Recurrent Anterior Skull Base Plasmacytoma

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CASE HISTORY

A 53-year-old man was referred for treatment of a mass that invaded the posterior nasal cavity, ethmoid sinuses, both posterior orbits, and anterior skull base. Eighteen years earlier, he had been diagnosed with a solitary plasmacytoma of the paranasal sinuses. Despite treatment with 4500 cGy radiation therapy, the tumor continued to grow and was subtotally resected 3 years later through a transfacial approach. Two years before his current referral, he underwent a second operation for a massive recurrent tumor. On that occasion, the highly vascularized tumor was completely removed anterolaterally through a subcranial approach. The patient elected not to have the pericarotid and perichiasmatic extensions of the tumor removed to spare these vital structures. The anterior skull base was reconstructed with the use of fascia lata, biological glue, and titanium miniplates. After surgery, the patient returned to his native country with the recommendation to undergo a complementary course of radiation therapy and chemotherapy.

The patient experienced intermittent epistaxis and nasal obstruction for 3 months before his current referral. He had received no radiological or chemotherapeutic intervention during the interven-

ing 2 years. On examination he appeared well. His speech and gait were normal, and he suffered no visual disturbances. Transnasal endoscopic examination showed that the anterior cavity was clean. A reddish mass obstructed the posterior nasal cavity and choana. Mirror laryngoscopy revealed normal vocal fold function and no suspicious lesions of the oropharynx, hypopharynx, or larynx. The oral cavity and tongue were normal. The neck was without adenopathy. Cranial nerves function was intact. The remainder of the physical examination revealed no musculoskeletal or cardiovascular abnormalities.

His erythrocyte sedimentation rate was 50 mm/h and his blood count was normal. Electrolytic, calcium, glucose, albumin, and liver functions were normal. Serum electrophoresis was normal, with serum globulin of 22 g/L and no monoclonal peak. Urinalysis was normal, and a test for Bence-Jones protein was negative. Pathological examination of the bone marrow aspirate showed a normal number of mature plasma cells with no evidence for monoclonal heavy or light chains.

Computed tomography (CT) and magnetic resonance imaging (MRI) of the skull base, paranasal sinuses, and orbits showed a 3- × 4-cm vascularized tumor involving the ethmoid cells and sphenoid sinus, eroding both orbits and invading

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Figure 1 (A) Sagittal T1-weighted MRI of the brain shows intracranial invasion of the tumor at the roof of the sphenoid sinus. The posterior ethmoid cells are also infiltrated by tumor. (B) Coronal T1-weighted MRI shows the tumor eroding the skull base and orbital apex on the left. The posterior ethmoidal cell and nasal cavity are invaded by the tumoral mass.

the anterior cranial fossa. Posteriorly, the tumor impinged on both the carotid arteries and optic nerves (Fig. 1).

Histopathological examination of the previous biopsy showed diffuse sheets of immature-appearing plasma cells (Fig. 2). Immunohistochemical staining confirmed the diagnosis of plasmacytoma.

ACTUAL MANAGEMENT

The patient underwent surgical excision of the tumor through a subcranial approach. The intracranial extension of the lesion was not excised. We believe that violating the dura in his condition (multiple surgical procedures and adjuvant radiotherapy to the involved area) would have been inappropriate.

The patient's postoperative course was unremarkable, and he was discharged with a recommendation to undergo chemotherapy (vincristine, doxorubicin hydrochloride, dexamethasone) and radiotherapy (2000 to 3000 cGy).

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Figure 2 Photomicrographs of the tumor. (A) The field consists entirely of immature-appearing plasma cells. Moderately dispersed chromatin and occasional nucleoli are present (hematoxylin and eosin stain $\times 400$). (B) Immunoperoxidase stain with antikappa antibody shows a monoclonal staining pattern for kappa light chain.

RESPONSES

We would consider offering the patient radical surgical excision of all macroscopic tumor using a combined approach from above and through the midface preserving vital structures such as the optic nerves and carotid arteries. We would identify the optic chiasm from above and follow it forward to decompress the optic nerves. We would recommend reconstructing the floor of the anterior cranial fossa with free revascularized tissue, such as a muscle-only rectus abdominis free flap, to introduce fresh tissue into the field that will withstand further radiation. In our hands, this strategy has reduced the likelihood of complications (*Plast Reconstr Surg* 1996;98:1159–1166). Recognizing that there is likely to be residual microscopic disease after surgery, we would strongly recommend a course of concurrent chemotherapy and further radiation using conformal techniques such as intensity-modulated radiation therapy to deliver a dose of about 55 Gy to a focused area in the previously radiated field.

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This very interesting patient demonstrates the dilemma posed by subtotal resection of malignant tumors that achieve various degrees of palliation, but invariably persist or recur. The remarkable feature of this case is that the patient has survived with what appears to be a good quality of life for 18 years. Another noteworthy aspect is that he has continued to have local recurrences of the plasmacytoma without developing multiple myeloma, which is the usual scenario in individuals with a solitary plasmacytoma, whether the primary lesion recurs after local therapy or not.

