Surgical management of midline skull-base tumors: a new approach

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Many surgical approaches to the clivus and upper cervical spine have been used in the treatment of skull-base tumors over the past 50 years. However, the outcome of surgery has been complicated by difficulties of access to the whole clivus, together with pharyngeal wound breakdown with subsequent development of cerebrospinal fluid (CSF) fistula and meningitis. A technique described recently utilized Le Fort I osteotomy to improve exposure of the clivus in the approach to vertebrobasilar aneurysms, facilitating control of the aneurysm and reducing the risk of posttraumatic CSF fistula. The same approach, via maxillotomy, has permitted partial or total tumor resection in 13 consecutive procedures carried out at Atkinson Morley's Hospital on 10 patients presenting with tumors of the skull base. Neurological status was either improved or unchanged in all patients postoperatively, and pain relief was obtained in five of eight cases in which this was a presenting symptom. No patient developed a CSF fistula following surgery. Cosmetic results were good, and no problems related to malocclusion were reported. This approach may be used to advantage in the surgical treatment of skull-base tumors, at initial presentation, and can be repeated without undue difficulty should there be tumor recurrence.

KEY WORDS • skull-base neoplasm • transclival approach • operative approach

NEOPLASMS involving the skull base are rare, probably accounting for less than 1% of intracranial tumors. They are also diverse in terms of pathology and can arise from any of the embryological rests or specialized tissues contained within the skull base. Despite this diversity in origin they behave in a rather uniform way: they are frequently benign or only locally malignant, they rarely metastasize, and they prove refractory to radiotherapy or chemotherapy; thus, radical resection is the treatment of choice in their management. Surgery may have to be repeated should the lesion recur, and ideally this repeat operation should not be attended by insuperable difficulties.

The clinical syndromes associated with these tumors extend from the optic nerves and chiasm down to the foramen magnum and encompass visual disturbances, pituitary dysfunction, orbital apex and cavernous sinus syndromes, nasopharyngeal masses leading to airway obstruction, cranial nerve palsies, brain-stem compression, cerebellopontine angle lesions, and foramen magnum syndromes. New imaging techniques such as computerized tomography (CT) and magnetic resonance (MR) imaging help in the identification of these tumors and increase the accuracy of their delineation. Surgical aids such as the microscope, the laser, and the ultrasonic aspirator have greatly facilitated the excision of these tumors. Modern anesthesia with controlled ventilation, hypotension, and advanced intensive care facilities for the postoperative phase have all helped to improve the operative field and the ultimate outcome.

In this paper a new approach to tumors in and around the clivus using a Le Fort I osteotomy is described. The need for teamwork in these complex operations is established. The combination of a maxillofacial surgeon and a neurosurgeon has been utilized successfully in the clipping of vertebrobasilar aneurysms. The technique provides improved exposure of the clivus and facilitates wide tumor excision with a much reduced risk of cerebrospinal fluid (CSF) fistula; if surgery is deemed necessary at a late date, it may be performed without adding materially to the difficulties.

Clinical Material

Since 1985, 13 procedures have been carried out on 10 consecutive patients presenting with tumors in and around the clivus. The patients' age range was 29 to 66 years at the first operation, with an average age of 43 years. Six patients were female and four male. The duration of symptoms before presentation to Atkinson
Morley's Hospital ranged between 4 months and 8 years. The presenting symptoms and signs are summarized in Table 1.

Of the 10 patients, seven had undergone previous surgery at the time of the first Le Fort I operation, and six had received radiotherapy. In three cases, the Le Fort I procedure was repeated for local tumor recurrence 12, 14, or 18 months after the original operation. The extent of tumor invasion was determined radiologically by CT or MR imaging with sagittal and coronal re-formations. Three-dimensional reconstruction from CT scans was executed where it was believed to be particularly helpful. Angiography was carried out in eight of the 10 cases before the first operation to demonstrate any involvement or displacement of major vessels. The histological diagnosis of the tumors encountered at operation are presented in Table 2.

