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# Extracranial Meningiomas of the Head and Neck

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Extracranial extraspinal meningiomas (less than 1% of all meningiomas) frequently occur in the head and neck. The radiographic findings of extracranial meningioma of the head and neck are not pathognomonic. However, the location of the tumor in the pathway of cranial nerves or adjacent to the skull base, the demonstration of a well circumscribed, solid, enhancing mass by computed tomography, and the faint tumor stain on arteriography should allow the radiologist to make a possible, although not definite, diagnosis of this rare tumor. Detection of possible intracranial extension and involvement of adjacent structures is mandatory before surgery.

Meningiomas originate from meningocytes (arachnoid or meningotheial cells) capping the arachnoid villi or pacchionian granulations [1]. Meningiomas constitute about 15% of all intracranial and 12% of intraspinal neoplasms. Outside the central nervous system, meningiomas are rare. Of 405 meningiomas reported from Memorial Sloan-Kettering Cancer Center, New York City, Farr et al. [2] considered that only three (less than 1%) were primary extracranial extraspinal meningiomas. Whicker et al. [3] reviewed 1768 patients with meningiomas and found 16 (about 1%) of the tumors to be extracranial and extraspinal.

We review the literature and describe the radiologic features of extracranial meningiomas of the head and neck in five patients.

## Materials and Methods

This series of five patients with extracranial meningiomas of the head and neck comprised four women and one man 19–44 years old. Presenting symptoms included pain, localized headache, hoarseness, nasal congestion, and ptosis. Neurologic examination revealed deficits in cranial nerves IX, X, XI, and XII.

Conventional radiography of the paranasal sinuses included posteroanterior (Caldwell), occipitomental (Waters), basal, lateral, and semiaxial views. Multidirectional tomography of the paranasal sinuses was done in the supine position at 5 mm intervals from the nasion to the clivus. Tomographic examination of the base of the skull was performed at 3 mm intervals. Computed tomographic (CT) scans in the coronal and axial views were obtained at 5 mm intervals through the base of the skull and paranasal sinuses before and after intravenous injection of iodinated contrast material (42.5 g I). Selective internal and external carotid arteriograms were obtained in four of the five patients.

## Case Reports

### Case 1

A 41-year-old woman had a history of right-sided headache that dated back to her teens and had been getting worse over the past several years. She also had a right-sided nasal obstruction without any change in her sense of smell. Three weeks before admission, a right nasal polyp had been found; excisional biopsy revealed a meningioma. The neurologic examination was normal with no cranial nerve deficit.

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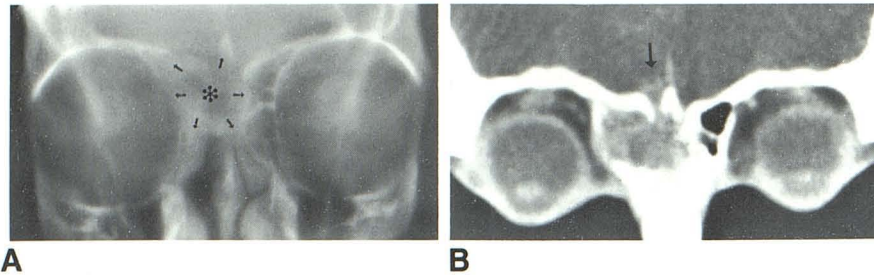


Fig. 1.—Case 1. **A**, Multidirectional tomogram shows soft-tissue mass (\*) in right nasal fossa and ethmoid sinus eroding cribriform plate, base of crista galli, and lamina papyracea (arrows). **B**, CT in direct coronal plane demonstrates well intracranial extradural extension of tumor (arrow).



Fig. 2.—Case 2. **A**, Multidirectional tomogram of skull base shows reactive sclerosis of right jugular foramen without destruction (\*). **B**, Right external carotid arteriogram shows large, well defined vascular mass (arrowheads) displacing right external artery and its occipital branch (arrows). Tumor encasement of occipital branch can be appreciated.

