Osteolytic Lesions of Skull in Pediatric Age Group

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Abstract: Lytic lesions of the calvarium in pediatric age group have a wide range of different etiologies, ranging from normal variants to congenital, traumatic, inflammatory and neoplastic lesions. They manifest as palpable masses, with or without associated pain. Imaging approach frequently begins with CT and MR being complementary and the method of choice for the assessment of bone and associated soft tissue masses. We are hereby reporting two such cases with review of literature.

Keywords: lytic lesion of skull, calvaria, eosinophilic granuloma, fibrous dysplasia.

I. Introduction

Primary skull lesions of skull are estimated to amount for 0.8% of all bone tumors. Dermoid and epidermoid account for 20-60% of all reported tumors with other lesion making up the remainder. Tumors of skull in children present as painless bumps, with benign lesions having long history while malignant lesions are multiple and more aggressive. Local pain and tenderness are less commonly seen. Imaging is advisable in most of the lesions, CT scan is the imaging modality of choice for bony lesions, it allows better evaluation of bony erosion or invasion and involvement of both inner and outer tables. MRI is recommended when intracranial extension is suspected. Definitive diagnosis we are here discussing is two such lesions in pediatric age group.

Case 1

Eight month old male child presented with progressive swelling of parietal region of head since two months as noticed by the parents. Not associated with pain or irritability. No history of constitutional symptoms. On examination child was active, appropriate milestones for his age, local examination showed diffuse bony swellings of parietal bones, non tender. Routine blood investigation was normal.

Legend 1

CT scan of brain with bony windows was done which revealed bilateral parietal bone had osteolytic lesion with ground glass appearance with no involvement of brain parenchyma.
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Legend 2

CT scan of the calvaria showing lytic lesion in left parietal bone and ground glass appearance in right parietal bone. Child was subjected for excisional biopsy, interoparatively there was widening of diploe cavity with pale granulation tissue in the space. Histopathology was suggestive of possibility of fibrous dysplasia. Child is under follow-up and is doing fine.

Case 2

A 10-year-old boy presented our department with a painful swelling over midline of the forehead for 30 days. There was no history of trauma, headache or vomiting. Systemic examination was unremarkable. Local examination revealed a circumscribed swelling of 4*4 cm in diameter in midline in the frontal region which was hard, not mobile and tender. Underlying bone was eroded. The overlying skin appeared normal. Routine hematological and biochemical investigations were within normal limits.

Legend 3

CT scan of brain with bony windows revealed circumscribed osteolytic lesion in the midline of frontal bone with erosion of both outer and inner table with irregular bony edges. Operative the lesion was seen below the scalp, reddish lesion in diploe cavity of the bone with eroded outer and inner table extending up to the dura but not infiltrating it. Curettage of lesion was done. HPE was suggestive of eosinophilic granuloma.

II. Discussion

Osteolytic skull lesions may have many different causes, anatomical variations being responsible for up to 60% of cases. In general population, seven diagnosis include 85% of all causes - by decreasing order, dermo/epidermic cysts, hemangioma, metastasis, multiple myeloma, Langherans histiocytosis, Paget disease of bone, fibrous dysplasia.
In adults, tumoral causes are predominant - metastasis and myeloma [8] - whereas in children, besides congenital defects, dermoid cysts and eosinophilic granuloma are themost frequent diagnosis.[7 benign lesions tend to have well-defined borders with sclerotic margins,a quite predictable location, mostly near the midline, and are usually solitary. On the other hand, those with a permeative appearance, multiple and randomly distributed, are probably solitary. In adults and elderly patients, metastases are by far responsible for the majority of lytic skull lesions, whereas children and young adults present more frequently one of the congenital, inflammatory, traumatic or benign neoplastic conditions mentioned below

Normal Variants
Transcalvarial venous lakes and venous channels
Arachnoid granulations
Enlarged parietal foramina

Congenital
Meningocele CSF lined by meninges
Gliocele CSF lined by glial tissue
Meningoencephalocele CSF and brain
Meningoencephaloceles Cedro CSF, brain and ventricules
Atreticcephalocele Sinus percranii

Traumatic
Skull defects
growing skull fracture or lepto meningeal cyst

Inflammatory
eosinophilic granuloma
sarcoidosis

Infective
Osteomyelitis
Tuberculosis

Benign lesions
fibrous dysplasia
Dermoid
epidermoid cysts
hemangioma
Osteoma
Ossifying fibroma

Malignant
neuroblastoma
osteoblastoma
ewings sarcoma
fibrous sarcoma
angiosarcoma
lymphoma
metastasis

fibrous dysplasia is a benign congenital disorder in which there is osteoblastic dysfunction causing replacement of normal bone with woven bone, lesions have ground glass appearance in CT scan less commonly cystic or sclerotic is also seen this condition is seen in children and young adults.[9]

Eosinophilic granuloma is a rare entity in frontal bone, hence discussed with review of literature Eosinophilic granuloma (EG) is a rare bone tumor, representing less than 1% of all bone tumors. The term “eosinophilic granuloma” was first introduced by Lichtenstein and Jaffe in 1940. Eosinophilic granuloma is the benign form of three clinical variants of Langerhans Cell Histiocytosis; the other two variants are Letterer-Siwe disease and Hand-Schuller-Christian disease. Langerhans Cell Histiocytosis (LCH) is a rare disease with unknown etiology. The incidence of LCH is estimated to be 0.2-0.5 cases per 100,000 per year. Bone is the most frequent site of this disease. It is usually considered to be a disease of childhood. Many patients are 1-15 years old, however the diagnosis frequently is made in adults and many cases of childhood onset progress into adult life.[2] The etiology...
of LCH is unknown and there is continuing debate whether this condition is neoplastic or non neoplastic. Inflammation, autoimmunity and loss of controlled proliferation of Langerhans cells are assumed etiologies.

