Paget disease is a common condition characterized by increased bone turnover and abnormal bone deposition in the skull and vertebral column. This article describes a previously undiagnosed case of Paget disease in which the manifesting clinical symptom was gradually progressive diplopia from sixth-nerve compression due to skull base hyperostosis. The sixth-nerve palsy, which was severe at the initial examination, slowly resolved after the patient was treated with intravenous zoledronic acid. To my knowledge, Paget disease has not previously been reported to manifest initially with diplopia, nor has diplopia due to this condition been reported to improve with medical therapy.

**Report of a Case.** A 74-year-old man was referred with a 4-month history of gradually worsening horizontal diplopia, worse on looking to the right. He also had gradually worsening right hemifacial pain and paresthesia.

On examination, visual acuity was 6/7.5 OU. There was a 40–prism diopter (PD) right esotropia for distance in primary position, with the right eye abducting to the midline but not beyond in attempted far right gaze (Figure 1 A). Other ocular ductions were normal. There was right corneal anesthesia as well as reduced cutaneous sensation to light touch on the right forehead and cheek, with normal lower facial sensation. There were no other cranial neuropathies.

The patient underwent magnetic resonance imaging of the brain, which revealed extensive skull base thickening. A subsequent computed tomographic scan demonstrated extensive diffuse demineralization of the skull base extending into the skull vault, a loss of cortical medullary differentiation, a cotton-wool bone appearance, and an overgrowth of bone at the petrous apex in the region of the Dorello canal bilaterally (Figure 2). Blood tests revealed an alkaline phosphatase level of 616 U/L (reference range, 30–115 U/L; to convert units per liter to microkatals per liter, multiply by 0.0167), and Paget disease was diagnosed.

Treatment was commenced with an intravenous infusion of 4 mg of zoledronic acid. Six weeks after the treatment, the patient began to notice a gradual improvement in the diplopia. Three months after the infusion, the right eye was able to abduct beyond the midline. A second infusion was given 6 months after the first. At last follow-up, 7 months after commencement of treatment,
the patient can achieve binocular single vision with a slight right head turn; there is an 8-9PD right esotropia in primary position and markedly improved right eye abduction (Figure 1B). The previously severe right hemifacial pain and paresthesia have completely resolved. Right corneal sensation has returned, although it is still subjectively reduced; there are no signs of neurotrophic keratopathy.

**Comment.** The gradual onset and slow progression of the sixth-nerve palsy suggest that progressive compression of the sixth nerve by skull base hyperostosis was the cause of the patient’s diplopia. Patients with known Paget disease have previously been described to have developed diplopia due to cranial nerve compression2; however, to my knowledge, the disease has not previously been reported to initially manifest with diplopia.

Bisphosphonates achieve a rapid reduction in bone turnover in Paget disease by inhibiting osteoclast resorption.3 Zoledronic acid is a potent new bisphosphonate. Intravenous infusion of this drug has recently been demonstrated to provide a greater and more rapid effect on bone metabolism than oral agents.4 The gradual improvement in the severity of the sixth-nerve palsy in this case presumably was due to slow reduction in the extent of bony compression of the nerve at the petrous apex.

Although diplopia due to Paget disease is rare, the condition should be considered in the differential diagnosis of slowly progressive diplopia in elderly patients. The use of new bisphosphonate agents may be an effective treatment for diplopia caused by this disease.

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**COMMENTS AND OPINIONS**

**Bilateral Orbital Infarction, Ophthalmic Artery Occlusion, and Cherry-red Spot**

In the March 2007 issue of the Archives, Maier et al1 described a case of bilateral orbital infarction syndrome due to ophthalmic artery occlusion following craniotomy.

Ophthalmic artery occlusion produces profound vision loss (light perception or no light perception) due to the simultaneous ischemia of the choroid and retina. A cherry-red spot can be an acute initial sign but is more suggestive of central retinal artery occlusion. In previoulsy reported series of orbital infarction, profound vision loss was seen in all of the cases, including those with a cherry-red spot.2,3 Close serial examination allowed the investigators to note the “disappearance of the cherry-red spot, suggesting choroidal ischemia.”2 Maier and associates did not provide such follow-up information, and more importantly, their patient did not have profound vision loss.

With respect to the fluorescein angiogram provided (Figure 2C in the original article), Maier and associates commented on the presence of retinal edema in the late frame. If retinal edema were present, hyperfluorescence with cystic pooling at the level of the retina would be expected; however, the late frame appears to demonstrate hyperfluorescence at the level of the choroid instead.

As a point of terminology, retinal edema refers to extravasation of fluid from the vasculature into the extracellular space producing cystic changes (eg, cystoid macular edema and diabetic macular edema), whereas the term retinal whitening refers to retinal thickening found in central retinal artery occlusion due to increased intracellular fluid content.

The case reported by Maier and associates is not consistent with previous reports of orbital infarction syndrome; rather, it appears to resemble a previous report of central retinal artery occlusion and ophthalmoplegia following spinal surgery.4

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In reply

We thank Dr Federici for his thoughtful letter referring to our recent article titled “Bilateral Orbital Infarction Syndrome After Bifrontal Craniotomy.”1 Herein, we would like to take the opportunity to reply to his points of criticism.

First, Dr Federici stated that “ophthalmic artery occlusion produces profound vision loss (light perception or no light perception) due to the simultaneous ischemia of the choroid and retina.” Our patient had profound and permanent visual loss from a visual acuity of 20/20 OU prior to the infarction to recognition of hand motion in both eyes. Although there is a significant difference between hand motion and light perception,3 the visual loss from a visual acuity of 20/20 to hand motion or to light perception is comparable (20/20 to hand motion is 23 logarithmic lines; 20/20 to light perception is 27 logarithmic lines). There is no proof that visual acuity bet-