Radiology Quiz Case 2

Tom W. Geurts, MD; Peter J. F. M. Lohuis, MD, PhD; Frank A. Pameijer, MD, PhD; Michiel W. M. van den Brekel, MD, PhD; The Netherlands Cancer Institute/Antoni van Leeuwenhoek Hospital, Amsterdam, the Netherlands

A 48-YEAR-OLD MAN presented to the emergency department with a severe headache and was seen by the attending neurologist. A nonenhanced computed tomographic (CT) scan of the lower sections of the brain revealed a low-density nasopharyngeal tumor with sharply defined margins and without signs of bone erosion (**Figure 1**). An otolaryngologist was consulted. The patient also complained of hyponasal speech, and according to his wife, he snored nightly. There was no history of nasal obstruction or hearing loss.

Physical examination showed a submucosal tumor arising from the left side of the nasopharynx that extended the median and expanded into the oropharynx. There were no signs of cranial nerve palsy, and the appearance of the eardrums was normal.

Fine-needle aspiration and biopsy of the lesion were performed, but the findings were inconclusive. A T1-weighted magnetic resonance imaging (MRI) scan at the same level as that seen in Figure 1 demonstrated that the lesion had a very high signal intensity (**Figure 2**). A sagittal MRI scan (**Figure 3**) showed the tumor extending from the nasopharynx into the oropharynx. In this particular STIR (short tau inversion recovery) sequence, the lesion had a markedly low signal intensity.

What is your diagnosis?

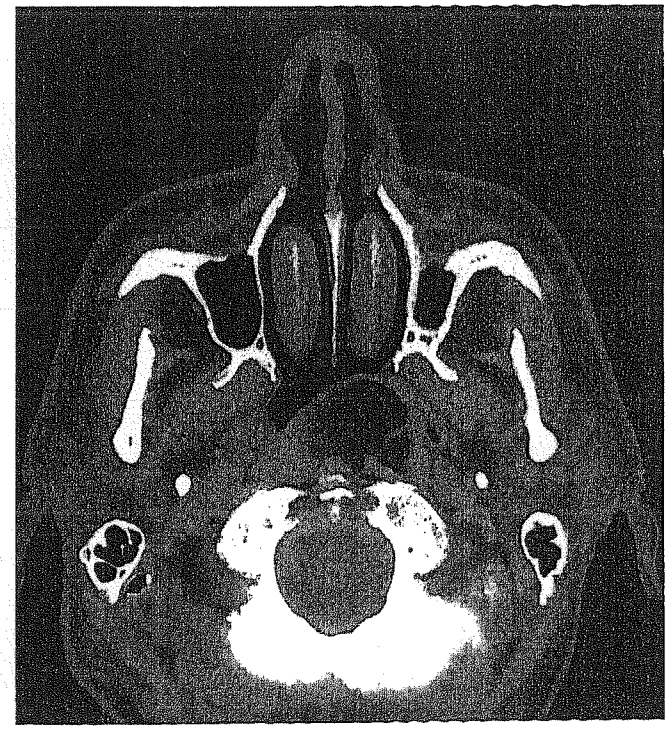

Figure 1.

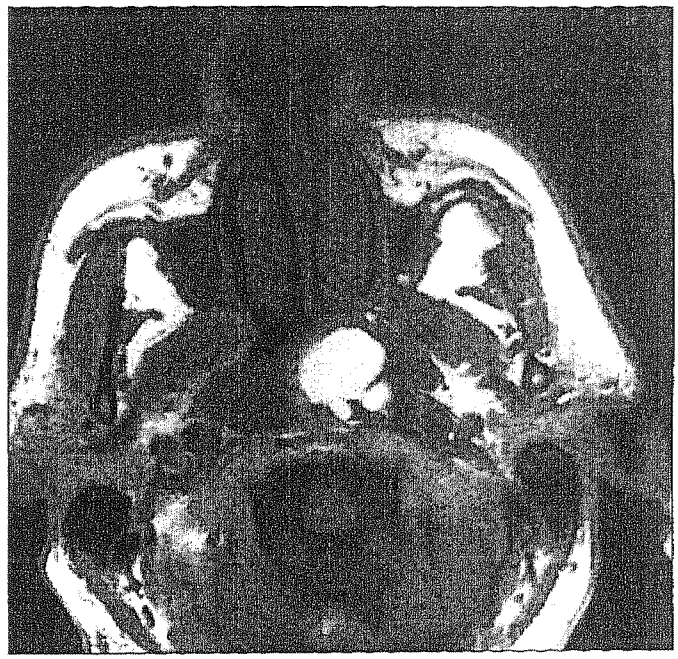

Figure 2.