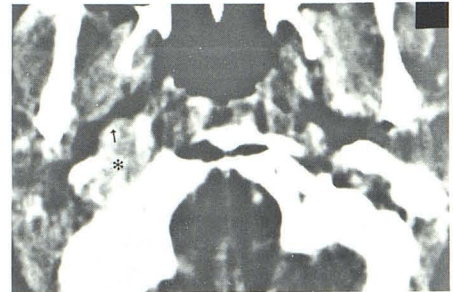


Fig. 3.—Case 3. Solid, enhancing mass (\*) arising from right jugular foramen and encroaching on parapharyngeal space (arrow).

Conventional radiographs demonstrated a soft-tissue mass in the right frontal sinus. Multidirectional tomography of the paranasal sinuses revealed a tumor mass in the superior part of the right nasal fossa and frontal and ethmoid sinuses, with erosion of the cribriform plate and thinning of the lamina papyracea (fig. 1A). CT disclosed the small intracranial extradural extension of the tumor (fig. 1B) with no definite intracerebral or intraorbital involvement.

The patient underwent craniofacial surgery with complete resection of a gray, firm, polypoid mass in the right nasal fossa and frontal sinus. Histopathology revealed meningioma. The patient's postoperative course was uneventful except for the loss of smell in the right nasal passage.

#### Case 2

A 19-year-old man had ptosis of the right eyelid and constriction of the right pupil 6 months before admission. He also had noted hoarseness and a mass in the right side of the neck. Physical examination revealed a large, firm, fixed, retromandibular mass, Horner syndrome on the right side, and right vocal cord paralysis.

Multidirectional tomograms of the base of the skull detected reactive sclerosis of the base of the skull at the right jugular foramen (fig. 2A). CT showed a large, solid, enhancing tumor mass medial to the right mandible distorting the right parapharyngeal space. The carotid sheath could not be identified. A separate small, solid mass that was interpreted as an enlarged lymph node was identified posteromedial to the atrophic sternocleidomastoid muscle. Arteriography demonstrated anterior displacement of the right internal and external carotid arteries by a faintly hypervascular mass that also displaced and encased the occipital artery (fig. 2B). The mass ex-

tended from the base of the skull, near the jugular foramen, down to the level of the hyoid bone. Internal jugular venography was not performed.

At surgery, a large, yellow gray, rubbery mass fixed to the carotid sheath and involving the eleventh and twelfth cranial nerves and possibly the sympathetic trunk was resected. The tumor occluded the internal jugular vein at the jugular foramen. Histologic examination revealed a meningioma with extensive perineural invasion. The specimen of the enlarged lymph node was diagnosed as reactive follicular hyperplasia.

#### Case 3

A 27-year-old woman was seen at the University of Texas M. D. Anderson Hospital and Tumor Institute in Houston with a 2 year history of right-sided facial pain and hoarseness. Recurrent right laryngeal nerve paralysis was detected by physical examination.

A CT scan of the base of the skull and neck disclosed a well circumscribed, solid, enhancing mass extending from the right jugular foramen to the angle of the mandible, encroaching on the parapharyngeal space posteromedially (fig. 3A). Right external carotid arteriography demonstrated a hypervascular tumor lateral to the right lateral masses of the C1 and C2 vertebrae, partly obstructing the internal jugular vein just below the jugular foramen. At surgery, a firm, fusiform, encapsulated tumor was readily dissected from the base of the skull at the jugular foramen. There was some fixation to the tenth and twelfth cranial nerves. The pathologic diagnosis was meningioma with extensive infiltration of the nerve sheaths. The patient's postoperative course was uneventful.



