Simple Partial Seizures in a 70-Year-Old Female

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The patient's episodes likely represent simple partial seizures with leftward eye deviation as an initial event. The seizures localize to the right frontal lobe with evidence of involvement of the right eye fields (leftward eye deviation) and possibly right Rolandic cortex (lip/chin quivering and dysarthria). A right frontal lesion is also supported by the isolated finding of mild weakness of left finger extension.

The chronicity of the seizures suggests a benign and non-infectious process. It is noteworthy that the seizures began near to the time of a motor vehicle accident although no overt head trauma was evident at that time.

My differential diagnosis for this presentation would include a low grade neoplasm, namely a meningioma. Vascular possibilities would include an infarct or vascular malformation. Less likely explanations would include a traumatic cortical scar.

This calcified mass is most suspicious for a vascular malformation, especially a cavernoma, although it does not have the typical “popcorn” appearance of this entity. A simple infarct or traumatic scar are not tenable considerations. I would recommend craniotomy for resection, definitive diagnosis and treatment.

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Preoperative diagnostic considerations were numerous but favoured a mid- to high-grade glioma or metastasis, based on heterogeneous enhancement and abundant perilesional edema. There was no evidence of a primary systemic lesion on preoperative investigations. After discussion of surgical options, the patient was taken to the operating room for craniotomy and resection of the lesion.

Intraoperatively, the cortex overlying the lesion had a pale grey hue, deep to which a 3 x 2 x 2 cm nodular lesion of stony consistency was encountered. The lesion and adjacent parenchyma were friable and bled easily but a clear plane was found on all aspects.

The patient was neurologically intact post-operatively and had no seizure activity. She was continued on Dilantin, placed on a tapering dose of dexamethasone and discharged home on post-operative Day 3.

**DISCUSSION**

Fibro-osseous lesions, also known as calcifying pseudoneoplasms, are uncommon lesions of the neuraxis with 43 cases reported in the literature (Table). The initial description of this entity was contributed by Rhodes and Davis who described it as...
an unusual fibro-osseous component in intracranial lesions and proposed a metaplastic process as its origin.\textsuperscript{1}

**Epidemiology**

Cases have been reported in patients ranging in age from 6 to 83 years with a slight male predominance (27 males: 16 females). Twenty-seven cases were intracranial, of which 15 were intra-axial and 12 extra-axial (including one intraventricular). Sixteen spinal cases have been reported, eight in the cervical spine and four in each of the thoracic and lumbar regions. Fourteen spinal lesions were extra-dural within the spinal canal, one was intra-dural and one was intraosseous arising from the body of C2 (Table).

**Clinical Presentation**

The clinical presentation is largely determined by location, however several (11 of 43) were neurologically silent. Seven of the latter cases were discovered at autopsy (all intracranial) and four were incidental findings on computed tomogram (CT) scans\textsuperscript{1-4}. Of symptomatic lesions, 7 of 15 intracranial cases presented with seizures. The remainder presented with headache or focal neurological deficit. The lesions predominantly presented with neck or back pain, however radiculopathy and myelopathy have also been reported\textsuperscript{5-9}. All cases had an insidious onset with duration of symptoms ranging from months to years (Table).

**Imaging**

The most consistent imaging feature is hyperdensity on unenhanced CT scans, signifying the presence of calcification. In a series of six fibro-osseous lesions evaluated by MRI, Aiken et al found hypointensity on T1 and T2 weighted images in all cases, with a variable enhancement pattern; two had linear enhancement, two had ring-enhancement, one enhanced in a homogeneous pattern and one did not enhance\textsuperscript{2}. This prompted the authors to conclude that fibro-osseous lesions should be considered in the differential diagnosis when a heavily calcified lesion is found on CT with hypointensity on T1-weighted and T2-weighted images, minimal linear rim or serpiginous internal enhancement, and limited to no edema\textsuperscript{2,3}. The present case was unusual for abundant perilesional edema but imaging features were otherwise similar to previous reports. Differences observed on imaging likely reflect differences in lesion composition, as described by Bertoni et al\textsuperscript{4}.

**Gross, Microscopic and Ultrastructural Features**

Fibro-osseous pseudotumours are well-circumscribed, stony hard, somewhat granular masses that can measure up to 10 cm in diameter. Most are single discrete masses, although multiple clustered lesions with extensive reactive changes in surrounding tissues have been reported\textsuperscript{4}.

