Diagnostic criteria for Otitis media with ANCA associated vasculitis (OMAAV)

(A) At least one of the following clinical courses:
1. Intractable otitis media with effusion or granulation, which is resistant to antibiotics and insertion of tympanostomy tube.
2. Progressive deterioration of bone conduction hearing levels.
(B) At least one of the following features:
1. Already diagnosed as AAV (GPA, MPA, EGPA) based on the involvement of other organs.
2. Positivity for serum MPO- or PR3-ANCA.
3. Histopathology consistent with AAV, i.e., necrotizing granuloma with infiltration of giant cells, necrotizing vasculitis predominantly affecting small vessels with or without granulomatous extravascular inflammation.
4. At least one of the following accompanying signs/symptoms of AAV-related involvement:
① upper airway involvement other than ear, scleritis, lung, and kidney involvement, ② facial palsy, ③ hypertrophic pachymeningitis, ④ multiple single neuritis ⑤ improvement in symptoms/signs with administration of 0.5 to 1.0 mg/kg prednisolone (although recurrence occurs on discontinuation of treatment).
(C) Differential diagnosis (the followings are excluded):
(1) cholesteatoma, (2) cholesterol granuloma, (3) eosinophilic otitis media, (4) tuberculosis (5) malignant otitis externa, skull-base osteomyelitis, (6) neoplasms (malignancy, inflammatory myofibroblastic tumor, etc.), (7) otitis media or inner ear inflammation caused by autoimmune diseases and vasculitis other than AAV.

A diagnosis of OMAAV can be made when the above three criteria (A, B, C) are fulfilled.

ANCA: anti-neutrophil cytoplasmic antibody
AAV: ANCA-associated vasculitis
GPA: granulomatousis with polyangiitis

MPA: microscopic polyangiitis
EGPA: eosinophilic granulomatousis with polyangiitis
PR3: proteinase 3
MPO: myeloperoxidase