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A Rare Intradural Extramedullary Calcifying Pseudoneoplasm of the Spine Presenting with Radiculopathy

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Authors' contributions

This work was carried out in collaboration between all authors. Study conception and delineation by authors KR and JR. The manuscript was composed by authors KR and JR. Authors KR, DS and JR conducted critical reviews of the manuscript. The study was supervised by authors DS and JR. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Aims: Calcifying pseudoneoplasms of the neuroaxis (CPN) are rare, poorly understood lesions of the central nervous system that pose a diagnostic challenge because they mimic the more common calcified lesions of the neuroaxis. We highlight the relevant clinical presentation as well as radiological and histopathological features unique to intraspinal CPNs.
Presentation of Case: We present the case of a 44-year-old Hispanic male with lumbar radiculopathy, radiological features of an indolent, intradural extramedullary mass, and a histopathological evaluation consistent with CPN. The patient underwent successful surgical resection and remained neurologically intact at long-term follow-up.
Discussion: Epidural CPNs have been described in the literature. However, intradural CPNs are

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exceedingly rare lesions, and as a result, are not routinely included in the differential diagnosis of calcified, intraspinal lesions. Although there are currently no consensus guidelines for the diagnosis and management of intraspinal CPNs, understanding the clinical presentation and radiological features of these lesions is crucial for spine surgeons and neurosurgeons because surgical resection may offer a cure.

Conclusion: Calcifying pseudoneoplasms may present as intradural abnormalities that mimic more prevalent lesions such as meningiomas. Surgical resection should be considered as first-line treatment because it is associated with low morbidity and may be potentially curative.

Keywords: Calcifying pseudoneoplasm; fibro-osseous lesion; intradural calcified mass; radiculopathy.

1. INTRODUCTION

Calcifying pseudoneoplasms of the neuroaxis (CPN) are rare lesions of the nervous system that have been reported previously as fibro-osseous lesions and brain stones [1-6]. Experts agree that CPN is a reactive process given its slow growth rate and the inflammatory features found on histological examination [1,3,7]. Since 1978 when Rhodes et al. [8] first described CPN, there have been only 40 descriptions of this entity in the literature with the preponderance of lesions located in the skull base or in the spinal epidural compartment [9]. The common presenting neurological symptom in cranial CPN is headache and associated cranial neuropathies while in spinal CPN it is axial back pain with or without myeloradiculopathy. Although there have been reports of local recurrence as well as peri-lesional edema suggestive of more aggressive pathophysiology, no distant metastasis has ever been documented and gross total resection is still believed to be curative [1,10-12].

2. PRESENTATION OF CASE

2.1 History

A 44-year-old, right-handed Hispanic male presented to the outpatient clinic with three years of progressively worsening lower back pain with radiation into the left anterior thigh and left lateral calf. Physical examination was normal, except that left straight leg-raise testing was positive at 30 degrees with radicular symptoms in the L4 distribution.

Non-contrast computed tomography (CT) scan revealed a round, calcified lesion in the left paramedian central canal at the level of the L4 vertebral body (Fig. 1a). Over a three-year period the mass increased from 9 x 9 x 9 mm to 12 x 10 x 10 mm (Fig. 1b). Magnetic resonance imaging (MRI) scan revealed an 18 x 13 x 11 mm mass with low signal attenuation on T1- and T2-weighted images posterior to the L4 vertebral body (Fig. 1c) causing significant compression of the exiting L4 nerve root and displacement of the traversing L5 nerve root (Fig. 1d). There was a nodular, albeit limited, pattern of enhancement (Figs. 1e and f). There was no evidence of surrounding edema or bony changes. Differential diagnoses were broad and included a calcified and sequestered herniated disk, calcified hematoma, calcified synovial cyst, dural-based metastasis, loculated abscess, calcified meningioma schwannoma, tuberculosis, or sarcoidosis.

2.2 Operative Technique

Given the progressive symptomatology and the need for tissue diagnosis, surgical decompression was offered. A standard L4 laminectomy was performed. The L4 and L5 nerve roots were displaced laterally by the calcified mass towards the patient's left side. The calcified tumor was friable and fragmented easily as it was dissected off the adherent nerve roots. Intraoperative pathology consultation revealed a stromal fibrocollagenous-type tumor with extensive calcification and a capsule of meningoepithelial cells without evidence of malignancy. Cultures of the calcified mass and cerebrospinal fluid were obtained intraoperatively but ultimately proved to be negative. The patient had complete resolution of his L4 radiculopathy at discharge and remained ambulatory and asymptomatic at 48 months.

