



Dural Amyloidoma Located in the Falx Cerebri and Spinal Dura: Two Case Reports

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ABSTRACT

Amyloidosis is a progressive disorder marked by the aggregation of insoluble fibrillar proteins in various tissues, leading to tissue damage. Localised amyloidosis, known as amyloidoma, is particularly rare in the central nervous system and may be mistaken for a neoplastic lesion due to similar radiological features. Consequently, the general treatment approach often involves total excision. However, it is important to consider that a biopsy of the amyloidoma, rather than total excision, may suffice for complete recovery when paired with appropriate systemic treatment. This report presents two rare cases of amyloidoma: one located in the falx cerebri and the other in the lumbar spine. Both cases were successfully operated on, with the patients recovering without complications following treatment. (1)

KEYWORDS: Amyloidoma, Amyloidosis, Dural amyloidoma, Cerebral amyloidoma

ABBREVIATIONS: MRI: Magnetic resonance imaging, CT: Computed tomography, CNS: Central nervous system

INTRODUCTION

Amyloidosis is a progressive, heterogeneous group of disorders caused by the abnormal deposition of toxic, insoluble fibrillar protein aggregates in various tissues, leading to tissue damage. For this reason, it is called a protein-folding disorder. The aetiology of amyloidosis remains unclear. The disease can be localised, wherein amyloid accumulates in different tissues and causes a mass effect. This condition is known as amyloidoma, and its clinical presentation varies depending on the affected tissue (1,4-6).

The incidence of amyloidosis is 1-2 per 100,000 per year in Western countries. In the United States, 1,275 to 3,200 new

cases are reported annually (3). However, specific data on amyloidoma are limited due to its rarity. Cerebral amyloidoma cases are reported predominantly in middle-aged individuals, with a female predilection. Amyloidoma is most frequently seen in the kidneys and gastrointestinal system, whereas brain involvement is uncommon. The involvement of the dura is also quite rare, and information about this condition is primarily based on case reports, challenging the acquisition of precise epidemiological data (2). Intracranial amyloid accumulation is extremely rare and can be seen in the cerebral parenchyma, skull base, pituitary gland and cerebellopontine angle. Parenchymal involvement is frequently observed in systemic amyloidosis, particularly in AL amyloidosis. Amyloid deposits

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in the calvarium often show a predilection for the skull base and are described as calcified lesions (5).

While the diagnosis of amyloidoma is confirmed through histochemical methods, it is often mistaken for other conditions before biopsy due to its radiological features. These lesions are contrast-enhancing and can mimic high-grade lymphomas and glial neoplasms, as demonstrated in a case report on a spinal extradural mass (1,2,4,6).

In this report, we present two cases: those of a young woman presenting with headache and dizziness, diagnosed with an amyloidoma involving the falx cerebri, and an older woman presenting with lower back and leg pain, diagnosed with a lumbar dural amyloidoma.

CASE REPORTS

Cerebral Dural Amyloidoma

A 32-year-old female patient presented with headaches, dizziness, and nasal bleeding lasting a few weeks. Her medical history, neurological examination, and blood tests were normal upon admission. Cranial imaging revealed a gadolinium-enhancing, hyperintense lesion on T2-weighted magnetic resonance images (MRI) and a hyperdense lesion on computed tomography (CT). The lesion measured 34 × 11 × 93 mm and was extra-axial, located in the frontal interhemispheric area and extending to both sides of the falx cerebri (Figure 1). Central nervous system (CNS) lymphoma, meningioma, or glial tumours were considered primary differential diagnoses, and the patient underwent surgery using an anterior interhemispheric approach. During the surgery, the tumour was observed to be associated with the dura. The lesion was successfully sub-totally removed. A pathological analysis confirmed the diagnosis of amyloidoma. Congo red and crystal violet histochemical stains were positive, and immunohistochemical analysis showed a polytypic reaction for kappa and lambda light chains (Figure 2).

Postoperative examinations were normal, and the patient was referred to the Haematology Department for further investigation of systemic amyloidosis. No systemic amyloidosis was detected, so systemic treatment was required. Three months after the surgery, follow-up imaging showed spontaneous regression of the tumour (Figure 3).

