SOLITARY PLASMACYTOMA OF PETROUS BONE

REPORT OF A CASE WITH NEUROLOGIC AND RADIOGRAPHIC REMISSION FOLLOWING 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.
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Fig. 1. High-power view of plasmacytoma (hematoxylin and eosin, ×400). Tumor is composed of round and polygonal cells fairly densely distributed in a scant connective-tissue matrix. The cells have a moderately eosinophilic cytoplasm with a densely staining solid basophilic nucleus which is often eccentric. The smaller cells, especially, resemble plasma cells.

No essential change in the patient's neurologic status occurred during the immediate postoperative period. A tumor dose of 3,000 r was administered to the base of the skull and she was discharged on March 23, 1957.

Subsequent Course. On a visit to the clinic in May, the patient stated that the headaches had disappeared and that the tinnitus in the left ear was much less severe. Examination in August revealed continuing subjective and objective evidences of improvement. She no longer complained of diplopia, and there was no deficit of the extraocular muscles. Her gait had become normal and she neither swayed nor fell on standing and walking. The left facial weakness had become less pronounced. The sternal marrow was re-examined and again it was normal. Total serum protein measured 7.1 gm. and the globulin was 3.1 gm. per 100 cc. A mild increase in the gamma globulin fraction was still evident on electrophoretic studies, however. In April 1958, the left facial weakness had disappeared completely. The only neurologic signs elicited were residual deafness in the left ear and slight deviation of the tongue to the left on protrusion. Blood studies were normal. Serum protein measured 7.4 gm. and the globulin fraction was 2.8 gm. per 100 cc. Mild elevation of the gamma globulin fraction was still demonstrated by electrophoretic studies, but this elevation was less than that found 1 year previously.

She was last seen in November 1958, approximately 21 months after admission to hospital and 7 years after the initial symptoms had developed. The striking neurologic improvement had been maintained. Clinical and chemical studies of the blood gave normal results. (Elec-
trophoretic studies of serum proteins were not carried out at this time.) No clinical or radiologic evidence of progression or dissemination of the disease process was apparent. Roentgenograms of the skull showed the old destructive process at the base on the left, and the bone defect left by the left suboccipital craniectomy. At this time, however, evidence of definite recalcification and formation of new bone in the region of the sella turcica was seen.

DISCUSSION

Many of the clinical features observed in this case are characteristic of the 25 proved cases of myeloma of the base of the skull with cranial-nerve palsy previously reported by Clarke.\(^3\) The duration of illness in our patient is longer than that in most of the cases outlined in his series, however. The average length of the disease from onset to death in Clarke’s series was 2 years—a figure that corresponds closely with the survival rates in generalized myelomatosis reported by other authors.\(^4,\)\(^5\) Three patients were alive more than 6 years after the onset of initial symptoms.

In this case, the myeloma presumably originated from the petrous temporal bone. In the past, however, similar tumors have been thought to have originated in extraosseous lymphoid structures of the nasopharynx and to have involved the base of the skull secondarily. This type of spread was seen in the case reported by Cappell and Mathers.\(^6\) While some authors\(^5,\)\(^7\)\(^8\) have suggested that lymphoid and myeloid tumors may arise from the mucous membranes of the nasopharynx or accessory air sinuses, this view was disputed by Stout and Kenney.\(^10\) The latter authors felt that mucous membranes were always involved secondarily. Jackson \textit{et al.}\(^5\) concluded that multiple myeloma, plasmacytoma and plasma-cell leukemia are all manifestations of one disease entity, however. On the basis of clinical and pathologic data, these authors stated that “multiple myeloma of the plasma type should be classified under the lymphomata.”

Multiple myeloma characteristically has a lytic effect on bone. Radiologically, the changes in the bone vary from generalized demineralization to complete destruction. The edges of the area of destruction are sharp without any evidence of secondary condensation. Recalcification and formation of new bone in areas previously destroyed, as seen in radiographs following therapy, are extremely rare. We have been able to find only 8 such cases. New bone was reported to have formed at the base of the skull in Case 1 of Clarke’s 2 years after a dose of 4,900 r had been delivered to the neoplastic area. Bailey\(^4\) also reported a patient with a severe destructive lesion of the humerus. Following biopsy study, radiation therapy was directed to this lesion although the exact dosage was not reported. Six years after radiation the patient was well and showed deposition of lime salts with resumption of radiographic characteristics of normal bone in the area previously destroyed. The tumor in this case was somewhat puzzling histologically, and the pathologists were never completely sure that this was a true case of myeloma. Presumably, the expanding myeloma in these cases, while destroying some bone, also caused decalcification and secondary pressure atrophy of bone in contiguous areas. When regression of the neoplasm was produced by roentgen-ray treatment, the atrophied bone, in which the matrices were still intact, recalcified and then served as a base for formation of new bone.

Although in the vast majority of cases of myeloma the tumors are multiple from the beginning or soon show evidence of multiple bony lesions, a few patients with solitary localized plasmacytoma have been observed for years and even decades\(^4,\)\(^7,\)\(^8\) without dissemination. They probably occur as \textit{formes frustae} of what is essentially a generalized neoplastic disease of the reticuloendothelial system. The following con-
considerations favor the concept that the tumor in the present case will remain solitary and that disseminated lesions will not develop. (1) The patient has now been observed for 7 years since the onset of symptoms, with no clinical or radiologic evidence of dissemination. (2) The results of examinations of urine for Bence Jones protein have been negative on repeated occasions. (3) Numerous samples of the peripheral blood have been examined and have failed to show abnormalities. (4) Total serum protein and albumin fractions have always been within the normal range. The serum globulin was mildly elevated at the time of admission to hospital in February 1957, but normal amounts of this component have since been obtained on successive follow-up examinations. (5) The local neoplasm in the skull has responded dramatically to roentgen-ray therapy and has remained dormant. (6) Two examinations of the sternal bone marrow have failed to show any reflection of a myeloid neoplasia which is not apparent clinically.

A cause for some concern, however, has been the mild, but persistent elevation of the gamma fraction of the serum globulin as determined electrophoretically. An elevation of this component is probably caused by an abnormal myeloma globulin which has been found to be immunologically different from the hyperglobulinemia seen in other diseases.6 This persistent elevation of the gamma globulin fraction may represent a generalized reticuloendothelial diathesis, which is not yet apparent clinically or histologically, but could eventuate in the characteristic picture of multiple myelomatosis.

REFERENCES