Imaging findings of the antineutrophil cytoplasmic antibody-associated vasculitis in the head and neck region: a systematic review of magnetic resonance imaging and computed tomography

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Learning objectives

To recognize the concept of antineutrophil cytoplasmic antibody-associated vasculitis (AAV)

To review imaging findings of AAV of the head and neck (H&N) region

To learn diagnostic clues of AAV of the H&N region

Background

AAV is defined as inflammation in small blood vessels that leads to damage to the tissue or organs supplied by those vessels and is associated with the presence of antineutrophil cytoplasmic antibodies (ANCA). About 75% of all vasculitis cases are ANCA positive with patients showing antibodies to MPO (myeloperoxidase) or PR3 (proteinase 3). Though the incidence of AAV varies little geographically, types of positive ANCA is different. MPO-AAV is observed more frequently in Japan and PR3-AAV in Europe and the USA. Therefore, some controversy remains in current classification criteria and disease definitions concerning the association of AAV with Wegener's granulomatosis, microscopic polyangiitis, and Churg-Strauss syndrome as well as polyarteritis nodosa. Diagnosis is based on clinical manifestations, ANCA testing, and histology.

AAV may affect various organs; common sites in the H&N region are the orbit, paranasal sinus, cavernous sinus, middle ear, and throat. Patients with AAV typically present with fever, fatigue, headache, hearing loss, double vision, blindness, and olfactory disturbance.

AAV also causes various intracranial lesions. In particular, hypertrophic pachymeningitis (HP) shows characteristic imaging findings, and its relevance in lesions of the head and neck is attracting a great deal of attention. HP is a rare disease characterized by inflammation, fibrosis, and thickening of the dura mater, mainly around the brain and occasionally around the spinal cord.

If it is possible to diagnose AAV, treatment with steroids can improve symptoms. Early diagnosis and treatment can significantly improve the quality of life of patients.

Radiologists should be familiar with the manifold imaging appearances of AAV.
Findings and procedure details

We describe the imaging findings of AAV in the H&N region, including those of otitis media, sinusitis, cavernous sinus syndrome, and orbital apex syndrome, and discuss the findings of AAV with regard to inflammation progression, such as in hypertrophic pachymeningitis.

## AAV-related head and neck findings

### Sinusitis

Sinusitis may be accompanied by nasal obstruction, olfactory disturbance, perforation of the nasal septum, and osteoclasis. Onset often accompanies mechanical blockage of natural orifice and may be caused by mass formation or inflammatory adhesion, which exhibits hypointense signals on T$_2$-weighted images. If the disease progresses, inflammation is known to extend to the cavernous sinus and eye socket. MR imaging shows a soft mass/pseudotumor with contrast-enhancing effects on contrasted T$_1$-weighted images in the ethmoidal sinus and anterior cranial fossa from above the upper nasal cavity. This often extends as well to the cavernous sinus and eye socket, and mass formation accompanied by perforation of the nasal septum or osteoclasis may be observed.

### Otitis media

Otitis media first impacts the auditory ossicles causing conductive hearing loss, which gradually develops into mixed hearing loss. Cases in the elderly may be caused by decreased function of the Eustachian tube or obstruction of the Eustachian tube due to a nasopharyngeal tumor or to postinflammation or postoperative adhesion. No bone erosion is observed, including in the auditory ossicles or mastoid wall. Otitis media has been reported in 25% cases of hypertrophic pachymeningitis. T$_2$-weighted images of MR imaging demonstrate hyperintense signals that indicate liquid retention in mastoid air cells. Cranial CT demonstrates this area with low density. Hypoplasia of the mastoid air cells may also be observed.

### Orbital apex syndrome

Orbital apex syndrome is a very rare complication of fractures of the facial skeleton as well as a variety of inflammatory, infectious, neoplastic, iatrogenic/traumatic, and vascular conditions, and it is characterized by blindness, fixed dilated pupils, proptosis, ptosis of the eye, and ophthalmoplegia. A third of pediatric cases are bilateral, whereas most adult cases are unilateral. Patients may ultimately exhibit intraorbital fibrosis following a course of repeated improvement and recurrence. Two-thirds of orbital infections are said to be influenced by sinusitis. This is thought to be due to the extreme thinness of the ethmoid plate that divides the paranasal sinus and eye socket and to the valveless vein-mediated communication between the paranasal sinus and eye socket. Many cases may
develop abscess along the medial orbital wall and are not accompanied by osteoclasis. T₁-weighted images show hypointense signals in the eye socket, and fat-suppressed T₂-weighted images show a pseudotumor with signals that are slightly more intense than those of the muscular tissue. Contrasted T₁-weighted images show soft tissue with contrast-enhancing effects in the orbit.

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**Cavernous sinus syndrome** results from a variety of disease processes and thus produces various signs and symptoms. The strong fascia supporting the cranial side of the epipharynx mucous membrane may impede inflammation progression. However, on the lateral side, the foramen of Morgagni, through which the cartilaginous part of the auditory tube and levator veli palatini muscle communicate, often forms a path for the early advance of maxillary cancer or inflammation to areas such as the parapharyngeal space. When inflammation advances into lymph nodes, the retropharyngeal space or parapharyngeal space may also exhibit inflammatory edema.