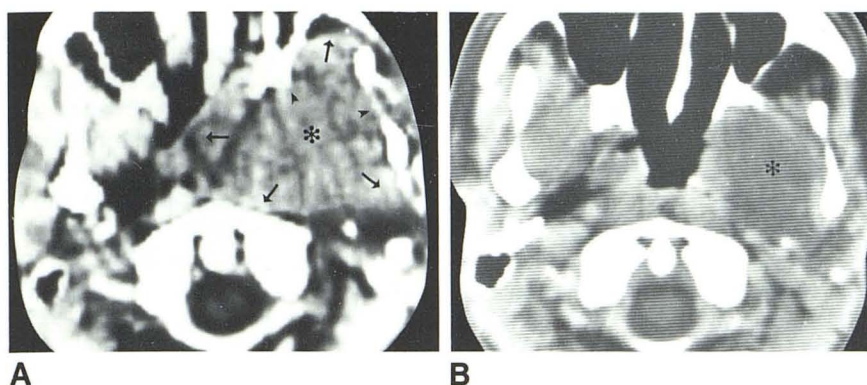


Fig. 4.—Case 4. **A**, Solid, enhancing mass (\*) in left infratemporal fossa and parapharyngeal space (arrows) encroaching on naso- and oropharynx. Remodeling deformity of left lateral pterygoid process and ramus of mandible (arrowheads). **B**, Lesion has diminished significantly with development of central necrosis after treatment. Only minimal encroachment of nasopharyngeal wall remains.

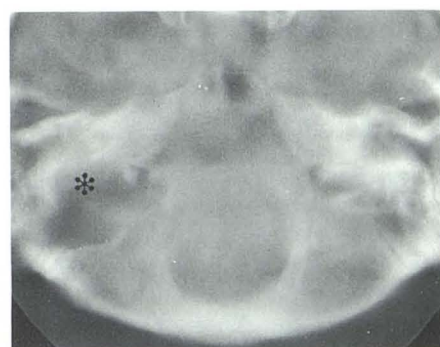


Fig. 5.—Case 5. Destructive lesion enlarging right jugular foramen and extending anterolaterally into petrous bone, medial mastoid air cells, ear structures, and lateral wall of foramen magnum (\*).

#### Case 4

A 24-year-old woman was admitted to the hospital because of nasal congestion, nasal speech, and snoring of 6 months duration. This had been preceded by epistaxis for 6 months. A left sphenoid wing meningioma removed 2½ years before had recurred and was subtotally resected 8 months before admission. For more than 1 year the patient had had difficulty in swallowing. On examination, her soft palate was found to be displaced inferiorly and deviated to the right. The buccal mucosa, lateral and posterior to the left upper molars, was severely edematous. A punch biopsy revealed a neoplasm of the same cell type as that of the previously resected meningioma, which subsequently had been diagnosed as a meningeal sarcoma.

Cranial CT showed chronic postoperative changes in the left temporal lobe. A large, solid, enhancing mass without calcification, extending along the parapharyngeal space to the level of the angle of the mandible and encroaching on the oropharynx and nasopharynx, was identified in the left infratemporal fossa (fig. 4A). A remodeling deformity was noted in the adjacent left pterygoid plate and in the border of the ramus of the left mandible. Left carotid arteriography revealed a complete obstruction of the M1 segment of the left middle cerebral artery, with leptomeningeal collateral circulation presumably related to the previously resected intracranial tumor. A large, heterogeneous, hypervascular mass supplied by minute branches of the left internal maxillary artery was opacified in the left parapharyngeal space from the infratemporal fossa to the level of C2.

The patient was then treated by radiotherapy and chemotherapy. Six months later, CT demonstrated a slight decrease in the size of the tumor, which exhibited central necrosis (fig. 4B).

#### Case 5

A 44-year-old woman was admitted with a history of persistent pain and progressive hearing loss in the right ear, with tenderness over the right mastoid area for 11 months. She had experienced intermittent headache, diplopia, and transient blindness for 7 months. One month before admission, she had noticed a large mass in the right side of her neck. Examination revealed a bluish discoloration of the right eardrum, with a mass bulging into the middle and external ear from the upper posterior quadrant. A soft, multilobulated, mobile, nontender mass of 5 × 2 cm was palpated underneath the sterno-

cleidomastoid muscle just below the right mastoid tip. The neurologic evaluation detected a low-grade chronic bilateral papilledema, dysfunction of the ninth cranial nerve, and a conductive hearing loss on the right side.