2.3 Histopathology

Gross analysis of the surgical specimen revealed a lobular, friable, gray-to-white and somewhat gritty mass with finely nodular, discrete islands of calcification. Microscopically, the lesion consisted of a sparse to moderately cellular, richly vascular, fibrocollagenous component within which there were extensive confluent zones of

collagen degeneration. These zones were calcified and appeared amorphous at the center. At their periphery, mineralization could be seen in degenerating collagen bundles continuous with surrounding viable stromal collagen in a radial fashion (Fig. 2a). At the interface between degenerating calcified collagen and viable stromal components, there were numerous epithelioid cells (Fig. 2b), which were immunoreactive with epithelial membrane antigen (EMA), a meningothelial cell marker (Fig. 2c). In some areas, the stromal component had undergone bony metaplasia. The stromal component was non-reactive with CD34, a vascular marker. Focally there was also sparse mononuclear leucocytic infiltrate (Fig. 2d).

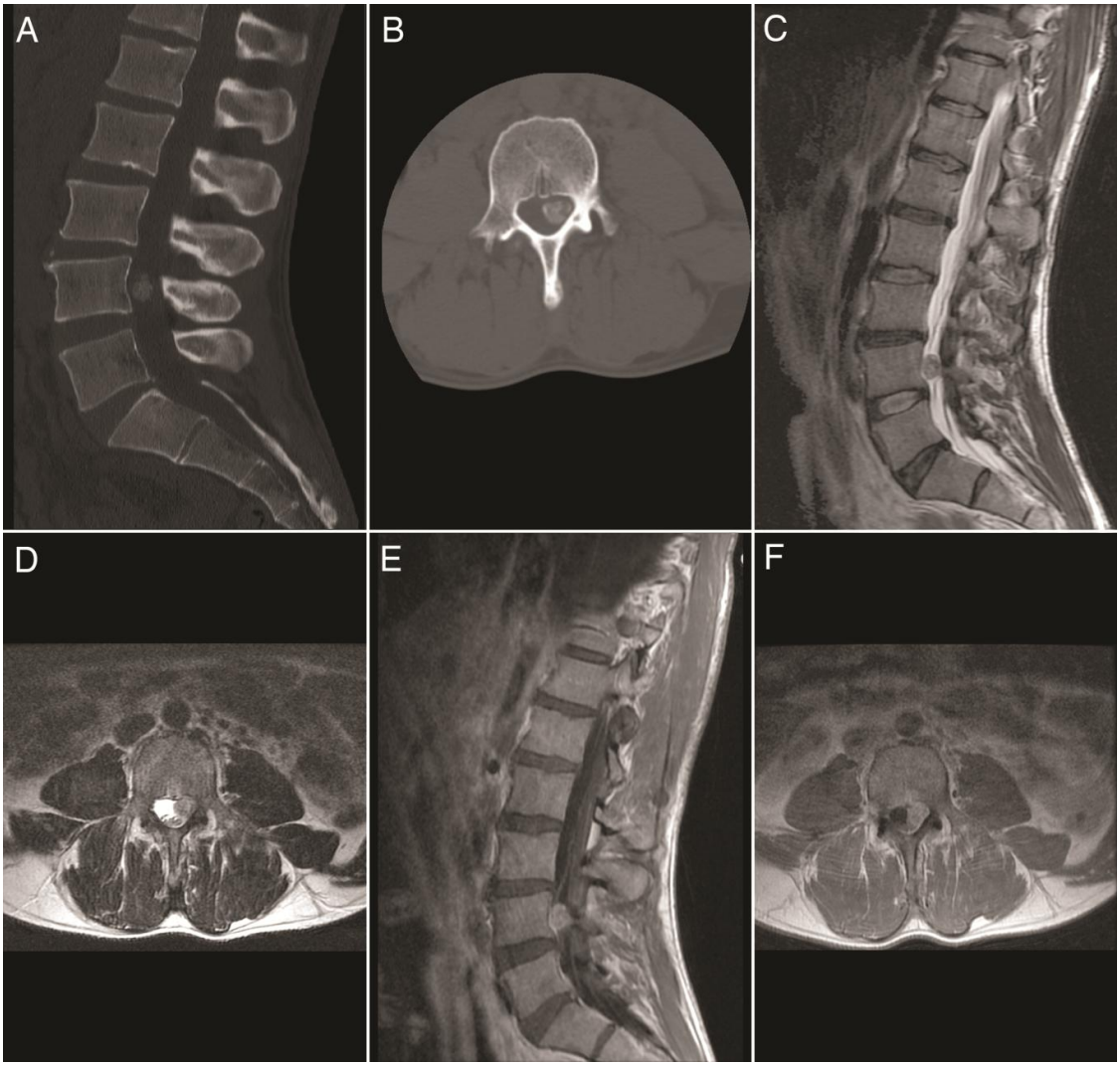

Fig. 1. Sagittal (A) and axial (B) non-contrasted computed tomography (NCCT) showing a round, calcified mass in the central canal behind the L4 vertebral body without any bony changes. Sagittal T-2 weighted magnetic resonance imaging (MRI) showing a mass behind the L4 vertebral body with low signal attenuation (C). An axial T-2 weighted MRI showing a mass with low signal attenuation abutting the left L4 pedicle and compressing the existing L4 and the traversing L5 nerve roots (D). A gadolinium-enhanced sagittal MRI showing a nodular enhancing mass posterior to the L4 vertebral body (E). A gadolinium-enhanced axial MRI showing a mass in the central canal with nodular enhancement (F)