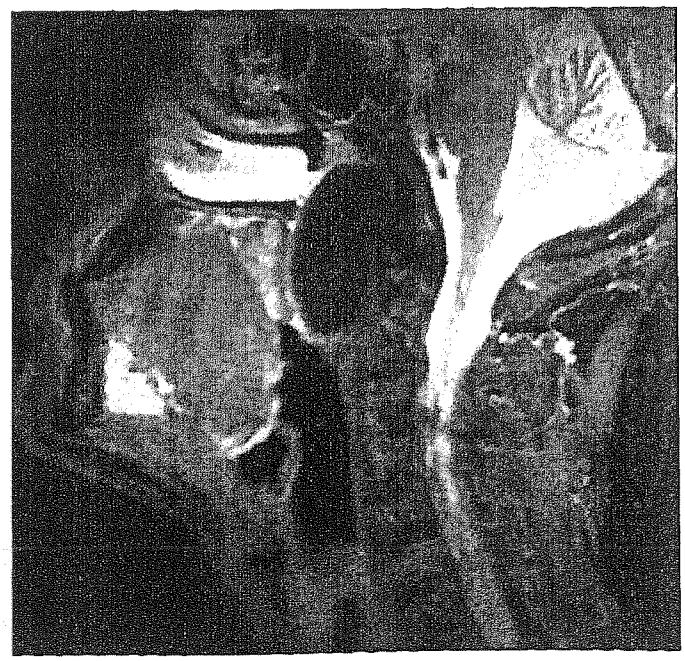

Figure 3.

Radiology Quiz Case 2

Tom W. Geurts, MD; Peter J. F. M. Lohuis, MD, PhD; Frank A. Pameijer, MD, PhD; Michiel W. M. van den Brekel, MD, PhD; The Netherlands Cancer Institute/Antoni van Leeuwenhoek Hospital, Amsterdam, the Netherlands

A 48-YEAR-OLD MAN presented to the emergency department with a severe headache and was seen by the attending neurologist. A nonenhanced computed tomographic (CT) scan of the lower sections of the brain revealed a low-density nasopharyngeal tumor with sharply defined margins and without signs of bone erosion (**Figure 1**). An otolaryngologist was consulted. The patient also complained of hyponasal speech, and according to his wife, he snored nightly. There was no history of nasal obstruction or hearing loss.

Physical examination showed a submucosal tumor arising from the left side of the nasopharynx that extended the median and expanded into the oropharynx. There were no signs of cranial nerve palsy, and the appearance of the eardrums was normal.

Fine-needle aspiration and biopsy of the lesion were performed, but the findings were inconclusive. A T1-weighted magnetic resonance imaging (MRI) scan at the same level as that seen in Figure 1 demonstrated that the lesion had a very high signal intensity (**Figure 2**). A sagittal MRI scan (**Figure 3**) showed the tumor extending from the nasopharynx into the oropharynx. In this particular STIR (short tau inversion recovery) sequence, the lesion had a markedly low signal intensity.

What is your diagnosis?

