A case of histologically confirmed Paget disease of the orbit produced a lesion that appeared both clinically and histologically similar to a cholesterol granuloma. This case is unique because of the unusual location of the lesion, its presentation in a patient with no other manifestations of Paget disease, and the histological picture produced by the disease.

Paget disease is a chronic disorder of unknown cause that affects the adult skeleton. It is characterized by excessive bone destruction, re-formation, and resorption, resulting in skeletal weakness and abnormal architecture of the diseased bone. About one third of cases are monostotic, affecting only 1 bone or part of a bone. The remaining cases are polyostotic, affecting several bones. The bones most often affected by Paget disease are the pelvis (65%), spine (42%), femurs (37%), and sacrum (30%). The skull is affected in 25% to 65% of cases.

Orbital involvement is rare in Paget disease. It almost always affects the posterior aspects of the greater or lesser wings of the sphenoid bone and anterior clinoid process, causing narrowing of the superior orbital fissure and optic canal, with subsequent proptosis, limitation of eye movement, optic neuropathy, or a combination of these manifestations. We describe a unique case of solitary Paget disease of the orbit characterized by an enlarging painless right supraorbital mass, the histopathologic appearance of which superficially resembled that of an isolated cholesterol granuloma.

REPORT OF A CASE

A 53-year-old man had a 14-year history of drooping of the right eyelid and a 3-month history of a nontender prominent area in the region of the right supraorbital ridge. There was no history of previous trauma to the area, and the patient denied any pain, blurred vision, or double vision. Magnetic resonance imaging showed a 2-cm multicystic bony mass beneath the right frontal lobe that involved the roof and the superior portion of the lateral wall of the right orbit. The mass appeared to contain old blood. The patient was referred to the Orbital Unit of The Johns Hopkins Medical Institutions, Baltimore, Md, for further evaluation.

On examination, the patient's visual acuity was 20/20 OU, color vision was normal, and visual fields were full. Pupils were isocoric and reacted normally to light and near stimulation; there was no relative afferent pupillary defect. The patient was orthophoric in primary position at distance and near; however, the right globe was displaced inferiorly. There was moderate limitation of elevation of the right eye, with the patient developing a right hypotropia on attempted upward gaze. There was no proptosis. Palpation of the right orbit revealed marked enlargement of the right superior and superolateral portions of the bony rim. The skin overlying the bony abnormality was neither warm nor erythematous, and the area itself was nonpainful and nontender. A soft tissue mass could be felt just beneath the orbital rim. The remainder of the examination results were normal.

Standard axial, coronal, and 3-dimensional computed tomographic scans of the orbits revealed a 2.4-cm multicystic le-
sion that expanded and eroded the bone of the right orbital roof and the superior portion of the right lateral orbital wall (Figure 2). The lesion also extended upward against the undersurface of the right frontal lobe and downward into the right orbit.

The patient underwent a frontal craniotomy with an orbitozygomatic extension. The orbital roof was exposed using an extradural approach and limited retraction of the frontal lobe. The contour of the orbital roof was markedly irregular, with areas of superior expansion. As the dura was dissected away from the orbital roof, it became apparent that the dura was markedly thinned in some areas, with brownish-blue bony expansions below these areas. The dura was then opened and the right frontal lobe was elevated, exposing the roof of the right orbit. The contour of the orbital roof was markedly irregular, with areas of superior expansion. Separation of the dura from the abnormal orbital roof exposed a large area of discolored and abnormal bone. A resection of all grossly abnormal bone was performed using a high-speed drill. The periorbita was then separated from the undersurface of the roof and lateral wall of the orbit. Once this was accomplished, it could be seen that the periorbita underlying the area was thinned in some places and absent in others, revealing normal orbital fat. The orbital roof and lateral wall were reconstructed using a split-thickness calvarial graft.

The central lytic portion of the resected lesion contained fibrous tissue with abundant stacks of slitlike clefts that contained dissolved cholesterol crystals (Figure 3, A). Many of the spaces were adjacent to foreign-body–type giant cells. In the central portion of the lesion were numerous foci of residual native bone, which showed a distinct mosaic pattern of remodeling lines (Figure 3, B). Numerous osteoclasts were juxtaposed to these residual islands of bone.

The bone from the rim of the lytic area showed a zonal pattern of

Figure 1. Magnetic resonance imaging results. T1-weighted coronal image shows an irregular mass in the superotemporal portion of the right orbit. The right globe is compressed and displaced inferiorly by the mass, which also extends superiorly to elevate and thin the roof of the orbit in an irregular fashion. The center of the mass shows marked hyperintensity, consistent with blood.

Figure 2. Computed tomographic scanning results. Coronal section through the orbits (bone-density window setting) reveals marked irregularity of lateral wall and roof of the right orbit associated with a diffuse mass with a slightly decreased density in its center. Note infradisplacement of the right globe and a small area in the orbital roof in which the bone appears to be absent.

Figure 3. Photomicrographs showing histopathologic characteristics of the lesion. A, The central lytic portion of the lesion shows fibrous tissue and stacked cholesterol clefts, some of which are surrounded by foreign-body–type giant cells (hematoxylin-eosin, original magnification ×190). B, An island of entrapped bone in the central portion of the lesion shows a distinct mosaic pattern of remodeling lines diagnostic of Paget disease (hematoxylin-eosin, original magnification ×210). C, Bone from margin of the lytic portion of the lesion has a zonal pattern, showing a transition from dense orbital bone (right portion of photograph) to highly resorbed, pagetic bone (left portion of photograph) (hematoxylin-eosin, original magnification ×60). D, Higher-power magnification of the resorbing bone shows numerous resorption cavities and pagetic osteoclasts (hematoxylin-eosin, original magnification ×240).
transition from dense facial bone to bone that was highly resorbed (Figure 3, C). The resorbing bone was directly adjacent to the central fibrous portion of the lesion, and, in this area, there were numerous large osteoclasts (Figure 3, D). These features were thought to be characteristic of the active (lytic) phase of Paget disease.

COMMENT

A variety of orbital lesions are characterized histopathologically by the presence of cholesterol clefts and altered blood products. These include aneurysmal bone cysts, epidermoid cysts, and cholesterol granulomas.7,8 In our case, the lesion was initially thought to be a cholesterol granuloma, because it contained numerous cholesterol crystals surrounded by granulation tissue and blood-derived debris; however, the lesion had features not associated with a cholesterol granuloma. In particular, the bone adjacent to the central lytic area was being resorbed by numerous osteoclasts. The finding of a zone of osteoclastic resorption at the margin of the lesion, combined with the islands of retained bone in the central portion of the lesion, indicate lytic Paget disease. Although many resorptive processes are characterized by large resorption cavities, the mosaic pattern, indicating chaotic remodeling, is diagnostic of Paget disease.9 The results of neuroimaging in cases of isolated Paget disease of the orbit have only rarely been reported. Khetarpal et al9 described an 89-year-old woman who developed proptosis of the right eye associated with progressive loss of vision in the eye. Computed tomographic scanning revealed the extent of bony involvement of the orbit in this case. We are unaware of any previous cases of orbital Paget disease in which the magnetic resonance imaging findings have been described.

Patients with Paget disease of the orbit who develop neurologic or visual complications of the disorder may benefit from a variety of drugs, particularly calcitonin.3,10 Others, such as our patient, may require only local excision of the lesion.

Our patient’s case indicates that Paget disease can present not only as a diffuse osteolytic process but also as an indolent, painless, solitary lesion of the orbit, the imaging and histological characteristics of which may mimic those of a cholesterol granuloma.