Low-Grade Fibrosarcoma of the Anterior Skull Base: Endoscopic Resection and Repair

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ABSTRACT

Background: *Fibrosarcomas of the paranasal sinuses and skull base are uncommon tumors. Traditionally, "open approach" surgery remains the mainstay for treatment of choice for these tumors.*

Methods: *A 49-year-old man underwent resection of a right anterior skull base fibrosarcoma using the endoscopic approach.*

Results: *Close follow-up using both endoscopic and imaging methods over a period of four years has revealed a well-healed skull base with no evidence of recurrence.*

Conclusion: *Significant resistance exists at present for such a technique to deal with malignant diseases of the head and neck, but results from advanced centers continue to prove that this may be a technique worth mastering and improving on.* (American Journal of Rhinology 17, 347–350, 2003)

Fibrosarcomas of the nasal cavity, paranasal sinuses, and skull base are uncommon tumors. Only a few scattered small series and case reports have been published.[1–3] We are not aware of a single published case report of an anterior skull base fibrosarcoma. The largest series of sinonasal fibrosarcomatous neoplasms consists of 67 patients accumulated over 30 years in the files of the Otolaryngic Pathology Registry at the Armed Forces Institute of Pathology.[3] Another case series from the Institute Gustave Roussy reports a total of 42 patients with sarcomas of the nasal cavity and paranasal sinuses over a period of 33 years. Only 16 of these patients had fibrosarcomas and none originated from the skull base.[2] Surgery remains the mainstay for treatment of choice for these tumors. Traditionally, such tumors in a similar location would be managed using an "open approach" such as a craniofacial resection or a subfrontal approach with the aim of obtaining an *en bloc* resection. Adjuvant radiotherapy and chemotherapy are used in cases of incomplete excision. We report a patient with right anterior skull base fibrosarcoma involving the entire roof of the ethmoid cavity. The lesion was excised in total using the endoscopic approach. No adjuvant radiotherapy or chemotherapy was used postoperatively.

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CASE REPORT

A 49-year-old truck driver from North Carolina presented to his general practitioner with a chief complaint of frontal headaches. He was found to be hypertensive and was treated medically. The hypertension and headaches were corrected adequately, but magnetic resonance imaging (MRI) was done to rule out intracranial pathology. The MRI showed a bony lesion of the right anterior skull base/ethmoid roof with associated mucosal thickening and localized polyposis. He was referred to the local otolaryngologist who performed a septoplasty and polypectomy with biopsy of the bony lesion. A preliminary histological diagnosis of low-grade osteofibrosarcoma was made. Therefore, the patient was referred to the Georgia Nasal and Sinus Institute for definitive therapy. A 1-mm axial computed tomography scan with coronal and sagittal reconstruction revealed a bony lesion extending from the anterior ethmoid artery to the roof of the sphenoid sinus posteriorly and from the lateral cribriform plate lamella and middle turbinate to the medial orbital wall laterally (Fig. 1). The entire right ethmoid roof measuring ~4 cm front to back and 1.5–2 cm from side to side was involved. Office endoscopy revealed bony shelves in the ethmoid cavity and a bony lesion superiorly at the level of the anterior ethmoid artery with a white round "cyst-like" structure anterior to the tumor at the skull base (Fig. 2). An MRI revealed no involvement of the dura and investigations for metastasis were negative. Endoscopic resection of the tumor was planned as a two-stage procedure. The first stage was designed to complete an