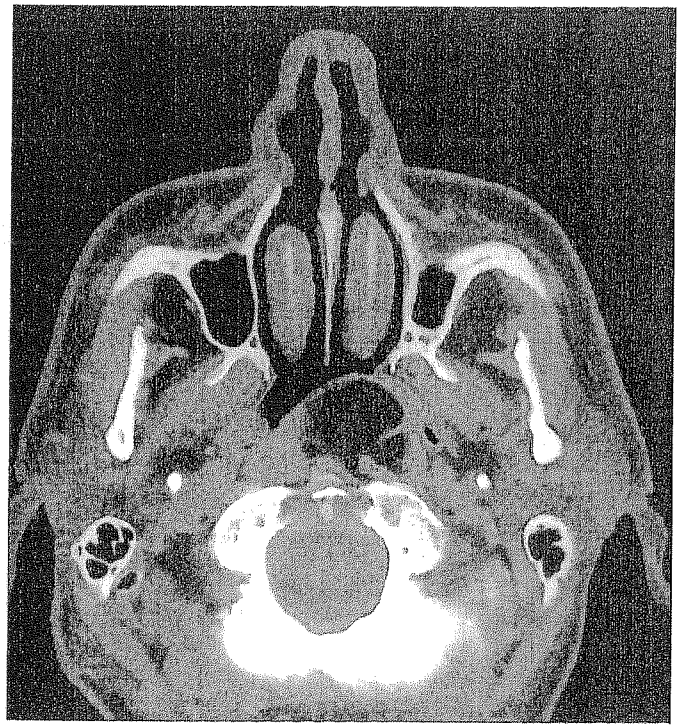

Figure 1.

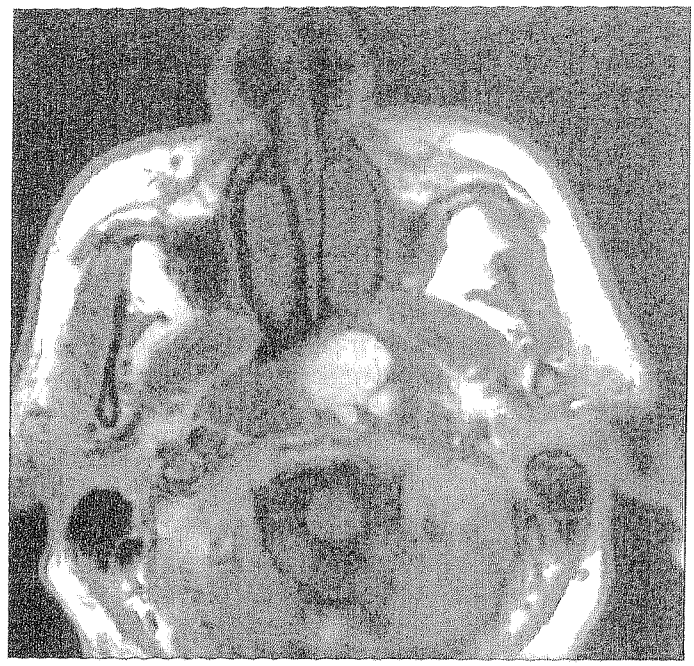

Figure 2.

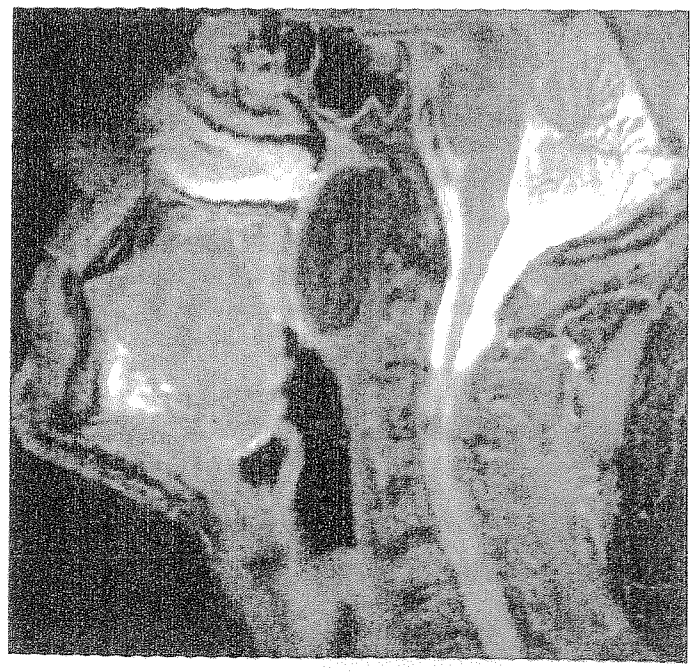

Figure 3.