Imaging findings show soft tissue/pseudotumor with somewhat hyperintense signals on T₂-weighted images from the cranial base to the epipharynx, the parapharyngeal space the carotid space, and the masticator space. Lesions may also be noted in the Eustachian orifice.

**Hypertrophic pachymeningitis (HP)** is caused by Sjögren's syndrome, rheumatoid arthritis, IgG4-related disease, sarcoidosis, tuberculosis, syphilis, fungal or other types of infections, and malignant tumors in addition to AAV. The chief complaint is headache, and findings of follow-up MR imaging correlate with clinical symptoms in 80% of cases.
MR imaging findings include hypointense signals on T₂ diffusion-weighted images that suggest fibrosis, thickening of the dura mater accompanied by contrast-enhancing effects on contrasted T₁-weighted images, and thickening of the dura mater described as isointense with brain in diffusion-weighted images, which suggests cytotoxic edema.

2. Findings of AAV related to inflammation progression in hypertrophic pachymeningitis (HP)

We reviewed CT and MR imaging findings of 7 cases of head and neck lesions of AAV (5 cases of otitis media, one of sinusitis, three of orbital apex syndrome, three of cavernous sinus syndrome, and five of epipharyngitis) with HP.

A. Pachymeningitis: lesions of the anterior cranial base and H&N region: nasal cavity, paranasal sinus

Dural lesions of the anterior cranial base may continue through the nasal cavity/paranasal sinus via the cribriform plate. Paranasal sinus lesions are likely related to the olfactory nerve and therefore cause olfactory disturbance.

B. Pachymeningitis: lesions of the middle cranial fossa and H&N region: orbit

The dura mater in the middle cranial fossa is present on the outside of the cavernous sinus. Dural lesions in the cranium continue from the cavernous sinus and pharynx to the eye socket. Because the optic, oculomotor, and trochlear nerves run through the eye socket, progression of inflammation into the eye socket may result in a more diverse range of neurological disorders, including visual disturbance, polyopia, and ocular motility disorder. The neurological disorders exhibited may be caused by progression of inflammation to the nerve itself, the exertion of direct pressure on the nerve by thickening of the dura mater, or neuritis as a result of circulatory failure.

C. Pachymeningitis: lesions of the middle cranial fossa and H&N region: epipharynx

Findings of pachymeningitis show connection of dural lesions in the middle cranial fossa to epipharyngeal lesions over the fascia through the foramen lacerum and oval foramen and from the parapharyngeal space through to the masticator space. The lesion may reach the epipharynx and Eustachian orifice, closing off the Eustachian orifice and resulting in secretory otitis media, findings consistent with the difficulty in determining characteristic findings of vasculitis via middle ear biopsy.

D. Pachymeningitis: lesions of the posterior cranial fossa and H&N region: epipharynx
Findings show connection of dural lesions of the posterior cranial fossa to epiphyaryngeal lesions through the jugular foramen from the parapharyngeal space to the carotid space. The mechanism described above may also cause secretory otitis media.

**E. Pachymeningitis: lesions of the posterior cranial fossa and H&N region: internal auditory meatus**

Because the posterior cranial fossa is adjacent to the internal auditory meatus, inflammation may progress continuously through the internal auditory meatus. Physical blockage or progression of inflammation into the auditory nerve itself could also cause auditory perceptual disorder.

Thus, we observed continuity between dural lesions and head and neck lesions and findings that suggested the direct progression of inflammation. However, we did not observe the unidirectional characteristics of the route of inflammation progression, and its bidirectional characteristics made it difficult to determine which lesions were affected first.

The very diverse appearances of vasculitis lesions make their one-dimensional description difficult.

The idea of progression of continuous lesions is consistent with the consideration of AAV as an inflammatory disease, and the idea of incidental lesion influence and progression is plausible if AAV is considered an autoimmune disease. AAV may also have the characteristics of both autoimmune and inflammatory disease. Accordingly, if olfactory disturbance, visual disturbance, and auditory disorder are observed, it is important to consider the possibility of a related head and neck lesion and to make a diagnosis after conducting appropriate imaging.