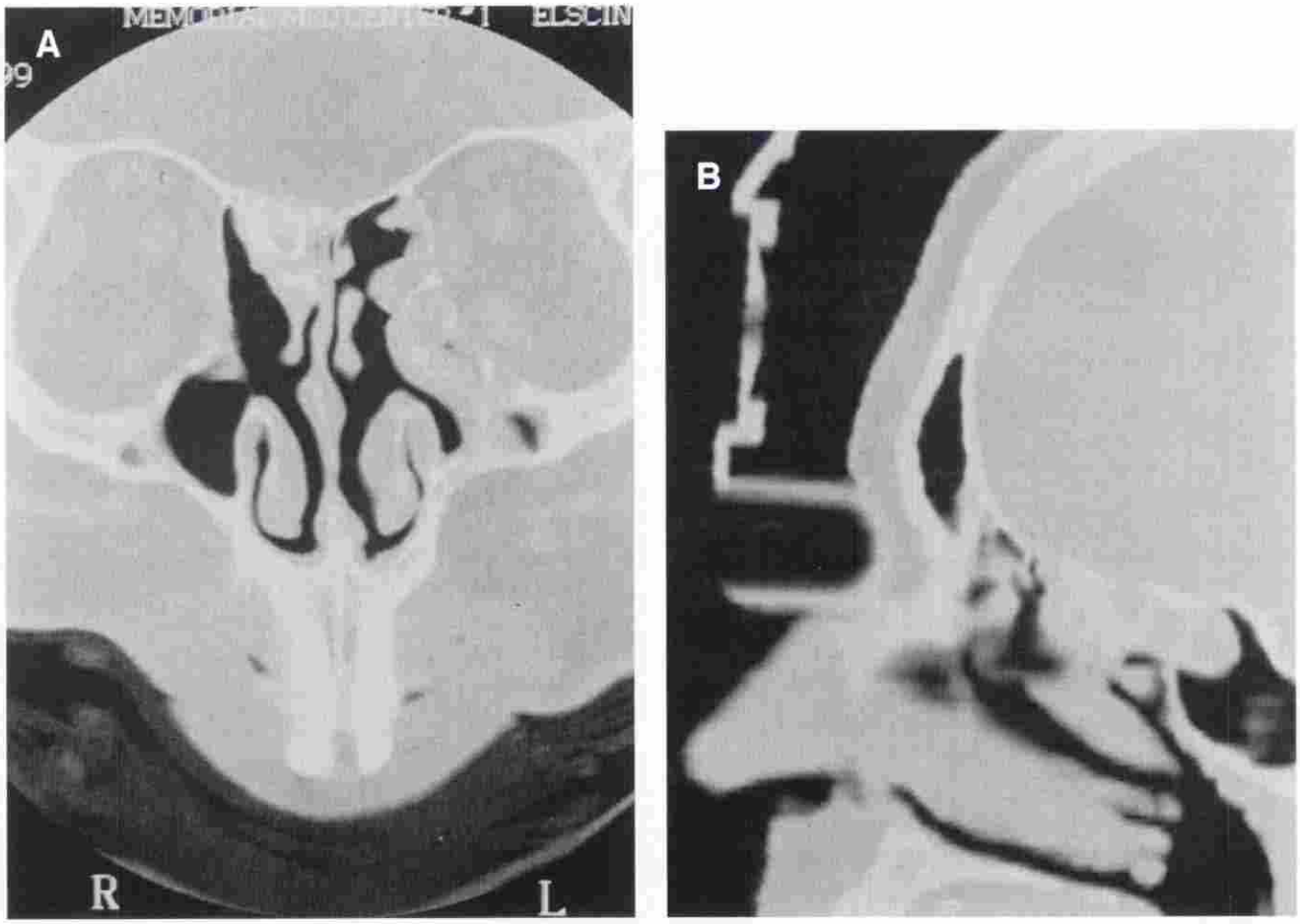

Figure 1. *CT showing tumor on an (A) coronal plane and (B) on sagittal reconstruction.*

endoscopic frontal sinusotomy, anterior and posterior ethmoidectomy, and sphenoidotomy as well as to debulk the skull base in preparation for the actual skull base tumor resection and repair during the second stage. During the first stage, the right anterior ethmoid artery was ligated also *via* an external approach so that endoscopic transection of the artery during tumor resection would be possible without unfavorable consequences. During the first stage, the fibrotic skull base lesion was removed in piecemeal fashion from anterior to posterior, leaving an intact skull base to avoid a cerebral spinal fluid (CSF) leak. The anterior white "cyst-like" structure was identified as an encephalocele during the first procedure. A fleshy mucocele identified and resected from within the anterior-superior aspect of the sphenoid sinus was positive for sarcoma on frozen section. Stereotactic computer-assisted navigation was used throughout the procedure.

Two weeks later the patient was taken back to the operating room for a completion procedure. A definitive resection of the skull base was performed from the sella turcica to the anterior ethmoid artery followed by repair of CSF leaks and reconstruction of the skull base. The skull base was removed using a Medtronic debrider (Xomed Surgical Products, Jacksonville, FL) fitted with a straight diamond burr to thin the bone and create posterior, medial, and lateral grooves at the margins of the resection. Then, the bony lesion was removed from posterior to anterior. Two CSF leaks occurred, one anteriorly at the level of the anterior ethmoid artery and one posteriorly at the margin of the septum and ethmoid roof.