Operative Technique

Operations were performed under general anesthesia with orotracheal intubation, except in two cases where preoperative tracheostomy was carried out because of airway obstruction. A lumbar spinal drain was inserted preoperatively to allow continuous CSF drainage. Prophylactic antibiotic cover was commenced at induction of anesthesia, using an intravenous cephalosporin (Ceftaxime) and metronidazole. The patient was positioned supine on the operating table, with a 15° head-up tilt, and the oral cavity was cleansed with an aqueous povidone-iodine solution.

TABLE 1

<table>
<thead>
<tr>
<th>Symptoms &amp; Signs</th>
<th>No. of Cases</th>
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<tbody>
<tr>
<td>symptoms</td>
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<tr>
<td>neck pain/headache</td>
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<tr>
<td>visual disturbance</td>
<td>7</td>
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<td>4</td>
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<td>1</td>
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<tr>
<td>unsteady gait</td>
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<tr>
<td>signs</td>
<td></td>
</tr>
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<td>nasal/airway obstruction</td>
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<tr>
<td>cranial nerve palsies</td>
<td>10</td>
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<tr>
<td>long-tract signs</td>
<td>3</td>
</tr>
<tr>
<td>ataxia</td>
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TABLE 2

<table>
<thead>
<tr>
<th>Histological Diagnosis</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>chordoma</td>
<td>4 (2 recurrent)</td>
</tr>
<tr>
<td>chondrosarcoma</td>
<td>5 (1 recurrent)</td>
</tr>
<tr>
<td>meningioma</td>
<td>1</td>
</tr>
<tr>
<td>pituitary adenoma</td>
<td>1 (infiltrating bone)</td>
</tr>
<tr>
<td>neurilemoma (9th nerve)</td>
<td>1</td>
</tr>
<tr>
<td>renal carcinoma</td>
<td>1</td>
</tr>
<tr>
<td>total tumors</td>
<td>13</td>
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An incision was made above the mucogingival reflection extending from one upper molar area to the other. A bilateral low-level bone cut was then made with an air-powered sagittal saw through the malar buttress to the maxillary tuberosities, thus exposing the mucosa of the nasal floor and giving access to the pterygomaxillary fissure (Fig. 1 left). Before proceeding further, the positions of the Luhr compression plates necessary for fixation of the maxilla at the completion of the operation were marked on either side of the saw cut. The nasal septum was then divided from the maxilla in the midline, and a curved chisel was used to separate the lateral pterygoid laminae from their maxillary attachments. The maxilla could then be “down-fractured,” its blood supply being maintained by the greater palatine arteries and the mucosal blood supply of the faucial pillars. This mobilization allowed the maxilla to be dropped downward through the normal range of mouth opening. The mucosa of the nasal floor was removed on each side to expose the nasal septum and vomer. The inferior turbinates were excised using heavy scissors, and the vomer was removed piecemeal to expose the roof of the nasopharynx and the clivus behind it.

A modified Dingman gag was then inserted to retract the cheeks laterally and displace the maxilla downward, thereby exposing the mucosa of the posterior pharyngeal wall. Thus, the clivus was accessible from the middle ethmoid sinuses to the foramen magnum and the anterior arch of the atlas (Figs. 1 right and 2). The mucosa overlying it was incised in the midline and retracted laterally. With a high-speed drill the clivus was removed down to the dura and the incision was extended using a Kerrison bone punch. Any bleeding from dural sinusoids at this stage was controlled by packing with oxidized cellulose gauze. Where necessary, the underlying dura was coagulated and opened to facilitate access to the tumor. The amount of bone

Fig. 1. Sagittal views of a dried skull showing the line of osteotomy (left) and the orientation of the retractor (right). Arrows indicate the arc of exposure.
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removal depended on the extent of tumor involvement, but it was possible to gain access via this approach to the middle ethmoid sinuses, the sphenoid sinus, and the clivus down to the foramen magnum, and to the lateral recesses on either side. Total macroscopic removal of tumor, piecemeal with rongeurs or using an ultrasonic surgical aspirator, was achieved in all except three cases. The dura was opened in three cases, but appeared intact in the others.

