الورم الوعائي الشوكي كتشخيص تفريقي لألم الظهر المزمن مسببا العجز العصبي واستسقاء الرأس.

الكلمات المفتاحية: استسقاء الرأس؛ الارتشاح؛ الشلل النصفي؛ ورم وعائي شوكي؛ تخفيف الضغط على العمود الفقري

Abstract

Angiolipoma is a rare subtype of lipoma that contains both mature adipocytes and a rich vascular component. Infiltrating spinal angiolipomas causes surgical challenges with potential on-table injuries to adjacent structures. We present a rare case of infiltrating spinal angiolipoma in a 76-year-old woman who presented with paraplegia and hydrocephalus. At the time of presentation, the patient was bedridden for 3 months due to a complete loss of motor power. Imaging examination revealed a lesion involving the T6 vertebra with compressive myelopathy. The patient underwent spinal decompression and resection of the lesion, which turned out to be an infiltrating angiolipoma by histological examination. This is a unique case because spinal angiolipoma is extremely uncommon and such lesions generally manifest without infiltration. This case report highlights the existence and importance of spinal angiolipoma as a differential diagnosis of chronic backache resulting in neurological deficits and hydrocephalus.

Keywords: Hydrocephalus; Infiltrating; Paraplegia; Spinal angiolipoma; Spinal decompression

Introduction

Lipomas are also known as 'universal tumours', capable of arising anywhere in the body except in the brain parenchyma. They arise from adipocytes, and are usually benign, slow-growing tumours; therefore, lipomas are typically harmless. Histopathologically, they are well capsulated, allowing easier and complete removal.

Angiolipoma is a less well-known entity of lipoma, accounting for 5–17% of all lipomas. In 1960, Howard and Helwig described it as comprising both mature adipocytes

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and a rich vascular component, as cited by Han et al.[1] The vascular component could be capillaries, venous, sinusoidal, arterial, or even a combination of elements. A more detailed description given by Lin includes the following significant criteria for angiolipomas: gross evidence of tumour formation regardless of encapsulation, microscopic evidence of mature lipocytes comprising at least 50% of the tumour population, and microscopic evidence of complex angiomatous proliferation with fibrin thrombi within the tumour.[2]

According to their nature, angiolipomas can be either infiltrating or non-infiltrating. The difference between these two distinct natures is due to the presence of encapsulation. Non-infiltrating angiolipomas are more common than the former, are similar to lipomas, and can be removed entirely due to similar encapsulation. However, they stand out clinically as tender nodules. The infiltrating angiolipomas' unencapsulated histology allows infiltration, and infiltrating spinal angiolipomas cause surgical difficulties and impose on-table injury risk to adjacent structures. In addition, they can recur despite removal due to the residual infiltrating tissue. Angiolipomas can be classified according to sites, although they are more commonly found in the subcuticular layer of the forearms, trunk, and face; a few unusual sites are spinal angiolipomas, adrenal angiolipomas and gastric angiolipomas.

Statistically, spinal angiolipomas account for 0.14–1.2% of all spinal tumours, 3% of extradural spinal tumours, and 24% of spinal lipomas.[3,4] We present a rare case of infiltrating spinal angiolipoma in a 76-year-old woman, resulting in paraplegia and hydrocephalus.

Case report

A 76-year-old woman presented to our Orthopaedics department with insidious onset of lower backache for 6 years, which gradually worsened over time. It started as a dull aching pain, and it was then associated with radiation to the bilateral lower limbs. She also developed weakness in both her lower limbs. Initially, she experienced difficulty in walking, especially while climbing the stairs, and it gradually worsened over a period of time. At the time of presentation, she was bedridden for 3 months due to a complete loss of motor power.

Upon further probing of her history, she was found to have changes in bladder habits. She had urge incontinence of increasing severity for the past 3 years. She complained of frequent episodes of headache for 1 month with increasing intensity. She also had hypertension and diabetes mellitus, and was under regular medication.

On local examination of the back, there were no abnormalities. Her vital signs were normal and hypertension was well-controlled. The Glasgow Coma Scale was $E_4V_5M_6$. Her higher mental function and the examination of cranial nerves and upper limbs were remarkably normal. There was spasticity with grade 0/5 motor power in both lower limbs. There was a sensory deficit from the level of T6 dermatome downwards for soft touch and pinprick. Bilaterally, the knee jerks were exaggerated, and the extensor plantar reflex was noted, suggesting an upper motor neuron pattern of neurological involvement. Other systemic examinations were normal.

Routine blood investigations (complete blood picture, renal function tests, and liver function tests) revealed no abnormalities. Plain radiographs of the thoracic spine did not show any lesions in either view (Figure 1). Bone marrow aspirate showed only reactive marrow changes with no evidence of myeloma.