Diagnosis Radiology Quiz Case 1

Diagnosis: Vascular leiomyoma of the larynx

Angiomyoma and *angioleiomyoma* are synonyms for vascular leiomyoma, but the latter is a more accepted term, because it is more accurately descriptive and widely used.[1] Vascular leiomyoma is an uncommon type of smooth muscle tumor and is rarely seen in the head and neck. In a review of 562 cases by Hachisuga et al[2] over a 17-year period, only 48 were seen in the head and neck. Most commonly, vascular leiomyomas arise in the female genital tract, gastrointestinal system, and pilar arrector muscles of the skin. Their exact pathogenesis is not known, but most authors agree that they probably originate in smooth muscle cells within blood vessel walls.[3,4] In the head and neck, vascular leiomyomas are found in the lips, auricle, nostrils, skin of the occipital region, and cheeks. They very rarely occur in the larynx.[5]

Most vascular leiomyomas present in middle-aged and elderly men; laryngeal subsites seem evenly represented. When the lesions occur in the larynx, they result in hoarseness or dyspnea. Pain and dysphagia are uncommon symptoms. The tumors are slow growing, and the duration of symptoms is reported to range from 2 to 16 years.[6] The most common symptom before admission is a rapidly progressive dyspnea, for which tracheostomy is often a necessary life-saving measure. Sometimes, however, a globus sensation, cough, or laryngeal pain is the only presenting symptom.[5]

Histopathologic classification of the vascular leiomyoma is complicated by its similarity to another benign laryngeal lesion, the simple leiomyoma. The World Health Organization recognizes 3 types of benign smooth muscle tumors: leiomyoma (simple leiomyoma), angiomyoma (vascular leiomyoma), and epithelioid leiomyoma (bizarre leiomyoma or leiomyoblastoma). The majority (7) of the 9 reported cases of laryngeal leiomyoma reported in the recent literature were vascular leiomyoma.[7]

Vascular leiomyomas are spindle cell neoplasms with numerous blood vessels but lacking mitoses. They have been reported to be encapsulated and made up of smooth muscle bundles among numerous vascular spaces.[6] The spindled fascicles stain strongly with actin, supporting the diagnosis of vascular leiomyoma. Degenerative changes such as fibrosis, calcifications, and giant cell reactions can also be present. The list of histopathologic differential diagnoses includes hemangioma, leiomyoblastoma, angiomyolipoma, and vascular leiomyosarcoma.

The pathogenesis of benign smooth muscle tumors is unknown. Duhig and Ayer[8] proposed that these lesions develop sequentially: smooth muscle proliferation within a vascular hamartoma leads to the formation of an angiomyoma, and continued smooth muscle proliferation results in a simple leiomyoma. Subsequent studies have refuted this theory of histogenesis, leaving the relationship between simple and vascular leiomyomas unclear.[9] An etiologic role for estrogen has been considered, but estrogens appear to have little influence on leiomyomas outside the uterus, in that such tumors have shown no female predominance.[7]

Submissions

Residents and fellows in otolaryngology are invited to submit quiz cases for this section and to write letters to the ARCHIVES commenting on cases presented. Quiz cases should follow the patterns established. See "Instructions for Authors."

Material for CLINICAL PROBLEM SOLVING: RADIOLOGY should be mailed to the Editor.

Reprints not available.

Morimoto[10] classified vascular leiomyomas into 3 histologic subtypes in 1973: solid (capillary), cavernous, and venous. Venous is the most common type found in the head and neck area. The solid type has smooth muscle bundles that surround the vascular slitlike channels. The cavernous type has dilated vascular channels with lesser amounts of smooth muscle. The venous type has vascular channels with thick muscular walls that are easily distinguished from smooth muscle bundles. In some lesions, features of all 3 types may be found.[10] Malignant variants of vascular leiomyomas are rare. The absence of mitosis is the most useful indicator of a benign lesion.[11]

Surgical extirpation is the treatment of choice for vascular leiomyomas. There are 3 reports of recurrent vascular leiomyoma of the head and neck, to our knowledge.[1] One was of a recurrent vascular leiomyoma of the larynx in which some mitotic figures had been noted histologically.[3] This suggests that complete resection of such lesions is important and that the patients involved should be monitored for recurrence on a regular basis after surgery. Some small tumors have been resected through microlaryngoscopy; larger tumors may require external approaches. If laryngeal vascular leiomyomas are treated endoscopically, Nuutinen and Syrjanen[6] recommend preliminary tracheostomy, general anesthesia, and, in the case of profuse bleeding, an external laryngeal approach.

