32 year-old man with Olfactory Hallucinations and Paresthesias

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Presentation of Case

- 32 year-old man of African and Caribbean ancestry, born in the United States
- Olfactory hallucinations
- Intermittent numbness and paresthesias on the left side
- Dental caries
- Taking levetiracetam and dextroamphetamine
- No systemic symptoms
- Mass in the right temporal lobe
Differential Diagnosis of Mass Lesion

Olfactory hallucinations suggest focal seizures in the uncus

Lesion may be neoplastic or nonneoplastic in origin

Imaging studies were inconclusive
- Enhancing mass located in the medial right temporal lobe (arrows)
- Leptomeningeal foci along right basis points and medulla oblongata (arrowheads)
Differential Diagnosis of Mass Lesion

Imaging studies showed calcification

Long list of tumors that cause calcification, nonneoplastic lesions such as tuberculous and sarcoid granulomas may also calcify

Prepontine meningeal enhancing lesion indicative of inflammatory process, argues against neoplasm

Noncontiguous meningeal disease suggests granuloma

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### Table 1. Brain Tumors with Calcification

<table>
<thead>
<tr>
<th>Intraaxial</th>
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</thead>
<tbody>
<tr>
<td>Low-grade astrocytoma (which is diffuse, linear, punctate, multifocal, and can follow white-matter tracts)</td>
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<tr>
<td>Subependymal giant-cell astrocytoma (which is manifested by chunks or nodules)</td>
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<tr>
<td>Pilocytic astrocytoma</td>
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<tr>
<td>Other astrocytoma and glioblastoma multiforme (rare calcification)</td>
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<tr>
<td>Oligodendroglioma (which is central, peripheral, punctate, ribbonlike, and often located in the walls of tumor vessels; 90% calcification)</td>
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<tr>
<td>Medulloblastoma (which is clump-like or nodular; 90% calcification)</td>
</tr>
<tr>
<td>Gangliocytoma (&lt;50% calcification)</td>
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<tr>
<td>Ganglioglioma (40% calcification)</td>
</tr>
<tr>
<td>Dysembryoplastic neuroepithelial tumor</td>
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<tr>
<td>Metastasis, such as osteosarcoma or breast carcinoma (rare calcification)</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Extraxial</th>
</tr>
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<tbody>
<tr>
<td>Meningioma (which is focal, diffuse, coarse, sandlike, and can have a rim around the periphery; 20-60% calcification)</td>
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<tr>
<td>Pineal tumor</td>
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<tr>
<td>Craniopharyngioma (30-40% calcification)</td>
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<tr>
<td>Dermoid or epidermoid tumor</td>
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<tr>
<td>Pericapsular lipoma</td>
</tr>
<tr>
<td>Intraventricular epidymyoma</td>
</tr>
<tr>
<td>Choroid plexus papilloma or carcinoma</td>
</tr>
<tr>
<td>Central neurocytoma</td>
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* The percentage of calcification indicates the percentage of an individual tumor that is usually calcified.

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### Table 2. Causes of Intracranial Lesions That Mimic Neoplasms

<table>
<thead>
<tr>
<th>Infectious</th>
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</thead>
<tbody>
<tr>
<td>Fungal: aspergillus, mucormycosis, cryptococcus, cladosporium, actinomycosis, blastomycosis, candida, coccioidoides, and nocardia</td>
</tr>
<tr>
<td>Bacterial: spirochete infections such as syphilis and borreliosis; pyogenic infections such as brain abscess</td>
</tr>
<tr>
<td>Parasitic: amebiasis, cisticercosis, schistosomiasis, and progressive multifocal leukoencephalopathy</td>
</tr>
<tr>
<td>Noninfectious or immune-mediated</td>
</tr>
<tr>
<td>Sarcoidosis</td>
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<tr>
<td>Inflammatory pseudotumor</td>
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<tr>
<td>Behçet’s disease</td>
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<tr>
<td>Lupus erythematosus</td>
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<tr>
<td>Sjögren’s syndrome</td>
</tr>
<tr>
<td>Granulomatous angitis</td>
</tr>
<tr>
<td>Demyelinating</td>
</tr>
<tr>
<td>Tumefactive multiple sclerosis</td>
</tr>
<tr>
<td>Vascular</td>
</tr>
<tr>
<td>Infarction</td>
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<tr>
<td>Vasculitis</td>
</tr>
<tr>
<td>Amyloid</td>
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<tr>
<td>Amyloidoma</td>
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<tr>
<td>Amyloid angiopathy</td>
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</tbody>
</table>
Pathological Discussion