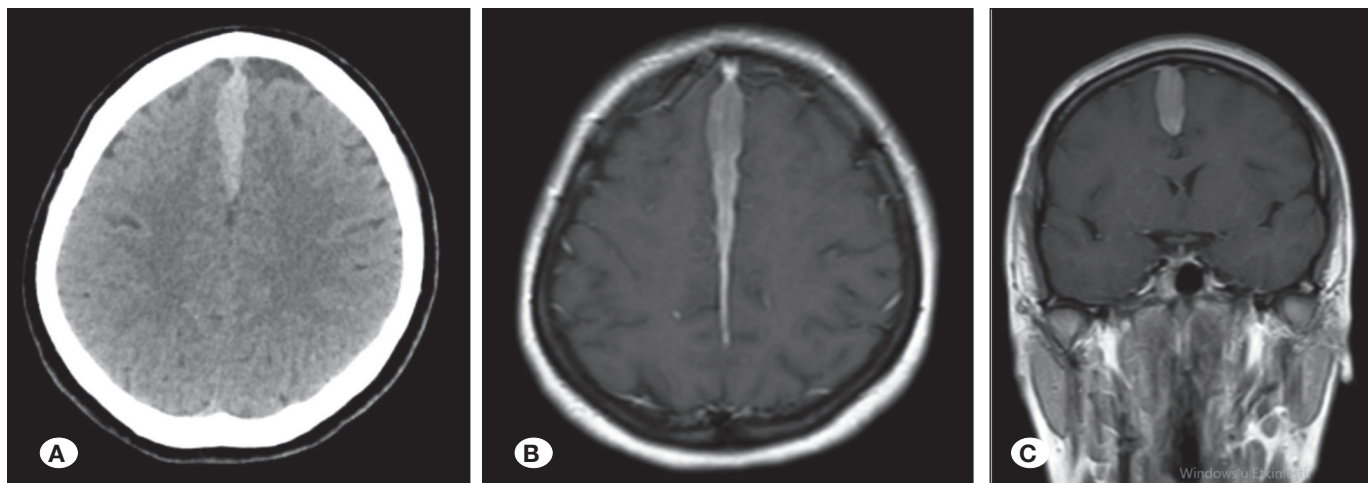


Figure 1: The patient's non-enhancing axial CT (A), gadolinium-enhanced axial (B), coronal (C) T1W MR images show a parafalcine extra-axial mass.

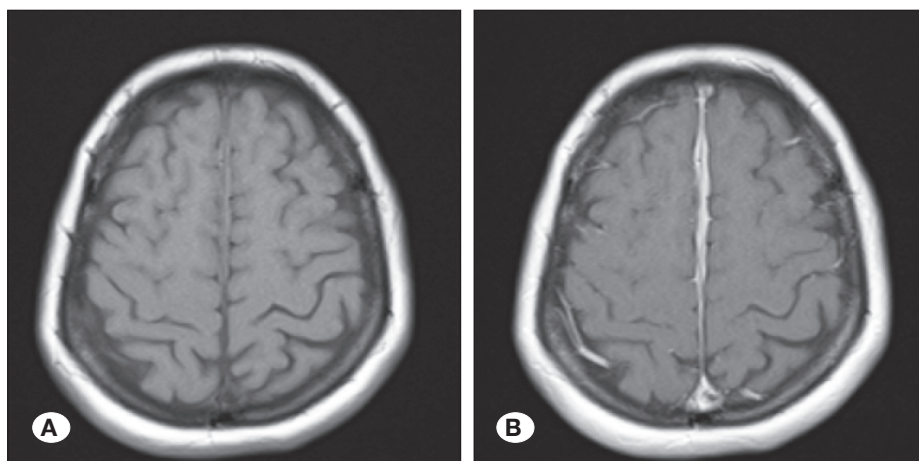


Figure 2: Post-operative third-month axial gadolinium-enhanced (A), and non-enhanced (B) T1W MR images show that the tumour spontaneously regressed.

Spinal Dural Amyloidoma

A 70-year-old female patient presented with lower back pain, right-sided leg pain, left-sided leg numbness, and left foot paresis lasting for a month. Her medical history and blood tests were normal. Lumbar spinal MRI showed spinal stenosis at the L3-4 level, with a suspected space-occupying lesion on the posterior wall of the spinal canal and the interspinous area. Gadolinium-enhanced MRI showed a contrast-enhancing lesion on the right side of the posterior wall of the spinal canal, possibly involving the dura (Figure 4). Surgery was performed using a classical posterior spinal approach. Bilateral L3 hemilaminectomies were performed, the ligamentum flavum was excised, and neural tissues were decompressed. During the surgery, the ligamentum flavum appeared normal except for hypertrophy and increased calcification. Following the flavectomy, the lesion was observed to be associated

with the spinal dura. Tissue samples were obtained from the interspinous and supraspinous ligaments, and these, along with the excised ligamentum flavum, were sent for histopathological examination.

After the surgery, the patient's pain was relieved, and the paresis remained stable. Pathological examination revealed amyloidoma. Immunohistochemical analysis showed kappa light chain positivity, but amyloid-A and lambda were negative. Congo red and crystal violet histochemical stains were positive (Figure 5).