Computerised tomography (CT) and magnetic resonance imaging (MRI) of the thoracic spine were performed. In the sagittal and coronal views of the MRI, a lesion involving the T6 vertebra causing compressive myelopathy was found

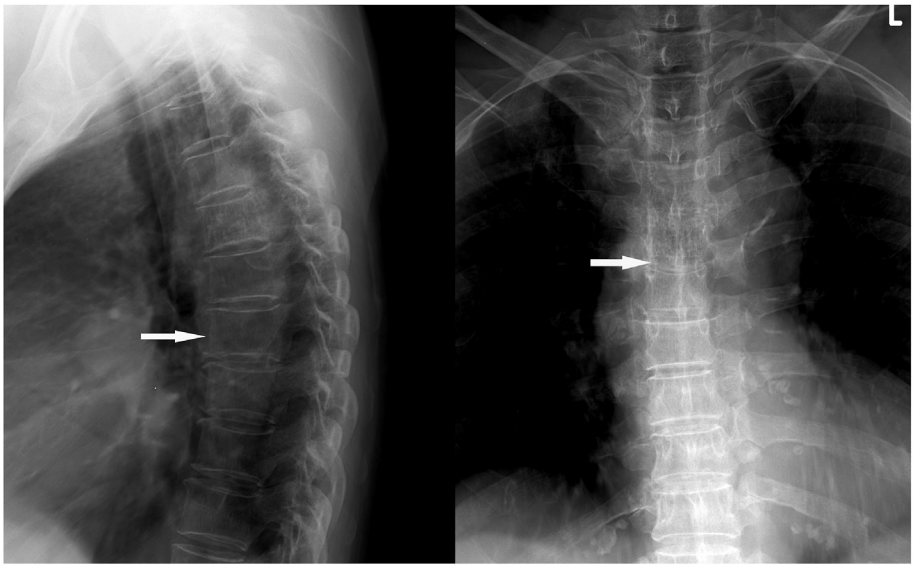

Figure 1: Plain radiograph of the anteroposterior and lateral views showing no abnormality in T6 vertebra. Arrows are pointing to the T6 vertebra in both views.

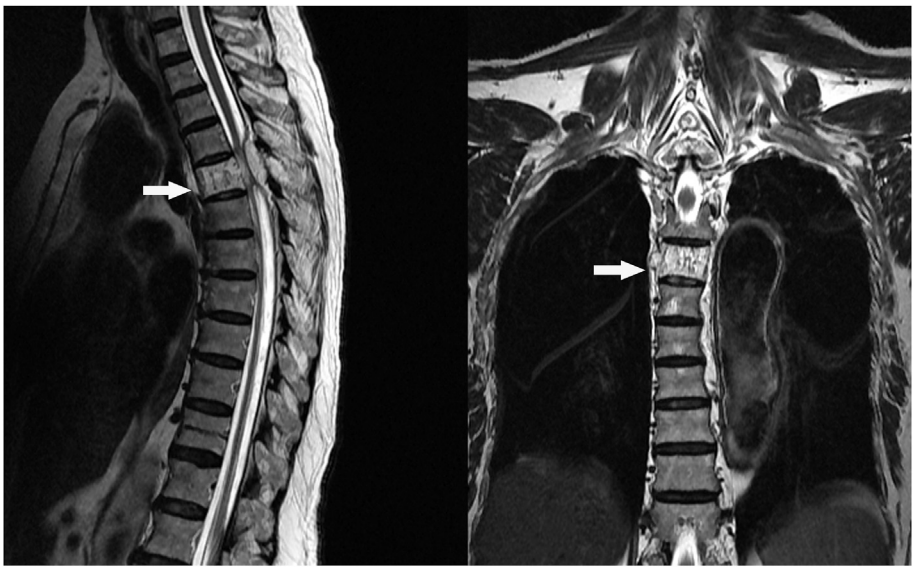

Figure 2: Sagittal and coronal sections of T2 weighted magnetic resonance imaging showing lesion in the T6 vertebra (arrows) and compressive myelopathy. Note the posterior cord compression.

(Figure 2). Axial CT (Figure 3A) and MRI (Figure 3B) showed that the lesion involved the body and posterior elements of the T6 vertebra. CT of the brain was performed because of her headache, and there was an enlargement of both lateral ventricles, which was suggestive of hydrocephalus (Figure 4).

A posterior spinal decompression was conducted and a tissue sample was obtained for histopathology. Accordingly, spinal decompression and surgical stabilisation were performed. Intraoperatively, the tumorous tissue was found in the epidural space after the T6 lamina was removed. Incomplete resection of the tumour was performed as the margins were not well demarcated. Histopathological examination showed haematoxylin-eosin-saffron (HES)-stained adipose tumour. It was composed of mature adipocytes admixed with large and small ecstatic thrombi-congested vessels infiltrating the marrow space (Figures 5A and 5B). She was diagnosed with an infiltrating spinal angiolipoma. The recommended treatment for this is total surgical resection. Typically, the surgical approach needs to be individualised as per the extension of the tumour. Adjuvant radiation therapy should not be used in the treatment of patients with spinal angiolipomas since the prognosis of the infiltrating group, even with incomplete resection, is very good.