The striking microscopic feature is the presence of focal amorphous nodules of chondromyxoid matrix surrounded by palisading spindle and/or epithelioid cells which express epithelial membrane antigen. The variably calcified and ossified matrix is composed of coarse fibrillar material in linear or anastomosing patterns. Ghost cells may be present in the lesion’s core. Cellular atypia and mitotic activity are absent or minimal. Chronic gliosis in the surrounding parenchyma may give rise to Rosenthal fibre formation. Changes reminiscent of meningioangiomatosis have been reported in adjacent cortex\textsuperscript{4}, raising a tenuous association with neurofibromatosis\textsuperscript{9,10}.

Ultrastructural examination of central hypocellular areas reveals masses of electron-dense amorphous material. These masses are largely composed of collagen\textsuperscript{11} and other fine fibrillary elements. Palisading cells at the periphery display ultrastructural features of fibroblasts including abundant intracytoplasmic filaments, prominent rough endoplasmic reticulum and no junctional complexes. Extracellular basal lamina-like material may be present and may represent the residuum of degenerated vascular channels\textsuperscript{5}.

**Nature/Origin**

Given the favourable clinical outcomes and morphological features, most authors have proposed a non-neoplastic proliferative or reactive etiology with which we concur. Other proposed explanations include an unusual expression of tumour calcnosis or abortive membranous bone formation, with osseous metaplasia. Various cells of origin have been proposed including: i) arachnoid cap cells; supported by a common location of this lesion in the CNS (i.e., dural or leptomeningeal) and epithelial membrane antigen or vimentin immunoreactivity, ii) fibroblasts or other mesenchymal cells; supported by ultrastructural studies and examples arising outside the neuraxis, and, iii) astrocytes; supported by glial fibrillary acidic protein immunoreactivity among palisading cells in a single case\textsuperscript{9}. In one report, a concurrent calcifying pseudoneoplasm and ependymoma were described\textsuperscript{12}. In the latter case the ependymoma was low-grade and displayed prominent reactive changes in the form of piloid giosis at its periphery. The latter case also lent support to the suggestion that calcifying pseudoneoplasms may be part of an exuberant reaction to an underlying pathologic process, such as inflammation or neoplasia.

**Differential Diagnosis and Treatment**

Heavy lesional calcification engenders a wide differential diagnosis including chronic inflammatory and infectious entities, granulomatous lesions, low grade tumours (pilocytic astrocytoma, gangliogioma), chordoma, chondrosarcoma, chondroblastoma, menigioma, longstanding intracerebral hematomas, aneurysms and vascular malformations\textsuperscript{13}. In most cases the imaging is not conclusive, necessitating tissue sampling for definitive diagnosis.

The recognition of this distinctive pseudotumour has practical importance in the avoidance of aggressive diagnostic, surgical or therapeutic measures. Excision appears to be curative with only one reported case treated by partial excision that recurred three years later\textsuperscript{4}. In all other cases of gross total or subtotal resection, patients were asymptomatic post-operatively with no evidence of recurrence or progression on follow-up imaging. Morbidity appears to be primarily associated with lesion location\textsuperscript{4}.

**CONCLUSION**

At six-week follow-up, the patient was neurologically intact with no further seizure activity. Three months after craniotomy, MRI examinations revealed no definite residual or recurrent tumour.
<table>
<thead>
<tr>
<th>Reference</th>
<th>Pt 1</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Presentation</th>
<th>Calcification</th>
<th>Multiplicity</th>
<th>Therapy</th>
<th>Follow-up</th>
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<tbody>
<tr>
<td>Jun et al. 1984</td>
<td>85</td>
<td>58</td>
<td>M</td>
<td>Intra-axial, Corpus callosum</td>
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<td>Garen et al. 1989</td>
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<td>44</td>
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<td>Extra-axial, Dura of Meckel’s cave</td>
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<td></td>
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<td>N/A</td>
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<td>Total removal</td>
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</table>

N/A=Not available, NR=No Recurrence, D/D=Degenerative Joint Disease, H/A=Headache, N/V=nausea dnd vomiting, CN=cranial nerve, F/U=follow-up, CV A=cerebrovascular accident.
This case demonstrated many typical clinical, radiographic and histopathological features of a fibro-osseous pseudotumour. The present case was atypical for the presence of abundant perilesional edema on imaging. Although uncommon, awareness of this entity and its inclusion in the differential diagnosis of long-standing calcified lesions along the craniospinal axis has practical importance in directing optimal investigations and treatment.

REFERENCES