SOLITARY PLASMACYTOMA OF PETROUS BONE
REPORT OF A CASE WITH NEUROLOGIC AND RADIOGRAPHIC REMISSION FOLLO WING ROENTGEN-RAY THERAPY

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Multiple myeloma involving the vertebral column frequently produces neurologic symptoms secondary to compression of the spinal cord and spinal nerve roots.3,8,9 Similar neurologic manifestations of intracranial involvement are extremely rare, however. In an exhaustive survey of this subject, Clarke3 reviewed the world literature and found only 24 cases of multiple myeloma involving the base of the skull and producing cranial-nerve palsies. At that time he reported 1 additional case of his own. Ten of these 25 cases were presented as examples of solitary plasmacytoma of the base of the skull. In several of his cases, however, the data and length of follow-up observation were insufficient to sustain a diagnosis of the solitary form of multiple myelomatosis.

The following case report, presenting an additional patient in whom neurologic structures at the base of the brain were involved by myeloma of the surrounding bony structures is deemed of interest because of two additional unusual features:

(1) Follow-up studies have been carried out in this case for 7 years from the time of onset of the disease and for 2 years since the disease was verified by pathologic study and treatment was initiated. At present there is still no evidence of dissemination of the disease beyond the base of the skull. Thus, the diagnosis of solitary plasmacytoma might, at least presumptively, be entertained at this time.

(2) The patient’s response to subtotal resection of the neoplasm and secondary roentgen-ray treatment of the involved area has been striking, both clinically and radiologically. Before this therapy, the neurologic deficit secondary to involvement of brain stem, cerebellum, and cranial nerves was profound and incapacitating. At the present time (2 years after treatment) virtually no evidence of neurologic disease is present except for a residual unilateral loss of hearing. Similarly, 2 years after roentgen-ray therapy directed to the base of the skull, evidence of recalcification and formation of new bone is apparent in areas previously completely eroded by the expanding neoplasm.

REPORT OF CASE

A 55-year-old housewife was admitted to the hospital on Feb. 27, 1957. Five years previously she had noticed the onset of deafness and tinnitus in the left ear. Both of these symptoms became progressively more severe over the ensuing years. About 1 year before admission headaches and pain in the left suboccipital and temporo-occipital regions developed. In December 1956 she began to complain of intermittent diplopia, especially on left lateral gaze. At that time she and members of her family noted intermittent “twitching” of the left side of the lower part of the face and “watering” of the left eye; these signs were associated with

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drooping of the left side of the mouth. All of these symptoms became progressively more severe. During the month before admission she began to complain of intermittent difficulty in swallowing food and liquids, and occasional inability to articulate clearly. Her gait was becoming impaired with staggering, especially to the left, on attempting to walk. She consulted her family physician who referred her to the Lahey Clinic.

Examination. There was mild right hemiparesis with accentuated deep tendon reflexes on that side. The patient displayed severe truncal ataxia manifested by inability to stand upright without swaying and falling backward. In addition, pronounced ataxia occurred on walking, with falling to the left side. The heel-to-shin test was poorly performed on the left. Movements of the left upper extremity were slow, and a slight intention tremor of the left hand was noted.

The right pupil measured 2.5 mm. and the left pupil 3.5 mm. in diameter. Both pupils reacted well to light, but convergence of the left eye was impaired. A mild left abducens paresis was noted. No deficit of corneal reflexes was observed, nor was there any evidence of impairment of the motor or sensory functions of the trigeminal nerve on either side. A facial weakness of the lower motor neuron type was present on the left side. The patient was completely deaf in the left ear and Weber’s test revealed that this was a nerve deafness. Caloric function was tested and complete paresis of the canal to hot and cold stimuli was demonstrated on the left. The tongue was atrophied on the left side with obvious fasciculation; deviation to the left occurred on attempted protrusion. No evidence of visual impairment was apparent, and the results of funduscopic examination were normal. There were no abnormalities of the nose and throat.

Hemoglobin measured 14.5 gm., and white blood cells numbered 6,200, with a normal differential count. Nonprotein nitrogen was 28 mg. per 100 cc. Cerebrospinal fluid pressure was normal. The fluid contained 1 red blood cell and no white blood cells; protein measured 30 mg. per 100 cc.

A roentgenogram of the chest was normal. Roentgenograms of the skull revealed an extensive destructive process at the base on the left side. This involved the left petrous pyramid and extended anteriorly to the petrous tip on the left side. The greater wing of the sphenoid process, the sella turcica and the clivus were also involved in this destructive process. Pneumoencephalography revealed evidence of a neoplasm arising from the area of bone destruction and expanding into the prepontine region and into the lateral cisternae on the left side.

Operation. A left suboccipital craniectomy was performed on March 4, 1957. The occipital bone just medial to the left mastoid process was eroded and partially destroyed by tumor. The neoplastic tissue was firm, purplish-grey and extremely tenacious. It appeared to be of a gelatinous consistency with irregular fibrous strands running through. The precise site of origin could not be determined, but the main bulk of neoplasm that could be visualized was located in the region of the left petrous pyramid which was severely eroded. The tumor was extradural and had displaced the dura mater covering the left cerebellar hemisphere toward the right; it extended into the cerebellopontine angle on the left and spread anterosuperiorly toward the region of the tentorial notch. No cranial nerves were visualized, as they were encased in the dense neoplastic stroma. A subtotal, but minimal, resection of the tumor was performed.

Microscopic examination revealed the lesion to be a typical plasma-cell myeloma (Fig. 1).

Postoperative Course. Roentgenograms of the lumbosacral region, pelvis, rib cage and long bones disclosed no evidence of multiple myelomatosis. Repeated testing of the urine revealed no evidence of Bence Jones proteinuria. Total protein and albumin values were normal. Equivocal elevation of globulin to 3.4 gm. per 100 cc. was found; 1 month later the globulin was normal—2.9 gm. per 100 cc. Electrophoretic studies of serum proteins showed evidence of an increase in the gamma globulin fraction.

No abnormalities were found in the peripheral blood smear, nor were plasma cells present. Examination of the sternal marrow on March 12 showed some generalized activity suggestive of normoplastic hyperplasia. No abnormal cells consistent with multiple myelomatosis were found.