In earlier cases, a Lyodura patch was applied to the dura followed by a free muscle graft but, in more recent procedures, the dural defect has been repaired by two layers of Lyodura, one inside and the other outside the dura, the “sandwich” being held together with human-derived fibrin glue (Tisseel). The bone defect was filled with layers of Oxycel and Tisseel before the overlying mucosal edges were approximated without sutures and sealed with the fibrin adhesive. Fixation of the maxilla was achieved with Luhr vitallium minicompression plates applied in predetermined positions. The mucosa was then repaired with interrupted silk sutures.

Before reversal of anesthesia, a nasogastric tube was passed under direct vision for postoperative feeding. In the three most recent cases, a puncture pharyngostomy was used instead, as it caused less discomfort to the patients. The endotracheal tube was then removed. Postoperatively, lumbar spinal drainage was continued for 5 days and antibiotic cover for 1 week. No oral intake was permitted for at least 48 hours, following which tube feeding was supplemented by clear fluids taken by mouth. A diet of soft foods was started approximately 5 days postoperatively, but no milk products were allowed. Once the wound healed, the tube was removed and the patient returned to a normal diet.

Operative Results

Symptom Relief

Airway obstruction was completely relieved in all cases presenting with this problem. Pain relief was obtained in five of the eight cases in which pain was a feature. The neurological status was improved in two cases and unchanged in the others. No patient experienced a postoperative increase in neurological deficits. Postoperative discomfort was slight and settled quickly. Cosmetic results were excellent, and there were no significant problems related to malocclusion.

Complications

One patient developed a transient unilateral sixth nerve palsy which resolved completely within 3 days. Two other patients had a poor cough and required insertion of a temporary minitracheotomy for tracheal toilet within 48 hours of operation.

Postoperatively, one patient developed pneumonia secondary to aspiration. She had presented with dysphagia and her chest x-ray film showed aspiration of barium from tests carried out prior to admission. As a result of the pneumonia, she developed *Pseudomonas* septicemia and died 18 days after surgery.

Deaths

There was one postoperative death, as described above. Another patient who underwent two Le Fort I procedures for recurrent clivus chordoma died 4 months after the second operation from respiratory arrest due to brain-stem involvement by tumor.

Follow-Up Findings

The follow-up period ranged from 1 month to 36 months after the first Le Fort I procedure. Three of the 10 patients developed recurrent tumor requiring repeat operation, which was performed without increased difficulty by removing the compression plates and dropping the maxilla again. Two of these patients had recurrent chordoma, and the other had a recurrent chondrosarcoma (Table 2). Seven of the surviving eight patients are well; one underwent partial excision of a ninth nerve neurilemoma and is known to have residual tumor, but her symptomatology remains unchanged. One patient with an extensive and aggressive chondrosarcoma, which had been operated on three times in the 18 months prior to her Le Fort I procedure, developed recurrent pain 3 months after surgery, with a bulging mass in the hard palate; however, no tumor was identified in the skull base. She is now abroad, and has not returned for review.

Discussion

The latest improvements in imaging technology and advances in surgical equipment have led to a new interest in tumors of the skull base. By virtue of their nature these tumors warrant radical resection and, because of their tendency to recur locally, the ideal operation should offer the opportunity for repeat surgery with only minimal increased difficulty should the need arise.

Over the years, various methods have been adopted to deal with skull-base tumors. From a neurosurgical point of view, transcranial approaches via the middle
or posterior fossa to the skull base have been the most common. These are, of necessity, intradural techniques, none of which is entirely satisfactory in terms of access because, although most of them give limited access to the midline, it is very difficult to proceed beyond that point without risking major damage to neurovascular structures. The frontotemporal or pterional approach has been described for aneurysms of the circle of Willis and has also been used for rostral clival tumors with lateral extension into the middle fossa and for tumors located in and around the anterior clinoid processes extending anteriorly. With such a limited approach, there is clearly a considerable risk of damaging vital structures when dealing with large tumors. In reality, access to the clivus is restricted to the region of the posterior clinoid processes. The frontozygomatic approach has been reported recently as offering improved exposure in this same region and we have been impressed by the increased access it gives to the anterior central skull-base region; however, we find it is not effective for lesions of the clivus.