The patient was referred to the haematology department for systemic amyloidosis evaluation. No systemic amyloidosis was detected during follow-up, and no recurrence was noted on control imaging (Figure 6).

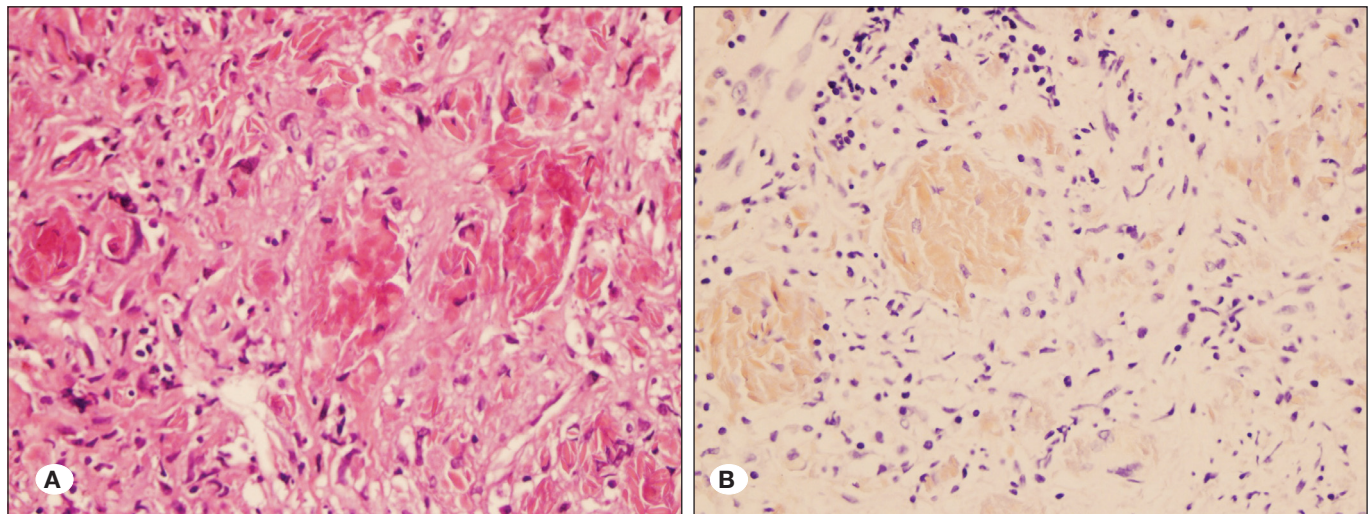


Figure 3: The accumulation of pink amorphous material in haematoxylin and eosin sections (200x) (A). Positively stained areas in Congo red-based histochemical examination (200x) (B).