Because of the tremendous vascularity of these tumors, there is a high risk of significant bleeding. Shibata and Komune[5] excised 2 small tumors through microdirect laryngoscopy. The patients involved lost a total of 600 to 1000 mL of blood during the primary procedure and from postoperative hemorrhage. Nuutinen and Syrjanen[6] used cryotherapy after partial resection to control profuse bleeding. Anderson and Weinstein[3] reported that the use of the carbon dioxide laser in the initial resection provided adequate hemostasis.

As surgical options are clearly not without risk, patients should be selected carefully. If there is evidence of malignancy, or compromise of the patient's airway or vocal quality, complete surgical excision is the recommended therapy. Because our patient was satisfied with the quality of his voice, and there were no findings to suggest a malignancy, we elected not to perform surgery. Instead, the patient will be followed up regularly over the upcoming years.

REFERENCES

1. Nall AV, Stringer SP, Baughman, RA. Vascular leiomyoma of the superior turbinate: first reported case. *Head Neck.* 1997;19:63-67.

2. Hachisuga T, Hashimoto H, Enjoji M. Angioleiomyoma: a clinicopathologic reappraisal of 562 cases. *Cancer.* 1983;54:126-130.
3. Anderson TD, Weinstein, GS. Recurrent angiomyoma (vascular leiomyoma) of the larynx after laser excision. *Otolaryngol Head Neck Surg.* 2000;123:646-647.
4. dEnzinger FM, Weiss SW. *Soft Tissue Tumors.* 2nd ed. St Louis, Mo: Mosby–Year Book Inc; 1983.
5. Shibata K, Komune S. Laryngeal angiomyoma (vascular leiomyoma): clinicopathological findings. *Laryngoscope.* 1980;90:1880-1886.
6. Nuutinen J, Syrjanen K. Angioleiomyoma of the larynx: report of a case and review of the literature. *Laryngoscope.* 1983; 93:941-943.
7. Hirakawa K, Harada Y, Tatsukawa T, Nagasawa A, Fujii M. A case of vascular leiomyoma of the larynx. *J Laryngol Otol.* 1994;108:593-595.
8. Duhig JT, Ayer JP. Vascular leiomyoma: a study of sixty-one cases. *Arch Pathol.* 1959;68:424-430.
9. Farman AG. Benign smooth muscle tumors. *S Afr Med J.* 1975;49:1330-1340.
10. Morimoto N. Angiomyoma (vascular leiomyoma): a clinicopathologic study. *Med J Kagoshima Univ.* 1973;24:663-683.
11. Soames JV, Moore UJ, Ord RA, McElroy JH. Angiomyoma of the temporal region. *Br J Oral Maxillofac Surg.* 1989;27:229-235.

Diagnosis Radiology Quiz Case 2

Diagnosis: Nasopharyngeal lipoma

Lipomas of the upper aerodigastric tract are rare. In the oral cavity, lipomas account for 2.2% of all benign tumors. More than half of these are located in the tongue, floor of the mouth, and lips. Lipomas have also been reported to occur in the lower pole of the tonsil, hypopharyngeal wall, aryepiglottic fold, retropharyngeal space, and parapharyngeal space.[1-4] To our knowledge, only 7 cases of nasopharyngeal lipoma have been described in the literature to date.[1-4]

Lipomas grow slowly and are often asymptomatic. If there are symptoms, they usually are caused by compression of adjacent structures. In the literature, patients with a lipoma in the nasopharynx complain of awareness of a nasopharyngeal mass in combination with dysphagia. Snoring, nasal obstruction, sleep apnea, and rhinolalia clausa have also been described.[1-4] Our patient had a history of snoring, sleep apnea, and rhinolalia clausa. His presenting symptom, the headache, had not previously been reported, although this particular symptom can occur in combination with parapharyngeal tumors and might be related to the obstructive sleep apnea syndrome.