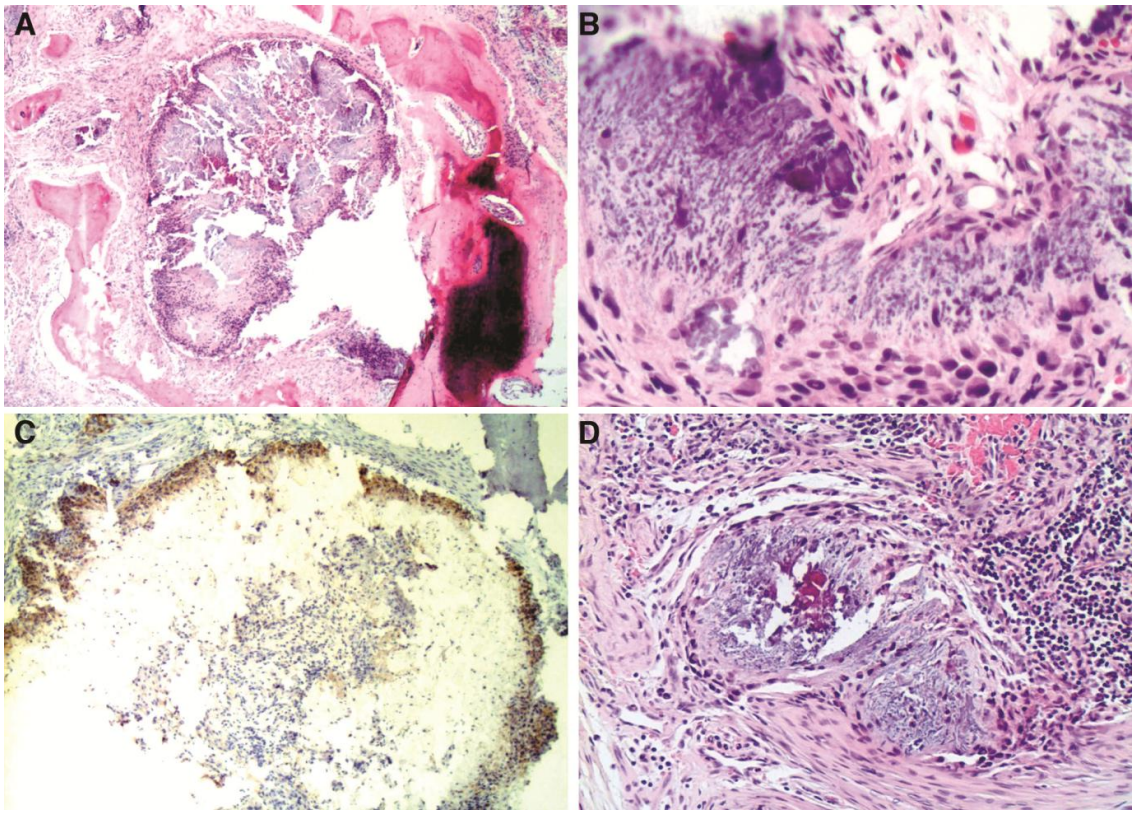

Fig. 2. A 2x H&E stain of CPN components: 1) fibrovascular, stromal lesion; 2) nodular collagen degeneration with central mineralization; 3) peripheral rim of epithelioid cells; and 4) bone metaplasia (A). A 20X H&E stain detailing periphery of nodular lesion with ropey, mineralized, degenerating collagen subjacent to bundles of epithelioid cells (B). An 8X EMA immunoperoxidase stain showing positive reaction of epithelioid cells at the periphery of the mineralized, degenerating collagen, indicating their meningiothelial nature (C). A 10x H&E stain of one nodular lesion depicting the fibrovascular stroma, mineralized and degenerated collagen, peripheral epithelioid cells and inflammation (D)

3. DISCUSSION

CPNS are rare and poorly understood lesions of the central nervous system. They pose a complex diagnostic challenge to neuro- and spine surgeons because their clinical presentation and radiographic features are nonspecific and closely mimic those of more common neuraxial lesions [13]. Since Rhodes et al. [8] described the first case in 1978 there have been only 40 CPN cases reported in the literature with the majority localized to the skull-base region or the extradural, intraspinal compartment [9,11,12,14,15]. We present a case of an intradural, extramedullary CPN causing mass effect and radiculopathy.

CPNs are termed fibro-osseous lesions based on their non-neoplastic histologic features. Our case has the morphological features characteristic of CPN: 1) a richly vascular, variably cellular, stromal component; 2) nodular zones of collagen degeneration with islands of calcification; 3) palisading epithelioid, EMA-positive cells at the periphery of degenerated, calcified stroma; and 4) bone metaplasia. These features suggest that CPNs are pseudoneoplastic lesions with a primary fibrocollagenous vascular component that undergoes focal degeneration with dystrophic calcification, associated meningothelial reactive proliferation, bone metaplasia, and a variable inflammatory response. The meningothelial cells are thought to originate from the pluripotent, ubiquitous, arachnoid cap cells of the leptomeninges. The triggering mechanism remains unclear.