Multidirectional tomography of the skull base (fig. 5A) demonstrated a destructive lesion of the right jugular foramen extending anterolaterally into the petrous bone, the medial air cells of the right mastoid, and the medial posterior wall of the right external ear canal. The tympanic cavity was opacified, and the lateral wall of the foramen magnum and the right hypoglossal canal were also involved. Right common carotid arteriography revealed a slightly vascular mass from the region of the jugular foramen to the C1–C2 level, displacing the upper cervical segment of the internal carotid artery anteriorly and the proximal occipital artery laterally. The right sigmoid sinus, jugular bulb, and proximal internal jugular vein were not opacified. CT delineated the destruction of the skull base in the region of the jugular foramen by a tumor mass. There were no intracranial abnormalities. The patient underwent surgery and was found to have an encephaloid tumor that involved the tympanic cavity and floor of the external ear canal and that projected into the neck within the jugular vein. A second surgical procedure using a mastoidectomy approach was carried out shortly afterward for excision of all of the grossly visible tumor that was invading the sigmoid sinus, inferior petrosal vein, and adjacent occipital bone.

Under light microscopy, the tumor was considered to be a chemodectoma. However, ultrastructural examination by electron microscopy disclosed tumor cells containing many microfilaments, and most were consistent with the diagnosis of meningioma. Also noted were attenuated cytoplasmic extensions of the more elongated tumor cells, curving and undulating to loosely occupy slender clefts between adjacent cells, characteristic of meningiomas. The final diagnosis was an aggressive intracranial meningioma with extracranial extension.

The patient subsequently received radiotherapy. Follow-up CT 2½ years later showed local tumor recurrence. The patient refused further treatment and was lost to follow-up.

#### Discussion

The mechanisms for the formation of extracranial meningiomas suggested by Hoyer et al. [4] include four types:



*Type A* involves direct extension from a primary intracranial tumor through the foramina of the base of the skull [3, 5–13]. Sometimes, extracranial components of the neoplasm project through the destroyed or eroded floor of the cranial fossa [6].

*Type B* involves extracranial growth from arachnoid cells within the sheaths of cranial nerves. Most of the reported primary extracranial meningiomas in the parapharyngeal region of the neck are related to the cranial nerves, particularly VII [2, 3, 13–15], IX, X, XI, and XII [2–4, 9, 10, 13, 16–19]. Ectopic meningiomas arising far from the base of the skull usually occur in association with spinal [20] or sympathetic [21] nerve roots.

*Type C* involves extracranial growth from embryonic rests of arachnoid without any apparent connection to the foramina of the skull base or cranial nerves. Extracranial meningiomas are reported to arise from the outer table of the skull, most often from the frontal bone [12, 13, 22, 23] or the temporal bone [24–27]. Orbital meningiomas without involvement of the optic nerve have been reported [28, 29]. The most frequently described extracranial meningiomas belonging to this group are those in the paranasal sinuses and naso-oral cavity [3, 6, 8, 9, 11, 12, 19, 30–50].

*Type D* involves distant metastases from intracranial meningioma. There have been cases reported of intracranial meningiomas metastasizing to the liver [51, 52], lung [53–56], and, in our own experience, bone. At times these have been considered aggressive meningiomas or meningeal sarcomas; however, they may have no malignant histologic characteristics.