**Images for this section:**
Fig. 1: A 79-year-old woman developed hearing loss in her right ear and headache in the occipital region. Laboratory findings were positive for myeloperoxidase (MPO)-antineutrophil cytoplasmic antibodies (ANCA), and she was diagnosed with antineutrophil cytoplasmic antibody-associated vasculitis (AAV) and refractory otitis media. Contrast T1-weighted imaging demonstrated thickening of the dura mater with contrast-enhancing effect in the right middle cranial fossa and posterior fossa (#). A diffusion-weighted image shows thickening of the dura in the right frontal region (#).
Fig. 2: A 71-year-old woman developed right ophthalmodynia, nose fullness, and headache. Laboratory findings were positive for myeloperoxidase (MPO)-antineutrophil cytoplasmic antibodies (ANCA), and she was diagnosed with antineutrophil cytoplasmic antibody-associated vasculitis (AAV) and interstitial pneumonia. A contrast T1-weighted image demonstrates thickening of the dura mater with contrast-enhancing effect on the right posterior fossa and left frontal region(#). Furthermore, contrast-enhancing effect is apparent in the orbital subperiosteal space at the back of the eye(#). Slightly contrast-enhancing effect along the right optic nerve sheath suggests inflammatory extension to the optic nerve(#). A diffusion-weighted image demonstrates thickening of the dura mater in the left frontal region(>).
Fig. 3: A 69-year-old woman developed hearing loss in both ears, visual disorder, and headache. Laboratory findings were positive for myeloperoxidase (MPO)-antineutrophil cytoplasmic antibodies (ANCA), and she was diagnosed with antineutrophil cytoplasmic antibody-associated vasculitis (AAV). A contrast T1-weighted image demonstrates thickening of the dura mater with contrast-enhancing effects on both sides of the middle cranial fossa and from both sides of the nasopharynx(#) to the Eustachian orifice.(#). T2-weighted images demonstrate high signal of the mastoid air cells on both sides(#). After treatment, thickening of the dura mater improved, and abnormal intensity of the left mastoid air cells disappeared(##).
Fig. 4: A 72-year-old man developed hearing loss in both ears and headache. Laboratory findings were negative for myeloperoxidase (MPO)-antineutrophil cytoplasmic antibodies (ANCA), but antineutrophil cytoplasmic antibody-associated vasculitis (AAV) was strongly suspected. A contrast T1-weighted image shows thickening of the dura mater with contrast-enhancing effects in the left middle cranial fossa(#) and from both sides of the nasopharynx to the Eustachian orifice(#). The contrast-enhancing effect in part of the left mastoid air cells is also seen(#). T2-weighted images revealed an area of high signal on both sides of the mastoid air cells(#). After treatment, the contrast-enhancing effect disappeared, and the abnormal intensity of the mastoid air cells improved(##).
Fig. 5: A 77-year-old man developed double vision and visual disorder in the right eye and headache. Laboratory findings were positive for myeloperoxidase (MPO)-antineutrophil cytoplasmic antibodies (ANCA), and he was diagnosed with antineutrophil cytoplasmic antibody-associated vasculitis (AAV). A contrast T1-weighted image shows
soft tissue with contrast-enhancing effect in the upper part of nasal cavity and ethmoid sinus and from the orbital apex to the right cavernous sinus(>). The unclear boundary of the intranasal soft tissue and dura of the frontal base suggests the continuity of inflammation(#). Soft tissue with contrast-enhancing effect that spans from the base of the skull to the nasopharynx and both sides of the parapharyngeal space is seen(#). A diffusion-weighted image shows thickening of the dura mater of the frontal region(>). T2-weighted images show a high signal on the left mastoid air cells(#). After treatment, the dura mater remained thickened(x#), but lesions of the head and neck improved(##).

**Fig. 6:** An 88-year-old woman developed hearing loss, double vision, visual loss, and pain in both legs. Laboratory findings were positive for myeloperoxidase (MPO)-antineutrophil cytoplasmic antibodies (ANCA), and she was diagnosed with antineutrophil cytoplasmic antibody-associated vasculitis (AAV). A contrast T1-weighted image shows thickening of the dura mater with contrast-enhancing effect on the front of the posterior fossa that involves the inner ear canal and contrast-enhancing effect from the left nasopharynx and parapharyngeal space to the carotid space through to the left jugular foramen(#). A coronal contrast T1-weighted image also shows contrast-enhancing effect on the left
cavernous sinus(##). After treatment, lesions improved overall(##), but findings for the cavernous sinus did not change(x#).

Fig. 7: A 71-year-old woman developed hearing loss, headache, and pain in both legs. Laboratory findings were positive for myeloperoxidase (MPO)-antineutrophil cytoplasmic
antibodies (ANCA), and she was diagnosed with antineutrophil cytoplasmic antibody-associated vasculitis (AAV). A contrast T1-weighted image showed thickening of the dura mater with contrast-enhancing effect in the right middle cranial fossa (#) and contrast-enhancing effect from the right nasopharynx to the parapharyngeal space and masticator space(#). Lesions of the nasopharynx and middle cranial fossa continue through the lacerated foramen, oval foramen, and spinous foramen. Even on computed tomography (CT), the spinous foramen is dilated(#). In T2-weighted images, high signal of the right mastoid air cells suggests mastoiditis(>). After treatment, thickening of the dura mater remained(x#), the contrast-enhancing effect disappeared, and the mastoiditis improved(# #).

**AAV in the head and neck region**

Fig. 8
Conclusion

Magnetic resonance imaging and computed tomography are powerful and significant instruments for diagnosing antineutrophil cytoplasmic antibody-associated vasculitis in the head and neck region. Understanding anatomic relationships and structures will allow correct diagnosis of AAV, and knowledge of its imaging features in this region will aid appropriate patient management.

Personal information

References