Although it is difficult to diagnose a nasopharyngeal lipoma by clinical examination alone, the CT and/or MRI characteristics of these fat-containing tumors are almost pathognomonic. On a CT scan, the uniform low density of fatty tissue is very specific. Also, fatty tissue has a characteristic high signal intensity on T1-weighted MRI scans, while it has a characteristic low signal intensity on highly T2-weighted STIR (short tau inversion recovery) sequences. In our patient's case, the density (on CT) and signal intensity (on MRI) of the lipoma were comparable to that of the subcutaneous fat, as can be seen in Figures 1 through 3.

In comparison to lipomas, liposarcomas have a less uniform density (on CT) and signal intensity (on MRI).[5] Most other nasopharyngeal tumors have an intermediate density (on CT) or signal intensity (on MRI) and enhance after intravenous contrast administration. Malignant nasopharyngeal tumors usually have ill-defined borders, may cause bone erosion, and often result in lymphadenopathy. Because recognition of a nasopharyngeal lipoma on CT and MRI scans is straightforward, fine-needle aspiration is not necessary for diagnosis. However, surgical excision and histopathologic analysis are required to exclude liposarcoma.

Macroscopically, lipomas are pale yellow to orange with an uniform greasy surface. They tend to be well delineated from the surrounding tissues by a thin capsule. Microscopically, they are composed of mature fat cells, which vary slightly in size and shape. The cells have a single, centrally located fat vacuole and a peripheral nucleus without atypia.[5] Lipomas sometimes contain fibrous, myxomatous, cartilaginous, osseous, vascular, or myomatous elements and are then called *fibrolipomas, myxolipomas, chondrolipomas, osteolipomas, angiolipomas,* and *myolipomas,* respectively.

The malignant opposite of a lipoma, a liposarcoma, is divided into 3 groups: well-differentiated/dedifferentiated liposarcoma, myxoid liposarcoma, and pleomorphic liposarcoma.[5] In general, a liposarcoma of the larynx or pharynx is well differentiated. A liposarcoma consists predominantly of mature fat with a variable number of spindled cells (precursors of fatty cells) with hyperchromatic nuclei and multivacuolated lipoblasts.[5] To distinguish liposarcoma from lipoma, one should look for these features and for the presence of stromal cells with atypia. It can be difficult to distinguish well-differentiated liposarcoma from lipoma on histologic examination.[5] This applies especially to fine-needle aspiration cytology, but also to histologic examination of a biopsy specimen or complete excision specimen.

The therapy of choice for a nasopharyngeal lipoma (or liposarcoma) is surgical excision, particularly when there are symptoms. Our patient had clear symptoms of severe headache and wanted to exclude the possibility of a malignant tumor. For the excision, we used a transoral approach. The soft palate was split in the midline to gain sufficient exposure for radical extirpation. The literature reports that surgical extirpation of a nasopharyngeal lipoma is usually curative.[1] Postoperative radiotherapy is of no use because lipomas seldom relapse, and reexcision is almost always sufficient in such a case. In case of a liposarcoma, surgical excision is also the therapy of choice. Well-differentiated liposarcomas do not metastasize and seldom relapse, which usually makes postoperative radiotherapy unnecessary.

In summary, in case of a submucosal tumor of the nasopharynx, a lipoma should be included in the differential diagnosis. In such a case, CT and/or MRI scans are invaluable diagnostics.

REFERENCES

1. Kalan A, Ahmed-Shuaib A, Tariq M. Lipoma in fossa of Rösenmuller. *J Laryngol Otol.* 2000;114:265-266.
2. Chaudhry S, Sirpal YM. Lipoma: a rare tumour of nasopharynx. *Indian J Cancer.* 1997;34:177-178.
3. Hong KA, Seo SY, Lee DG. Chondrolipoma of the nasopharynx. *J Laryngol Otol.* 1998;112:75-76.
4. Oddie JW, Applebaum EL. Lipoma of the nasopharynx. *Arch Otolaryngol.* 1982; 108:57.
5. Weiss SW, Goldblum JR. Benign lipomatous tumors and liposarcoma. In: Weiss SW, Goldblum JR, eds. *Enzinger and Weiss's Soft Tissue Tumors.* 4th ed. St Louis, Mo: Mosby–Year Book Inc; 2001:571-693.