Eighty-four cases of extracranial meningiomas of the head and neck were compiled from a review of the literature. Thirty-nine cases originated in the paranasal sinuses and naso-oral cavity [3, 6, 8, 9, 11, 12, 19, 30–33, 35–50]; of those, 15 had documented intracranial extension [3, 6, 8, 9, 11, 12, 19, 33, 39, 46]. Nineteen cases were seen in the parapharyngeal regions [2–4, 9, 10, 13, 15–19, 57–61]; 15 of the 19 presented with definite involvement of cranial nerves VII, IX, X, XI, and XII. Fourteen cases were documented within the structures of the ear [3, 14, 62–68] and 12 cases originated from the outer table of the calvaria [12, 13, 22–27].

In our series, case 1 was most likely a meningioma arising from ectopic rests of arachnoidal cells without any obvious connection to the cranial nerves (*type C*). Despite the erosion of the cribriform plate, there was no olfactory deficit. The intracranial extension of the tumor was completely limited by an intact dura. Cases 2 and 3 should be grouped with those originating from the cells of the arachnoid of cranial nerves (*type B*). In case 2, cranial nerves X, XI, and XII and probably the sympathetic trunk were involved. In case 3, cranial nerves X and XII were within the tumor. Cases 4 and 5 were aggressive intracranial meningiomas with direct extracranial extension (*type A*). It is believed that case 5 originated intracranially and extended extensively into the petrous bone and venous sinuses without an obvious bulky intracranial mass.

In radiographic evaluation, extracranial meningiomas of the head and neck can be divided into two groups. In the first group are those meningiomas located in the nasal or oral cavity and in the paranasal sinuses where the lesions are

easily detected and are accessible for biopsy or complete excision. In these cases, radiographic evaluation before surgery is considered by some to be of no clinical importance. However, our review of the 39 reported cases in this group indicated that more than one-third of the patients also had intracranial extension. Radiologic demonstration of the intracranial involvement would change the surgical planning to a combined craniofacial approach. The lesion can usually be delineated by conventional radiographs. Multidirectional tomography can clearly define the involvement of the adjacent cranial fossa or orbit, in addition to the exact location of the neoplasm within the paranasal sinuses or naso-oral cavity. The advantage of CT is its ability to define bone as well as soft-tissue involvement. Direct coronal CT best detects intracranial extension. Most of the recently reported cases of meningiomas of the paranasal sinuses [9, 12, 13, 39, 60] were proved to have intracranial extension diagnosed by CT. We believe that direct coronal CT images provide the most pertinent preoperative radiologic information about possible intracranial involvement. Arteriography in this group of patients will be of academic interest only, although theoretically, a tumor stain should be present.

The second group includes those tumors in the parapharyngeal area and at or under the base of the skull. These neoplasms are neither palpable nor easily accessible for biopsy. Even when the tumor is diagnosed by biopsy as a meningioma, a detailed radiographic evaluation is still required before surgery can be contemplated to establish the specific location of the tumor and the extent of involvement of adjacent structures. Conventional radiographs of the base of the skull or neck are usually uninformative. Multidirectional tomographic findings offer variable degrees of assistance, depending on the location of the tumor and its relation to the base of the skull. Bone destruction is frequently seen when the meningioma originates at a foramen or fossa, and reactive sclerosis is common when the neoplasm is at or immediately adjacent to the base of the skull. CT is the most accurate and informative method used to define the tumor, which is usually well circumscribed and solid, with homogeneous contrast enhancement. Direct coronal images best demonstrate the relations and involvement of the base of the skull. Arteriography is definitely necessary to differentiate a meningioma from a chemodectoma, which is extremely vascular and necessitates a different surgical approach. A meningioma usually exhibits a well defined and prolonged faint tumor stain, which is not as intense as that seen in a chemodectoma, and frequently there are no predominant feeding arteries. Selective and sometimes superselective arteriograms are required to delineate the lesion. Subtraction technique may be mandatory to identify the faint tumor stain of a meningioma; however, the arteriographic findings are not specific and can also be seen in sarcoma, neurofibroma, lymphoma, or carcinoma metastatic to lymph nodes. Jugular venography may be of value in demonstrating extrinsic compression or direct tumor infiltration into the jugular bulb or internal jugular vein.

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