The best transcranial approach to the upper part of the clivus is via the subtemporal transtentorial approach, and if necessary the posterior limb of this incision can be extended over the squamous part of the occipital bone so that a lateral suboccipital craniectomy can be performed to permit an even more extensive resection. Great care must be taken throughout to preserve the vein of Labbé, and Malis has devised a resection. The advantages of the technique are those of any of the transcranial middle fossa approaches, namely the risk of brain retraction causing aphasia and epilepsy due to temporal lobe retraction. These deficits can be minimized by frequent relaxation of the retractors for 5 of every 20 minutes, but this does lead to a tiresome disturbance of concentration. The other hazards of the procedure are those involved with damage to cranial nerves and branches of the vertebrobasilar vessels, which can often cause permanent and disabling complications. These latter complications can also bedevil the suboccipital approach through which it should be possible to expose the lower end of the clivus down to the region of the foramen magnum. The lower cranial nerves lie between the operator and tumor, and manipulation or repeated minor trauma during a prolonged operation could lead to loss of function. Medial perforating vessels could be injured in a similar manner with even more disastrous results. Clearly, if the tumor involves the nerve roots and the vertebral arteries, there is little that one can do to effectively remove it whatever the approach to the lesion.

In addition to the intradural approaches, there are several extradural lateral approaches. The transcochlear approach was introduced by House and Hitselberger; this technique was accomplished by extending in an anterior direction the translabyrinthine opening they had popularized for operations in the cerebellopontine angle. This required mobilization of the facial nerve and would extend centrally as far as the ipsilateral internal carotid artery. The method is said to be useful for tumors of the petrous tip, as well as tumors arising directly from the clivus. Although suitable for small lesions, access would appear to be too restrictive to facilitate the removal of large lesions. The operation automatically results in ipsilateral deafness and a high risk of facial paraesthesia; thus, there is a considerable morbidity, which can be reduced by using other techniques. The advantage of the method is the same as for the other lateral approaches in that morbidity associated with cerebellar and brain-stem manipulation is usually avoided. The infratemporal fossa approach described and popularized by Fisch and others also involves unilateral deafness, a fair risk of facial paraesthesia, and loss of facial sensation due to division of the branches of the fifth cranial nerve. In common with similar approaches, this technique is best for tumors laterally situated on the clivus. In over 100 cases treated with the infratemporal fossa approach, there was a 7% complication rate of dural fistulae leading to a CSF leak, and approximately half of these patients developed meningitis. The lateral transtemporal transphenoidal approach to the skull base avoids the many problems that may accompany the transcochlear and infratemporal approaches, and the tedious dissection associated with the latter two techniques is replaced by a simpler and more straightforward procedure. Although it is essentially an extradural method, it could easily be adapted to deal with intradural pathology. It offers the same advantages as the frontozygomatic approach in that the zygoma can be mobilized and rotated out of the surgical field and, in addition, the temporomandibular joint can likewise be retracted inferiorly to provide access to the infratemporal region. The report describing the method by Holliday records four clival dissections in the series. The most common complications (in equal numbers) were facial numbness in the third division of the trigeminal nerve due to its section, mild self-limiting trismus, and loss of eustachian tube function.

The other lateral approach is via a high cervical incision involving mobilization of the distal portions of the extracranial carotid vessels, which may thereby be damaged, and the sacrifice of the upper cervical nerves. The technique was one of the earliest to be employed in the approach to the clivus, and it has been described again recently. Modifications of this technique intended to facilitate the surgical attack include labiomandibulotomy, but even with this extra space, the upper clivus must remain remote. The initial advantage of the lateral extradural method was that CSF leaks occurred into a sterile environment and not into the contaminated nasal or oral cavity. This argument has been largely countered by the efficacy of modern antibiotics, which has rendered meningitis less dangerous and also by the fact that CSF seals are now very effective.
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difficulty that would be experienced if an attempt was made to repeat the operation. There would be considerable difficulties with dissection through scar tissue, and it is perhaps significant that repeat ventrues are not recorded in papers on the methodology of these approaches.

