Osteolytic Lesions in the Craniofacial Skeleton of a Patient with Extrapulmonary Sarcoidosis

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Case report: A 52 year old African American woman with a history of pulmonary sarcoidosis and non-compliance with her medical management presents to the otolaryngology clinic complaining of nasal obstruction, crusting, and epistaxis. On exam she had significant nasal crusting, friable mucosa with a nodular appearance, non-tender area of erythema of the anterior hard palate, and cutaneous lesions consistent with sarcoidosis. A non-contrast maxillofacial CT revealed osteolytic lesions in the pre-maxilla, greater wing of the sphenoid bone, and the parietal bone. Biopsies were performed of the middle turbinate, the pre-maxilla, and a cutaneous lesion. All biopsies revealed non-caseating granuloma, consistent with sarcoidosis. The patient was referred to rheumatology and aggressive combination medical therapy was initiated.

Clinical Presentation: 10-15% of patients with sarcoidosis will have head and neck involvement. Disease may affect the nose and sinuses, salivary glands, the eye, facial skin, and exhibit otologic manifestations. Involvement of craniofacial bones, as in the patient described above, is very uncommon.

Location of bony lesions: While bone involvement has been described in 3% to 13% of patients with sarcoidosis, it is the small bones of the hands and feet that are most commonly affected. A review of the literature yielded only 38 reports of bony involvement of the craniofacial skeleton.

<table>
<thead>
<tr>
<th>Involved bone</th>
<th>Nasal bones</th>
<th>Maxilla</th>
<th>Mandible</th>
<th>Calvarial bones</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>3</td>
<td>12</td>
<td>11</td>
<td>12</td>
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Radiographic findings: Lytic bone lesions generally appear as rounded, punched out lesions involving cortex and medulla. In an advanced stage destructive lesions may be seen, characterized by fractures of devitalized cortex and sequestrum.

Histology: Biopsy reveals non-caseating granulomas made up of compact epithelioid cells with pale nuclei and multinucleated giant cells. Interspersed fibroblasts and and lymphocytes at the periphery are commonly seen.

Treatment: Management of bone involvement is typically the same as for other manifestations of the disease. Common agents include systemic corticosteroids, methotrexate, azathioprine, antimalarials (chloroquine) as well anti-TNF-α therapies.

Conclusion: Although an uncommon site of intraosseous sarcoidosis, osteolytic lesions can be found in the craniofacial skeleton. Radiographically, these lesions can mimic other diseases, such as malignancy and infection. This case illustrates the importance of understanding head and neck involvement in sarcoidosis and of obtaining tissue diagnosis so that appropriate medical therapy may be initiated.

References: