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ANCA-Associated Vasculitides (AAV) Presenting as Isolated Right Infra-auricular Abscess

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ABSTRACT

Anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis (AAV) is a rare autoimmune disease in children, typically affecting small- to medium-sized vessels and presenting with systemic manifestations. Salivary gland involvement is uncommon and may mimic infection or neoplasm, making early diagnosis challenging. We present the case of a 10-year-old boy with recurrent right parotid gland swelling and necrotizing granulomatous inflammation despite surgical drainage and antibiotic therapy. Laboratory studies revealed PR3-ANCA positivity, supporting the diagnosis of AAV. The patient responded well to oral prednisolone and co-trimoxazole, achieving clinical and sonographic resolution. This case highlights the importance of including AAV in the differential diagnosis of persistent or recurrent parotid gland lesions in children, as early recognition and initiation of immunosuppressive therapy can improve clinical outcomes.

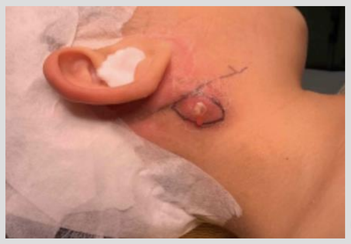
INTRODUCTION

Anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitides (AAV) are a group of rare autoimmune diseases characterized by necrotizing inflammation of small- to medium-sized blood vessels. AAV comprises three main clinical entities: granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA)[1]. The presence of ANCA—particularly antibodies targeting proteinase 3 (PR3) and myeloperoxidase (MPO)—serves as a key diagnostic and pathogenic hallmark of these conditions[2].

Epidemiologically, AAV primarily affects older adults, with peak incidence occurring between 65 and 74 years of age. The overall annual incidence is approximately 10–20 per million individuals in European populations [3]. In contrast, pediatric cases are exceedingly rare and less well characterized. Notably, geographic differences exist: GPA and PR3-ANCA are more prevalent in Western populations, while MPA and MPO-ANCA dominate in East Asia [4]. Despite the low frequency of pediatric presentations, they often pose significant diagnostic challenges, particularly when classical organ involvement is absent.

Salivary gland involvement in AAV is uncommon and rarely reported as the initial or sole manifestation. We present a pediatric case of ANCA-associated vasculitis manifesting as recurrent parotid swelling and necrotizing granulomatous inflammation. The clinical course highlights the need for high clinical suspicion when evaluating persistent head and neck lesions unresponsive to conventional treatment.

FIGURE



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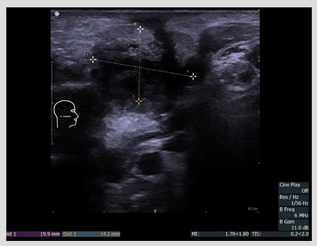

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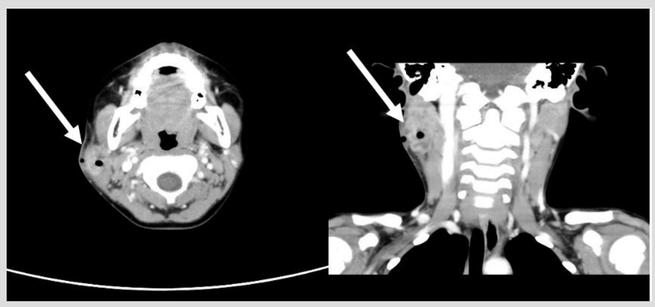

(3)

Figure (1). Preoperative image showing a recurrent subcutaneous lesion with central pustule and keloid changes over the right infra-auricular scar.
Figure (2). Sonographic examination of the right parotid gland revealed an ill-defined, irregular, hypoechoic lesion, measuring approximately 1.99 × 1.41 cm. The lesion demonstrated heterogeneous internal echotexture, suggesting an abscess.
Figure (3). The head and neck computed tomography (CT) reported heterogeneous enlargement of the right parotid gland with two air bubbles and internal necrosis (arrow)

CASE

A 10-year-old Asian boy had no congenital disorder. He had right infra- auricular tumor and received right parotid operation at other medical center in 2021/03. The pathology report revealed granulomatous inflammation and salivary gland tissue with abscess formation. One lobulated/cystic lesion along previous scar was noted by his family in 2022/07, so we arranged the head and neck computed tomography (CT) with contrast revealed heterogeneous enlargement of right parotid gland with two air bubbles and internal necrosis. Sonography at head and neck area showed a right parotid ill-defined mass with abscess formation and intra-glandular lymph nodes and skin keloid formation. We arranged right neck abscess incision and drainage and the pathology still revealed necrotizing granulomatous inflammation. We also sent the abscess for bacterial and tuberculosis culture and only some Coagulase-negative Staphylococcus was found but no Mycobacterium was isolated. The patient was referred to the pediatrician for further evaluation and treatment. The blood test of the patient showed c-ANCA positive and Anti-proteinase 3 (PR3) Ab=3.7 IU/ml , but Anti-myeloperoxidase (MPO) Ab was negative. Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides was diagnosed and the patient was treated with oral prednisolone and the pediatrician treated secondary bacterial infection with trimethoprim-sulfamethoxazole (Co-trimoxazole).

DISCUSSION

AAV typically presents with systemic features involving the respiratory tract, kidneys, or skin. However, extrapulmonary manifestations such as salivary gland involvement have been reported sporadically, mostly in adult patients [5-9]. In our case, the patient developed recurrent parotid abscesses without initial systemic symptoms, delaying consideration of a vasculitic process. Histopathology repeatedly showed necrotizing granulomatous inflammation, and the serologic finding of PR3-ANCA positivity eventually supported the diagnosis of AAV.

Salivary gland involvement as a presenting feature in pediatric AAV is exceedingly rare. Several adult case reports have described similar manifestations, including unilateral or bilateral parotid gland enlargement [6-9]. These atypical presentations emphasize the importance of broadening differential diagnoses in persistent or recurrent head and neck infections, particularly when microbiologic cultures are negative or reveal only skin commensals.

The underlying pathogenesis of AAV involves a complex interplay between genetic, immunologic, and environmental factors. PR3- and MPO-ANCA are believed to activate primed neutrophils, leading to endothelial injury and necrotizing inflammation [2,5]. Genetic studies have demonstrated stronger associations between ANCA specificity and HLA types than between ANCA subtype and clinical diagnosis, suggesting a mechanistic link rooted in immunogenetics [10]. Infectious triggers, including bacteria and viruses, have also been implicated in initiating or exacerbating disease activity [5]. In our patient, no pathogenic organisms were identified, but the persistent nature of the inflammation prompted further autoimmune workup.

Management of AAV in pediatric patients remains largely extrapolated from adult treatment guidelines. The use of systemic corticosteroids remains the cornerstone of initial therapy. Our patient responded to a combination of oral prednisolone and trimethoprim-sulfamethoxazole (Co-trimoxazole), which may have dual benefit in GPA-like phenotypes by reducing bacterial colonization and disease relapse [11-13].

CONCLUSION

This case illustrates a rare and atypical presentation of pediatric ANCA-associated vasculitis, with recurrent parotid gland inflammation as the dominant clinical feature. While salivary gland involvement is not a classical manifestation of AAV, it may occur and can mimic infectious or neoplastic conditions. Persistent or refractory head and neck lesions in children warrant consideration of autoimmune etiologies. Early recognition and appropriate serologic and histopathologic evaluation are crucial to avoid unnecessary surgical interventions and to initiate timely treatment. Ongoing research is needed to better understand the pediatric spectrum of AAV, including its pathogenesis, epidemiology, and optimal therapeutic strategies.

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