Table I. The staging system proposed by Greenberger et al.1981

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tbody>
<tr>
<td>I</td>
<td>a) Single monostotic bone lesion</td>
</tr>
<tr>
<td></td>
<td>b) Multiple lesions in one or multiple bone</td>
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<tr>
<td>II</td>
<td>&gt;24 months of age at diagnosis and having one of the following systems involved:</td>
</tr>
<tr>
<td></td>
<td>diabetes insipidus, teeth, gingivae, lymph nodes, skin, mild lung involvement</td>
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<tr>
<td></td>
<td>(i.e., infiltrates seen on chest radiograph without pulmonary symptoms or gross consolidation), focally positive bone marrow</td>
</tr>
<tr>
<td>III</td>
<td>a) Age &lt;24 months at diagnosis with any of the systems involved in stage II</td>
</tr>
<tr>
<td></td>
<td>b) Age &gt;24 months with involvement of liver and/or spleen, massive nodal involvement (nodes &gt; 5 x 5 cm in several sites above or below diaphragm), honeycomb lung (major involvement in all areas with apparent fibrosis), bone marrow packed</td>
</tr>
<tr>
<td>IV</td>
<td>Spleen &gt; 6 cm (palpable below costal margin) and fever &gt;1 month with or without any or all of the above systems involved</td>
</tr>
<tr>
<td>V</td>
<td>&quot;Special&quot; monocytosis in peripheral blood &gt; 20% of differential cell count, in addition to stage III or IV</td>
</tr>
</tbody>
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The staging system of Langerhans cell histiocytosis proposed by Greenberger et al has classified lesions primary limited to the bone as stage 1a or 1b (Table I). About 80% of patients show bone lesions at diagnosis and about 40% of them have the disease limited to the bone.

Table II. Anatomic distribution and frequency of bone lesions in LCH

<table>
<thead>
<tr>
<th>Bone</th>
<th>Frequency</th>
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<tbody>
<tr>
<td>Parietal</td>
<td>17%</td>
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<tr>
<td>Frontal</td>
<td>9.5%</td>
</tr>
<tr>
<td>Occipital</td>
<td>9.5%</td>
</tr>
<tr>
<td>Temporal</td>
<td>7.5%</td>
</tr>
<tr>
<td>Sphenoid</td>
<td>2.8%</td>
</tr>
<tr>
<td>Maxilla</td>
<td>0.9%</td>
</tr>
<tr>
<td>Mandible</td>
<td>13.3%</td>
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</tbody>
</table>

Solitary eosinophilic granuloma of skull is a rare condition, the natural history of which has not been defined completely. Characteristically the patient notices an enlarging tender skull mass for weeks to months; most commonly seen in parietal bone of the skull. Our patient had involvement of frontal bone with a gradual increase in size of swelling. Eosinophilic granuloma of any skull bone can be within diploe may or may not compress brain parenchyma without dural infiltration or it can be projecting into the cranial cavity compressing the brain parenchyma with dural infiltration [3] EG can be asymptomatic or can present as localized pain, tender swelling and fever. Rarely does it present with epidural hematomas, suppression of bone marrow and pathological fracture. Headache, neurological symptoms, chronic mastoiditis and exophthalmos may be seen in cases of skull lesion [4]. Langerhan cells of eosinophilic granuloma express HLA-DR, S-100 and CD1a.

Unlike normal resident dendritic cells, it is the co-expression of CCR6 and CCR7 which allows the langerhan cells to migrate into the tissues that express the relevant chemokine-CCL20 in skin and bone (ligand for CCR6) and CCL19 and 21 in lymphoid organs (ligands for CCR7). Langerhan cells can cause osteolysis by elaboration of interleukin-1 and prostaglandin-E2. Histopathological examination of the lesion provides definitive diagnosis of the condition. The lesions cellular components includepathologic langerhan cells, chronic inflammatory cells and eosinophils. Radiograph and computed tomography shows extensive frontal bone destruction [5] generally accepted treatment of choice for solitary bone lesions, especially for those affecting calvaria, is surgical excision when the lesion is readily accessible. Data of many authors indicate that surgical curettage is a very successful treatment. If necessary, excision is combined with concurrent bone grafting. However some authors report the higher risk of local recurrence after surgery alone.

 Persistence symptoms of disease, or expansion of the lesion after surgical intervention, may respond to the subsequent radiotherapy [1,6]. Eosinophilic granuloma of bone has been reported to resolve after low dose irradiation, intralesional corticosteroids, simple biopsy, subtotal curettage with or without postoperative low dose radiotherapy or chemotherapy and craniectomy with cranioplasty for lesions >4 cm [3]. Systemic evaluation and long term follow-up after any kind of treatment modality is mandatory in every case.

III. Conclusions
Calvarial tumors in childhood age group run from benign to malignant disease, they have to be carefully evaluated with imaging and surgical excision is usually mainstay of treatment in most of lesions.

References

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