Skull base reconstruction was performed using temporalis fascia and bone from the nasal septum. The dura was elevated around the periphery of the skull base defect and a 1.5×5 cm temporalis fascia strip was draped over the dura covering both dural defects. It was tucked between dura and bone around the margins of resection. Then, four pieces of septal bone were used to reconstruct the skull base. Three pieces were placed horizontally across the defect, which were held in place by the fourth piece, a narrow strut extending from the orbital margin anteriorly to the medial-posterior aspect of the septal margin posteriorly. Another piece of fascia was draped over the bone grafts and a mucous membrane graft obtained from the floor of the right nose then was placed over the fascia layer. Finally, the nose was packed with Gelfoam (Pharmacia & Upjohn Co., Kalamazoo, MI), followed by a layer of Gelfilm (Pharmacia & Upjohn, Co.). Then, Merocel 3000 sponges (Medtronic Xomed Surgical Products) were stacked to the floor of the nose for support. A lumbar drain was placed for 48 hours. The patient was discharged home on postoperative day 6.

Figure 2. Preoperative endoscopic view of right ethmoid roof. White cyst-like encephalocele is seen clearly.

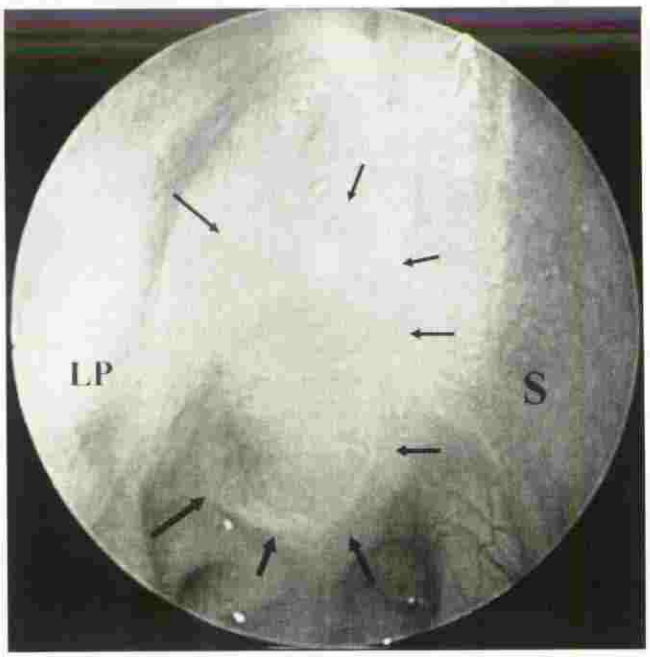

Figure 3. One-year postoperative endoscopic view of healed right skull base. Mucosal graft is seen clearly delineated from surrounding tissue.

Postoperatively, the patient did extremely well. All biopsy specimens taken at the margin of the resection were negative for malignancy and therefore no additional adjuvant treatment was deemed necessary. The patient returned to work 1 month after surgery. An MRI and computed tomography scan done at 4 months showed no evidence of disease with good support of the anterior skull base. The patient is now 4 years postoperative with no evidence of local or distant recurrence. One year postoperative endoscopy revealed a clean, well-healed skull base with the mucosal graft nicely delineated from the surrounding tissue (Fig. 3). Endoscopic views remain unchanged at the 4-year postoperative follow-up (Fig. 4). A final histopathological diagnosis of low-grade fibrosarcoma of the anterior skull base was obtained. An MRI obtained at 2.5 years postoperative reveals no evidence of intracranial or intranasal recurrence with a well-healed skull base (Fig. 5*A* and *B*). A repeat nonenhanced and enhanced gadolinium MRI scan obtained at the 4-year follow-up also revealed no evidence of intracranial or intranasal recurrence (Fig. 6)

Figure 4. Endoscopic view of the frontal recess at 4 years postoperatively.

DISCUSSION

Sarcomatous lesions of the sinonasal cavity account for 7–28% of all sarcomas of the head and neck region.[4,5] Fibrosarcoma is the commonest type of soft tissue sarcoma in the head and neck region and involves paranasal sinuses more frequently.[6,7] As seen in our patient, most sinonasal fibrosarcomas have an extremely low mitotic rate. Nuclear pleomorphism or anaplastic features usually are not prominent.[3] Because of these relatively bland histological features, there is a risk of these tumors being diagnosed benign and therefore being undertreated. There is a high rate of local recurrence with reduced risk of distant metastasis. Death usually results from intracranial extension of uncontrolled local disease.[8] The 5-year survival in patients with fibrosarcoma has been reported at 21%.[2,9] The sinonasal region has a poorer prognosis than the skin and soft tissue for two reasons: first, the more indolent nature of presentation and, second, the greater difficulty in obtaining tumor-free margins.[10]

Surgery remains the mainstay for the treatment of choice of these tumors. Wide local excision with radical margins has been recommended.[2,11] With continued advancements