- Performed open right temporal craniotomy & anterior temporal lobectomy
- Abundant granuloma forming lesions
- Consistent with sarcoidosis of brain (neurosarcoidosis)
Differential Diagnosis of Mass Lesion

Calcification and prepontine lesion point towards diagnosis of cerebral sarcoidosis

Lack of sicca syndrome?

- Can be caused by sarcoidosis or Sjögren’s syndrome
- Symptoms include dry mouth (xerostomia) and dry eyes (xerophthalmia)
- Does not rule out diagnosis of sarcoidosis or Sjögren’s syndrome
Sarcoidosis

- Dysregulated immune response that leads to the growth of granulomas, likely in response to an inhaled substance such as insecticide\(^2\)
Sarcoidosis

- Dysregulated immune response that leads to the growth of granulomas, likely in response to an inhaled substance such as insecticide
- Caused by a combination of environmental and genetic factors
  - ANXA11 detectable in human B-cell exosomes, induce immune responses or tolerances

Chromosome 5, ANXA11 gene location indicated, credit: Genome Decoration Page/NCBI
Sarcoidosis

- Dysregulated immune response that leads to the growth of granulomas, likely in response to an inhaled substance
- Caused by a combination of environmental and genetic factors
- Three times more common and more likely to be chronic and fatal in African Americans than European Americans\(^3\)
Sarcoidosis

- Dysregulated immune response that leads to the growth of granulomas, likely in response to an inhaled substance
- Caused by a combination of environmental and genetic factors
- Three times more common and more likely to be chronic and fatal in African Americans than European Americans
- Does not usually affect the Central Nervous System
More Diagnostic Tests

- CT
- Lymph nodes
- Right Hilia
Management of Neurosarcoidosis

- Immunosuppressive drugs
- Glucocorticoids vs Biologics
Management of Neurosarcoidosis

- Immunosuppressive drugs
- Glucocorticoids vs Biologics
- Glucocorticoids suppress cytokines in granulomas\textsuperscript{5}
  - Prednisone
Management of Neurosarcoidosis

- **Immunosuppressive drugs**
- **Glucocorticoids vs Biologics**
- **Glucocorticoids suppress cytokines in granulomas**\(^5\)
  - Prednisone
- **Biologics- TNF-\(\alpha\) inhibitors**
  - Suppresses cytokine TNF-\(\alpha\) in granulomas\(^1,6\)
  - Infliximab
Management of Neurosarcoidosis

- Prednisone 80mg
- Levetiracetam 1000mg
Management of Neurosarcoidosis

- Prednisone 80mg
- Levetiracetam 1000mg
- Prednisone 60mg
- Gabapentin 200mg
- Trimethoprim-sulfamethoxazole (pneumonia prophylaxis)
- Calcium and Vitamin D
Management of Neurosarcomiosis

- Prednisone 80mg
- Levetiracetam 1000mg
- Prednisone 60mg
- Gabapentin 200mg
- Trimethoprim-sulfamethoxazole (pneumonia prophylaxis)
- Calcium and Vitamin D
- Methotrexate
- Lamotrigine
Management cont.

- MRI revealed stability is lesion in right temporal lobe
- Spread to brain stem
- Prednisone and Infliximab