Our patient was not willing to undergo further treatment due to financial constraints, and she was discharged from the hospital against medical advice.

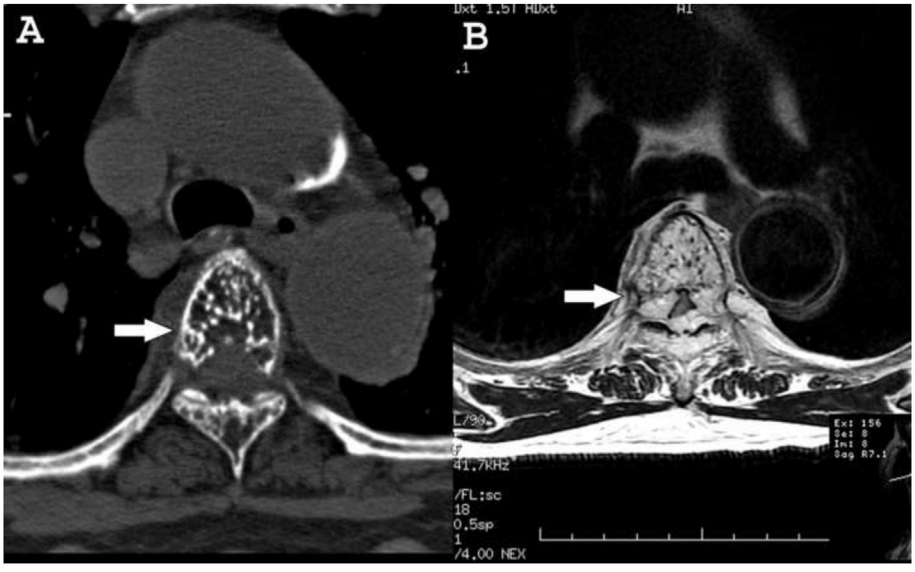

Figure 3: Axial views of computerised tomography (**3A**) and magnetic resonance imaging (**3B**) at T6 vertebra. The lesion can be observed in the entire vertebra including the posterior elements (arrow).

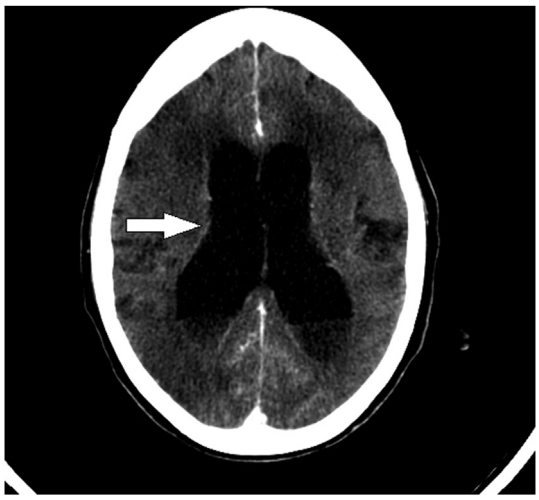

Figure 4: Computerised tomography of the brain showing dilated lateral ventricles (arrow) compressing the brain parenchyma.

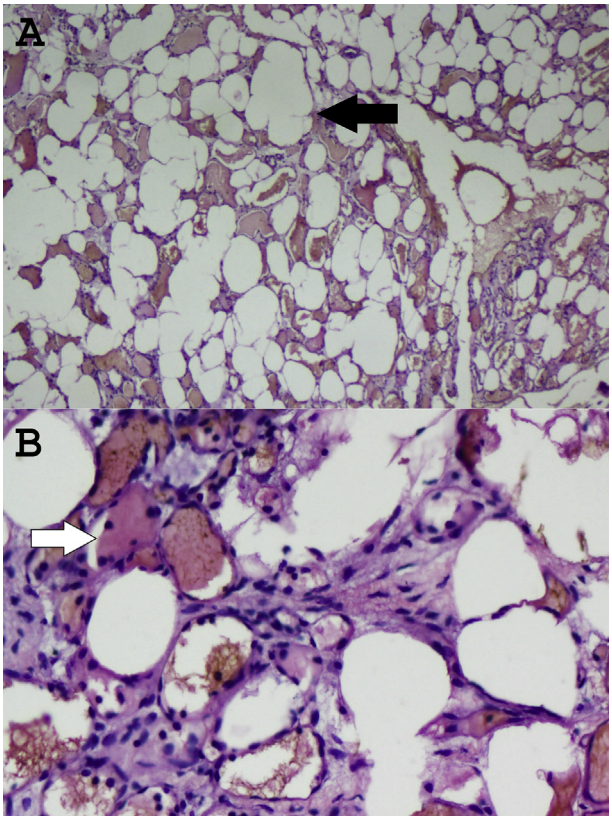

Figure 5: Histopathology at 10 × (**5A**) and 40 × (**5B**) of the T6 posterior elements showing tumour infiltrating the marrow spaces. The tumour consists of mature adipocytes (black arrow) with vascularity. Thrombi are noted in the blood vessels (white arrow).