In general, subtotal resection of head and neck malignancies, followed by radiation therapy or radiation coupled with chemotherapy, rarely results in a cure and often does not even provide a significant period of remission. However, it is important to note that most head and neck malignancies that pass through the skull base and extend intracranially are of epithelial or soft-tissue origin. The goal of surgery in these carcinomas and sarcomas is complete excision with tumor-free margins followed by a full course of radiotherapy if this latter treatment has not been given previously. The same principle applies to a solitary plasmacytoma, but this lesion is usually sensitive to both radiation and certain chemotherapeutic agents.

In this particular case, the irradiation may have slowed the growth of this tumor but was not curative. All resections that followed the irradiation were subtotal and predictably recurred. It is inaccurate to say that the subcranial approach performed preceding the most recent procedure was a complete resection when intracranial tumor was left behind. Following the prior procedure, the tumor inevitably recurred, as it will after the most recent resection. The reason given for not removing the intracranial component was concern about re-establishing the integrity of the dura and preventing cerebral spinal fluid leakage because of the prior surgical interventions and radiotherapy. This concern can be obviated by the use of a long pericranial flap fixed by sutures, passed through drill holes in the planum sphenoidale, and augmented by fibrin glue. If the pericranium has been used previously or is in poor condition and thus unusable, a dural graft covered by a radial forearm free flap with the donor vessels passing through a high pharyngotomy in the neck can be used.

The issue of dissecting the tumor from the pericarotid and perichiasmal area should not be a problem using the anterior subcranial (Raveh) approach with or without a frontal extension. This tumor is relatively slow growing and relatively radioresistant. A prolonged period of palliation and even a cure may be accomplished by surgery. Careful dissection under microscopic control by a

neurological surgeon skilled in skull base surgery should be able to achieve complete tumor removal or at least a resection with only microscopic residuum. Additional radiation is contraindicated because of the high risk to the optic nerves. Gamma knife irradiation would endanger these nerves even more seriously. Irradiation for this relatively radioresistant tumor must be given at a sublethal tumor dose and is likely to be attended by serious complications.

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A slow-growing plasmacytoma treated 18 years earlier with radiation therapy was followed by subtotal resection 15 years earlier and, again, 2 years before the current recurrence. Another operation with resection of the extracranial portion of the tumor (incomplete resection) violating the dura is considered to be inappropriate. The images show a huge tumor of the anterior skull base with an intracranial extension between the posterior wall of the frontal sinus and the beginning of the planum sphenoidale. Unfortunately, no computed tomography images are available for a clear statement. However, the tumor does not look as if it has grown through the dura. According to the available images, there seems to be no intradural extension.

Without treatment, further growth of the residual intracranial portion of the tumor is to be expected. There is some danger of intradural growth.

GENERAL TREATMENT OPTIONS

Because previous radiotherapy involved a relatively low dose of 4500 cGy and was performed 18 years earlier, another course of stereotactic radiotherapy might be possible. The outcome, however, would

be questionable. Chemotherapy obviously did not have a curative effect.

PERSONAL TREATMENT PREFERENCE

Through an interdisciplinary approach with a neurosurgical partner, I would again use the subfrontal approach and try to resect the entire tumor microsurgically. Because the patient's vision is normal on both sides, infiltration of the optic nerve is unlikely. Microsurgical resection of the tumor preserving both optic nerves and the internal carotid artery should be possible. Before surgery, a balloon occlusion test is necessary to determine if one internal carotid artery could be sacrificed. The patient is only 53 years old and in good general condition. Therefore, a curative surgical option in a relatively nonaggressive malignant lesion seems to be the preferred option.

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SURGICAL MANAGEMENT

Apart from the fact that solitary plasmacytoma is an extremely rare tumor of the head and neck region, the history of this case is most interesting. Due to this rarity there is a paucity of information on treatment approaches and outcome, leaving the optimum management poorly defined. According to the current literature, however, complete surgical resection with adjuvant radiation therapy is recommended, which we also endorse.

In this case, after initial radiation therapy (4500 cGy) 18 years earlier, subtotal tumor resection was performed twice in the ensuing years. As described, the reason for partial resection during the second operation, leaving the "pericarotid and

perichiasmatic" extensions, was to spare these structures. The question is: To what degree were these structures involved? Were they fully encased or just effaced by the tumor? The author notes that the extensions of the actual third recurrent tumor impinged on both the internal carotid artery and optic nerves apart from intracranial involvement.

The selection of the subcranial approach, as developed and described by us, was correct, particularly because this approach provides broad exposure of these locations under direct vision. Thus, removal of the tumor extensions effacing the optic nerves and carotid artery as well as the intracranial portion sparing vital structures is facilitated. With respect to the author's experience with this approach, there is no doubt that he could have performed this procedure. Accordingly, his concern was not related to the extensions impinging on the optic nerves or carotid artery but to violating the dura (after the previous procedures) and thus leaving the intracranial extension.

In regard to the time elapsed between the last operation and our experience in similar cases

(with different diagnosis), one layer of fascia lata fixed to the borders of the defect with sutures or glue and alignment of the skull base up to the posterior wall of the sphenoid sinus with two further layers have proved to be the most reliable strategy. Thus, given that recurrence of solitary plasmacytoma with subtotal resection is possible despite radiotherapy—particularly as manifested in this case, I would have opted for a radical resection. The treatment would have included the intracranial extension, resection and reconstruction of the eroded orbital walls, and adjuvant radio- and chemotherapy as suggested by the author. In my opinion this approach is justified to reduce the risk of a further recurrence.

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