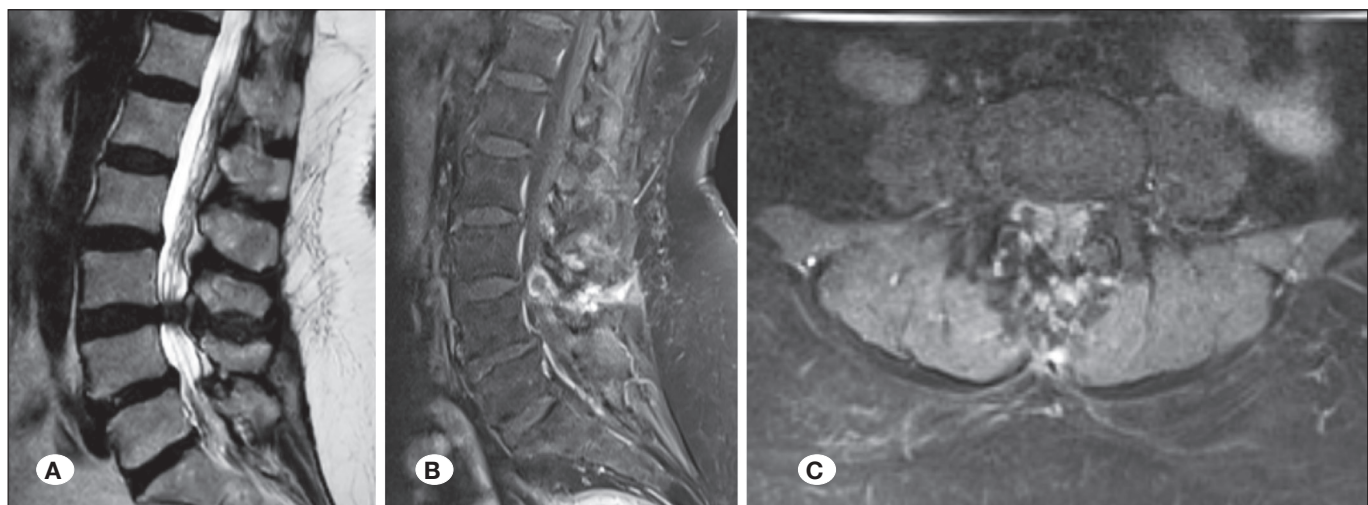


Figure 4: Preoperative T2W and gadolinium-enhanced T1W MR images show lumbar stenosis at the L3-4 level with contrast enhancement on the right side of the posterior wall of the spinal canal (A, B, C).

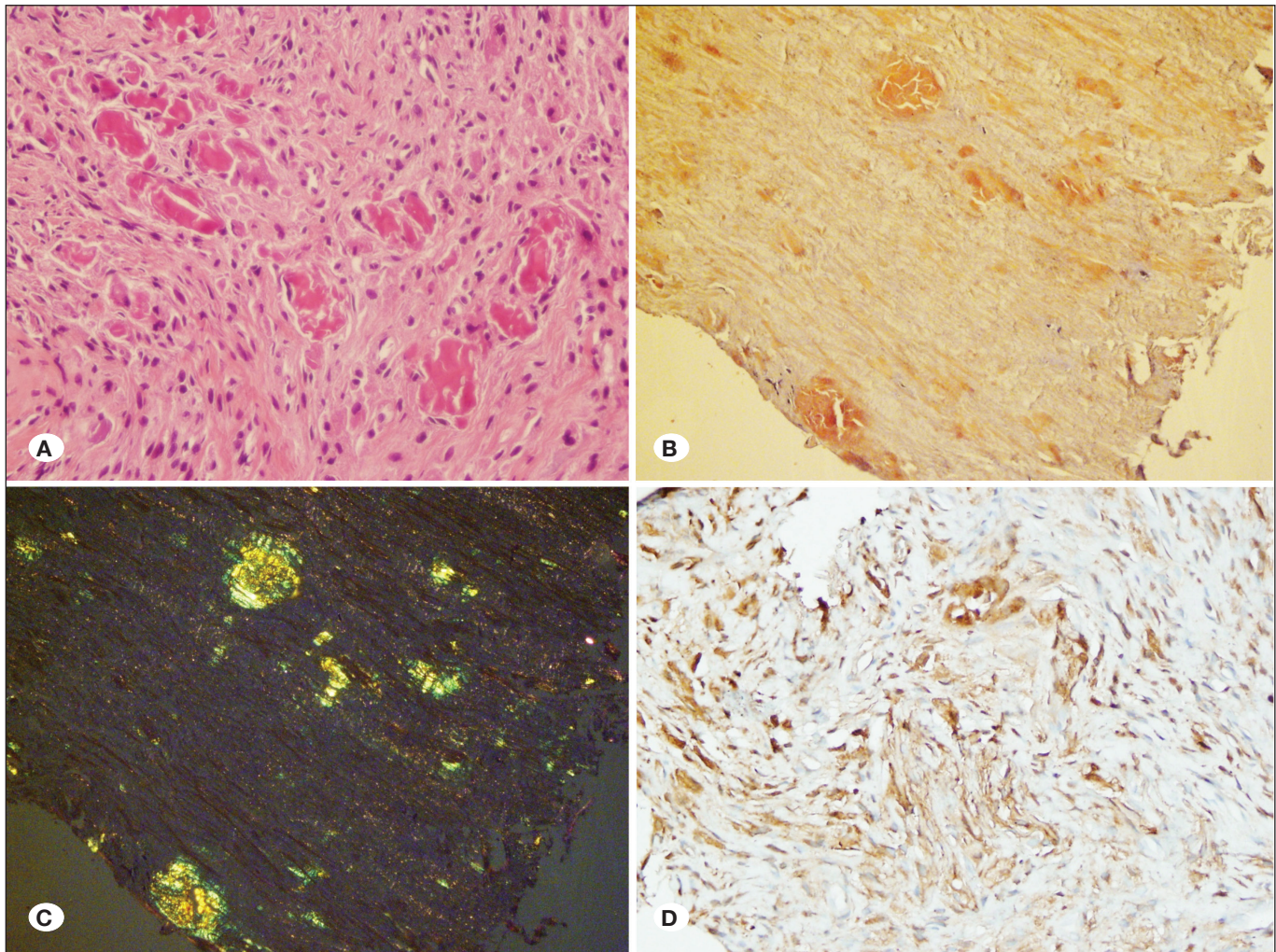


Figure 5: Accumulation of pink amorphous material in haematoxylin-eosin sections **(A)** (200x). Congo red-based histochemical examination shows positively stained areas **(B)** (100x), and an apple-green appearance under polarised light **(C)** (100x). Kappa light chain immunopositivity **(D)** (100x).