Discussion

A case of infiltrating angiolipoma with hydrocephalus is rare. This case is unique because spinal angiolipoma is a rare entity, and it usually manifests without any infiltration. Our patient presented with complete paraplegia, but showed no signs of increased intracranial pressure except headache. In view of paraplegia, a spinal decompression and stabilisation procedure were decided.

In 1890, the first published case of spinal angiolipoma was reported. It was a doctoral dissertation about a 16-year-old male patient with numerous cutaneous lipomas who developed progressive paraparesis with hyperreflexia. The autopsy revealed a thoracic cutaneous lipoma infiltrating the spinal canal with a predominantly vascular component and compressing the spinal cord from C6 to T5.[3] A decade later, Liebscher described a primary spinal angiolipoma for the first time.[3]

Spinal angiolipoma is more common among women, with peak incidence among the age group of 40–60 years.[5] This demographic detail tallies with our patient. It usually arises in the thoracic epidural space. This is believed to be due to the variation in the blood supply along the spinal cord. The mid-thoracic region is the least perfused region. In accordance with this, the tumour in our patient was also found to be at level T6.

Preul et al.[6] reported back pain, numbness, paraesthesia, and weakness of the lower limbs as the usual presenting complaints. Paraparesis, sensory disturbances, and hyperreflexia were the most common presenting signs. Spinal angiolipomas commonly arise from the epidural layer. Hence, the neurological symptoms depend on the level of the spine being subjected to compression of the tumour.[4] This explains the paraplegia and urge incontinence of our patient. A compression at T6 acts as an upper motor lesion affecting the impulse transmission of the nerves below, resulting in spasticity of the lower limbs. Urge incontinence usually occurs because of more parasympathetic activity owing to the injury of the sympathetic nerves. Our patient's symptoms were gradual due to the slow-growing nature of the tumour. The infiltration of the tumour from the paravertebral space into the spinal canal caused an obstruction of the cerebrospinal fluid flow, leading to a communicating hydrocephalus in our patient.[3] However, this is also a rare phenomenon. It generally does not infiltrate and is in the posterior or posterolateral locations.

This patient did not agree to undergo any further management due to financial constraints. We believe that a complete surgical excision of the tumour posed a higher risk because of her age. In addition, evidence of infiltration suggests that complete excision is not possible. Furthermore, long-standing compression of the spinal cord does not guarantee full recovery of the spinal cord. Surgical decompression of the spinal cord and spinal stabilisation were performed only in view of the rehabilitation of the patient.

In general, complete excision of the spinal angiolipoma has an excellent postoperative outcome.[7,8] The most common route is removal of the lamina process of the involved vertebrae.[9] Even when complete excision is not possible in infiltrating tumours, a partial excision promises a relatively good prognosis. However, partial excision should be performed only after considering other confounding factors, such as anaesthesia risk and other surgical comorbidities. Recurrence following surgery has not been reported as these tumours are slow-growing, and they do not have the capability to metastasise. Hence, surgical treatment is the best option.

Since spinal angiolipoma is a vascular tumour, some have used preoperative embolisation.[8] This is, however, debated as

bleeding is generally not reported during surgical removal.[9] Whether postoperative radiotherapy should be administered in infiltrating spinal angiolipoma is under debate. Some argue that it is unnecessary as the risk of recurrence is low, and even if it recurs, a second surgery is a better option. However, there is a consensus in not opting for neo-adjuvant therapy unless malignancy is suspected.[9]

In summary, spinal angiolipoma with an infiltrating nature is important because of its rarity. Slowly progressing neurological deficits develop due to infiltration and secondary canal stenosis. One needs to consider the association of infiltrating spinal angiolipoma with hydrocephalus. Despite its infiltration, spinal angiolipoma takes years to present and does not metastasise. Early recognition, surgical excision, and histopathological confirmation are essential.

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Conflict of interest

The authors have no conflict of interest to declare.

Ethical approval

Departmental Review Committee approval was obtained (KMC/Ortho/0829/2020 dated August 29, 2020) for submission to the journal.

Consent

Consent for publication of this report has been obtained from the nearest relative.

Authors' contributions

DV conceived and acquired the data, including articles relevant to the case report. SNB drafted the initial article and revised it critically for important intellectual content. SPM revised it critically for important intellectual content and approved the final version submitted. All the authors critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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