On logical and anatomical grounds, it would seem that an anterior transclival exposure would be superior to the lateral techniques. The direct transoral approach was first used for the drainage of retropharyngeal abscesses, but since then has been successfully used in a variety of conditions, including resection of the odontoïd for basilar impression, or atlanto-axial dislocation, congential cranial spine anomalies, excision of extradural and intradural tumors, and clipping of verteobasilar aneurysms. A number of modifications have been made to the technique to improve access, namely splitting of the palate or division of the mandible. Even with those improvements, access to the clivus is limited by retracted soft tissue, particularly by lateral retraction of the divided soft palate, and this interferes with exposure of the upper reaches of the clivus. If the dura has been opened, difficulties occur in obtaining a watertight seal, with consequent risk of meningitis. Such problems have led some authors to suggest that only extradural pathology should be tackled via this approach.

A transsphenoidal approach gives access only to the region of the sella and the uppermost part of the clivus. The technique of this elegant operation is well known. We have had experience of quite severe hemorrhage from the sinusoids in the dura at the upper part of the clivus with control rendered more difficult by the limited exposure when operating via the transsphenoidal route, and would agree with the suggestion that the exposure is only suitable for biopsies of soft tumors in the middle and upper parts of the clivus. The transbasal approach of Derome is said to provide access to tumors of the ethmoid sinuses and to the middle and lower parts of the clivus down to the arch of the atlas. The only part of the skull base that cannot be operated on is the region of the sella and that part of the clivus which is shielded by it when viewed from the anterior fossa. Although attractive in theory, it would appear that dural tears are bound to occur when stripping dura from the cribiform plates. Dissection of the lower part of the clivus must be difficult in view of the distance from the surface. The method, as described, calls for reconstruction of the skull base with autologous bone grafts, but we have not found this to be necessary, and indeed it would seriously restrict the capacity for maneuver should further surgery be necessary.

The midfacial degloving operation as described by Price offers an extensive approach to the central skull base. A wide range of lesions have been attacked using this method, including three clival resections. Complications are minor, consisting chiefly of infraorbital and dental sensory loss which usually resolves. The operation was repeated for recurrent pathology on three occasions without additional comment, implying that there were no insuperable problems.

The Le Fort I level osteotomy was first described by David W. Cheever in 1867 for excision of a nasopharyngeal tumor, although in 1859 von Langenback had described a slightly less extensive approach. It is frequently used in maxillofacial surgery to expose tumors of the nasopharynx or to correct deformities of the midfacial region, and complication rates are low. There have been reports of postoperative hemorrhage, subcutaneous emphysema, and unilateral abducens nerve palsy following the procedure, but these accounts are uncommon. One of our patients developed a transient left sixth nerve palsy, but this resolved completely.

The downward displacement of the maxilla provides a direct line of vision to the clivus and a good view laterally without obstruction by retracted soft tissues. This facilitates both the approach to the tumor and the closure of the defect. In addition to the excellent exposure provided by the Le Fort I osteotomy, two further aspects of the technique help to minimize the risk of CSF fistula. Continuous lumbar spine drainage for 5 days postoperatively maintains a low CSF pressure, and no difficulties have been encountered with leakage at the drainage site or with infection via that route. Human-derived fibrin adhesive provides rapid watertight closure without necessitating the difficult insertion of sutures at the depth of the operative field. There have been no cases of wound infection or dehiscence, and no CSF fistulae have developed since its use.

This technique allows good access to tumors in or around the clivus with a much reduced risk of wound breakdown and CSF fistula. In our hands, both extradural and intradural tumors have been successfully operated on by this approach. Wound healing is rapid, with little discomfort to the patient and excellent cosmetic results.

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