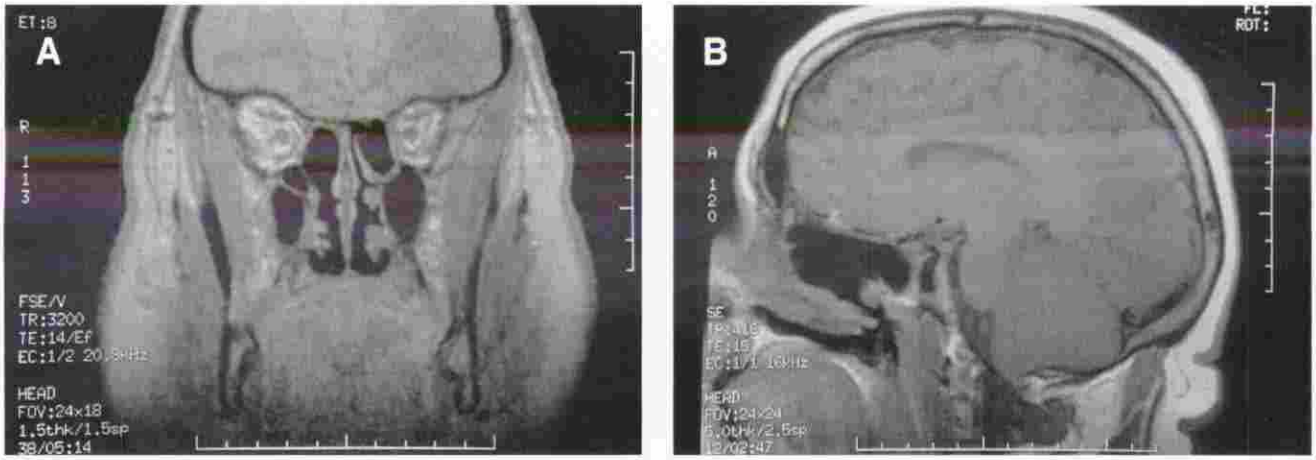

Figure 5. *MRI showing a well-healed skull base in the (A) coronal and (B) sagittal plane with no evidence of tumor recurrence at 2.5 years postoperatively.*

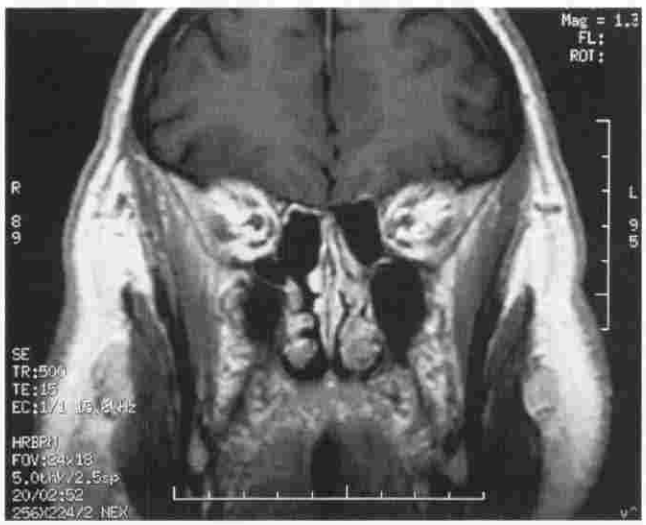

Figure 6. *MRI showing no evidence of tumor recurrence at 4 years postoperatively.*

in endoscopic surgery and image guidance, endoscopic removal of such tumors is being attempted at advanced centers with acceptable results. The greatest advantage of endoscopic surgery is the reduction in morbidity by avoidance of a major external procedure and, therefore, quicker recovery. However, the endoscope does not allow for an *en bloc* resection with radical margins. Instead, it results in a piecemeal excision of the tumor until it is excised adequately, at which point frozen section pathology can be obtained for margins, not unlike Moh's surgery for skin cancers. There is significant resistance at present for such a technique to deal with malignant diseases of the head and neck, but results from advanced centers continue to prove that this may be a technique worth mastering and improving upon.

Adjuvant radiotherapy has been recommended only when excisions were incomplete or for unresectable disease. For patients with macroscopically incomplete excisions, adjuvant radiotherapy has been shown to improve the survival rate significantly.[2]

In our patient, complete endoscopic resection of the tumor with free margins on the frozen section was obtained. Therefore, further adjuvant therapy was withheld. It must be noted that the follow-up period of 4 years as of this study may be insufficient to exclude a recurrence. Close endoscopic and radiological follow-up therefore has been maintained (Figs. 4–6).

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