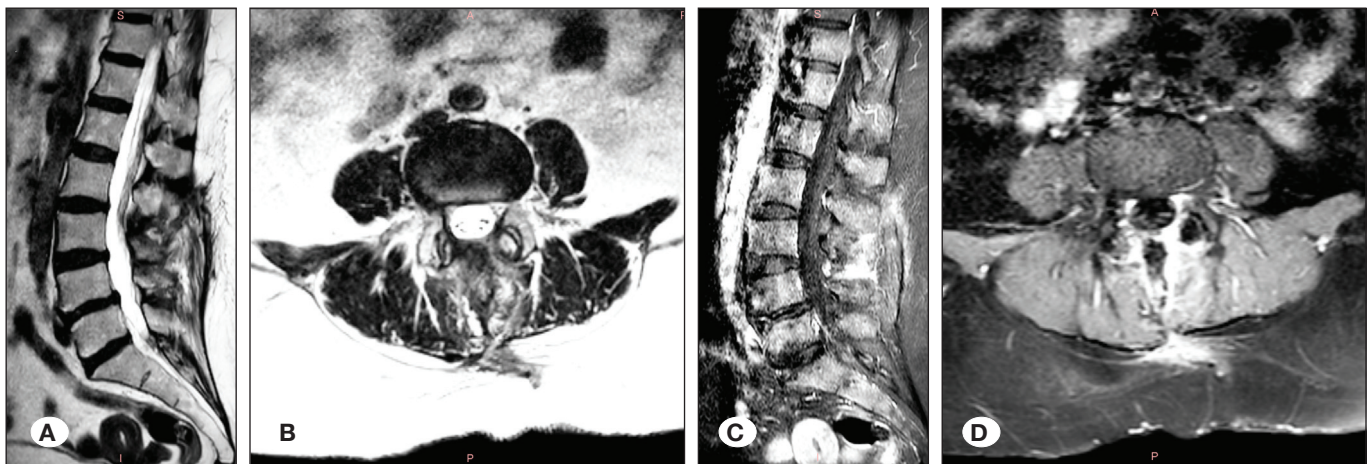


Figure 6: Postoperative T2W sagittal and axial **(A, B)**, and gadolinium-enhanced T1W MR images **(C, D)** show that the contrast-enhancing lesion disappeared and the neural tissues were decompressed.

■ DISCUSSION

The first patient presented with a homogeneously contrast-enhancing extra-axial mass on each side of the falx cerebri. Based on the radiological features, meningioma, lymphoma, plasmacytoma, and subdural hematoma, albeit less likely, were considered. Due to the lesion's location and elongated shape, a biopsy was recommended instead of total excision. Given the rare pathology, the patient was referred to the haematology department. Although cerebral amyloidoma is rare, it can be easily mistaken for more common space-occupying lesions. This case highlights the importance of considering biopsy rather than rushing into total excision when the pathology is uncertain.

The second patient presented with spinal stenosis caused by a contrast-enhancing lesion located in the spinal canal that initially appeared to be an extramedullary tumour due to its contrast-enhancing characteristics. Successful decompression was achieved. Pathological examination revealed amyloidoma, which had not been considered initially before the surgery. No systemic amyloidosis was detected during follow-up, and there was no recurrence.

■ CONCLUSION

Rare cases of amyloidoma located in structures covering the CNS, such as the falx cerebri and spinal dura, are presented and discussed. These lesions can easily be mistaken for other extra-axial and extramedullary space-occupying lesions of the CNS, such as meningiomas, high-grade gliomas, CNS lymphomas, or metastases. In these cases, the lesions appeared as contrast-enhancing masses and were initially considered tumours. Rare pathologies like amyloidoma should be considered, and decisions regarding surgical excision or biopsy should be made based on the patient's condition. A multidisciplinary approach is essential once the pathology is confirmed.

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AUTHORSHIP CONTRIBUTION

Study conception and design: IA, ASK

Data collection: MG, ASK

Analysis and interpretation of results: ASK, GGU, MG

Draft manuscript preparation: SYKO, ASK

Critical revision of the article: IA, ASK, EE

Other (study supervision, fundings, materials, etc.): GGU

All authors (IA, ASK, MG, SYKO, GGU, EE) reviewed the results and approved the final version of the manuscript.

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