Cystic angiomatosis of the skull presenting with extradural pneumocephalus

Case report

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The case is reported of a 40-year-old militiaman who presented 4 days after a blast injury with a huge swelling of the scalp. The bulge was determined to be an air mass collected extradurally via lysis of the air cells of the mastoid bone. Pathological study of the resected bones revealed cystic angiomatosis. The unusual clinical presentation, as well as the rarity of the pathological findings, prompted this report.

KEY WORDS • cystic angiomatosis • pneumocephalus • osteolytic skull lesion • skull • angioma

Cystic angiomatosis of the skeleton is a rare, distinct pathological entity. Osteolysis due to hamartous afflictions is well documented; however, involvement of the calvaria by the cystic variety is very rare. Cystic angiomatosis limited to the skeleton or skull without involvement of soft tissue and viscera has a more favorable prognosis and should be recognized to facilitate proper management. The present report demonstrates a rare case presenting with a massive extradural air mass due to mastoid air-cell penetration. The rare pathological findings and unique clinical presentation are discussed.

Case Report

This 40-year-old militiaman was evaluated in November, 1987, during the Persian Gulf war 4 days after suffering a blast injury. He was alert on presentation, with no report of unconsciousness or a blow to the head. He continued military service until the bulging of his scalp caused a minor ache. Several puncture holes had been made by the battlefront physicians to evacuate the suspected blood under the skin; however, instead of blood, air emerged then reaccumulated in minutes with mild pain and a strange feeling in the patient's throat and left ear. His medical history was unremarkable except for a healed laceration of the scalp sustained in childhood which overlay the bulge.

Examination. Physical and neurological examination was negative except for the large soft mass under scarred but healthy skin (Fig. 1). The mass was fluctuant on palpation, with no bruit audible. Skull x-ray films showed a large radiolucent defect with a honeycomb appearance over the left temporo-parieto-occipital area. The margin was not defined, and no sclerosis, calcification, or new bone formation was present. The osteolytic defect was most striking over the mastoid air cells. Destruction of the outer table was evident in the occipital bone (Fig. 2).

The patient was hospitalized with a tentative diagnosis of chronic osteitis, and another puncture was made to verify the presence of subgaleal air. Upon puncture, the bulging skin collapsed. Defective bone could be felt underneath and caused no apparent pain or discomfort. As before, the air reaccumulated in less than 15 minutes. Laboratory investigations were unremarkable. A 99mTc-medronate methylene diphosphonate (MDP) whole-body bone scintiscan revealed vascular perfusion which disappeared on the delayed scan.

Operation. Upon incision, the skin over the bulge collapsed and defective bone appeared beneath. Cavities of all sizes were present. No periosteum or cortical layer could be identified. Trabeculae, soft to the touch, were the only connections between the defective bone and the soft tissue above. Bleeding was moderate. Resection was performed with ease. The bone could be removed by placing slight pressure upon it. The margin of defect was uneven. Healthy bone could be identified by its consistency and the normal bleeding associated with it.
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while rongeuring the defect. Craniectomy was then extended downward toward the temporal bone. Here the lacunae were even larger and the air cells of the mastoid were obviously enlarged and defective. The lesion was entirely removed, revealing the dura beneath it to be intact. The trabeculae were detached and the cavity was irrigated with 1 liter of saline. The mastoid air cells were covered with pieces of beaten, antibiotic-treated muscle from the thigh.

Pathological Examination. Microscopic slides, prepared from superficial and deep layers of all specimens, showed spongiform bone infiltrated by neoplastic tissue containing an edematous fibroplastic component with minimal collagen fibers. On the background were numerous capillary vessels differing in number and extent in various slices. In places, very dilated capillaries bounded by one row of endothelial cells without anaplastic features were present side by side and in large numbers. The spicules of bone were composed of mature elements accompanied by a hyperosteoblastic reaction and a few osteoclasts. Most of the bone spicules were ossified, but there was some osteoid formation and ossified spicules with osteoid rimming (Fig 3).

Discussion

Benign hemangioma, defined as a lesion consisting of newly formed blood vessels either of capillary or cavernous type, is common in soft tissue, but seldom seen in the skeleton. The incidence varies from 0.85% to 1% of all bone tumors. The site most commonly affected is the vertebral column. Clinical, radiological, and histological features are well documented.3,8

Cystic angiomatosis, previously called hamartous hemolymphangiomatosis is a distinct pathological entity occasionally reported in the skeleton and soft tissue, especially in the spleen. The skeleton alone may be involved, but the condition is usually disseminated.8 The report of calvarial involvement alone or in combination with other bones is quite rare. Boyle1 and Schajowicz and coworkers6,9 have reported three cases each and reviewed the literature. Diagnosis of this clinicopathological picture is recommended for its prognostic significance as well as its pathological interest. In contrast to cases with visceral involvement, a more favorable prognosis is expected when only the skeleton is involved. Spontaneous regression of the cystic lesions has been reported in a few cases.1,9

The present case seems to be unusual in several aspects. The massive osteolytic lesion affecting three bones of the skull was obviously present for some time without any clinical symptom or sign. It became apparent through an unrelated mechanism. The lack of any clinical symptom was emphasized in the patient's his-

Fig. 1. Photograph of the patient 4 days after a blast injury showing his head disfigured by a bulge in the occipital area of the scalp.

Fig. 2. X-ray films, anteroposterior (left) and lateral (right) views, showing a large radiolucent bone defect involving the temporoparieto-occipital bones with a honeycomb appearance, without a margin or calcification.
The "moth-eaten" bone extending to the mastoid area with increasingly larger lacunae seemed to be a chronic lesion. The outer and inner diploe could not be distinguished. The moderate bleeding was probably due to infarction and/or thrombosis. The margin of the lesion was not defined and could only be distinguished by rongeuring the defect and reaching bone which was obviously healthy due to its normal consistency and bleeding, much like the usual osteitis of the skull.

The collection of air between the depressed dura below and the overlying scalp could only be attributed to the obviously large lacunae around the defective mastoid bone. Entrance of air into the cranial cavity has been reported in the literature only once, in an angiofibroma. However, the source of the slight pressure of air expanding the scalp above and depressing the dura below could only be speculated upon by assuming that the air had entered the cyst-like space through a one-way valve created by destruction of the mastoid and sudden opening of mastoid air cells into the middle ear prompted by minor trauma to the patient's skull in a blast injury. It remains a puzzle that the air should reaccumulate in such a short time.

Osteolytic destruction of the calvaria by an angiomatosis is a rare disease. Cystic angiomatosis of the skull affecting three adjacent bones is also rare. These lesions may be more common than expected and may not be associated with any clinical symptom or sign. Considering the relatively few reports available, the documentation of more cases is recommended.

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References