Importantly, the presence of the four aforementioned morphological features is highly variable, and may account for the inconsistent MRI characteristics of CPN that make radiographic diagnosis so challenging. Most CPNs are circumscribed lesions with variable T1- and T2-weighted signal characteristics depending on the composition of the CPN matrix [4,16]. However, low signal attenuation on T1- and T2-weighted images, a characteristic of any calcified lesion, with a nodular pattern of enhancement indicative of a vascular stromal matrix, should raise suspicion of an indolent, calcified lesion such as CPN, meningioma, or even meningioangiomatosis.

No natural history studies on CPN have been published to date, therefore, our understanding of this entity with variably complex histopathology and dubious radiographic characteristics has been limited to small case series and case reports. Moreover, there are no consensus guidelines for the management of CPNs. The current CPN treatment paradigm is gleaned from small case series that have advocated surgical resection, though it is unclear whether surgical resection may provide long-term benefits to patients as compared to expectant management [9,12,15]. In our case, resection was indicated because of the patient's progressively neurological deterioration. It is noteworthy that complete resection may not be feasible in cases where the tumor is densely adherent to surrounding eloquent structures. Local recurrence is rare and has only been reported in cases of incomplete resection [9].

4. CONCLUSION

Spinal CPNs are rare, benign lesions that may present with nonspecific clinical findings such as radiculopathy or myelopathy. These lesions pose a diagnostic challenge by mimicking more common lesions of the central nervous system. CPN should be considered in the differential diagnosis when a well-circumscribed, heavily calcified mass encountered on CT is found to have low signal attenuation on T1- and T2-weighted images with a nodular pattern of gadolinium enhancement. In symptomatic patients, surgical resection should be considered because CPNs are curable and have a low rate of recurrence.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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