GRANULOMATOSIS WITH POLYANGIITIS IN CHILDREN: RARE DISEASE

SEFAKO MAKGATHO HEALTH SCIENCES UNIVERSITY

JN Mpholefole, N Diale, A kganyago, RS Masela

Department of Otorhinolaryngology (ENT), Sefako Makgatho Health Sciences University

INTRODUCTION

Granulomatosis with polyangiitis (GPA) is one of the antineutrophil cytoplasmic antibody (ANCA)-associated vasculitic disorders, previously called Wegener's granulomatosis. It is a rare autoimmune disease of unknown cause characterized by necrotizing granulomatous inflammation and vasculitis of small and medium sized vessels. (1) It is believed to be triggered by environmental events on a background of genetic susceptibility. (5)(6) This disease is rare in childhood, but awareness is crucial for the prognosis of the child. (4)

CASE PRESENTATION

History: 9 years old male presented in June 2022 with 15 months history of generalised skin lesions which started from the lower limb and spread to the upper body, constitutional symptoms, upper respiratory symptoms. Three months history of hoarseness of voice, dyspnoea, rhinorrhoea, left otalgia and otorrhea. Previously treated for PTB in 2021 at a local clinic.

Examination: Revealed generalised granulomatous skin lesions, hoarseness, inspiratory stridor with tracheal tug, left otorrhea with inflamed tympanic membrane, mucopurulent rhinorrhoea, inflamed nasal mucosa. Gingivitis on oral examination. Flexible fibre optic scope showed narrowed airway with fused oedematous epiglottis, aryepiglottic and arytenoid.

Investigations: Direct laryngoscopy showed oedematous, fragile, and scarred laryngeal mucosa with narrowed supraglottic area and subglottic stenosis. Tracheostomy was performed.

Blood results: anti-proteinase 3 antibody is positive, elevated ESR, normal kidney functions

Imaging: CT showed sinusitis, supraglottic narrowing and no lungs lesions

Audiogram: normal hearing

Histology: no features of a well granuloma or vasculitis

Treatment: patient was referred to paediatric rheumatology department at Charlotte Maxeke Academic Hospital for treatment. He was put on steroids and azathioprine.

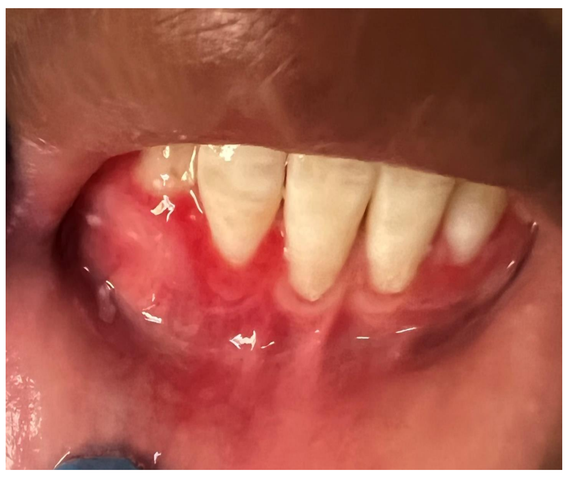

Fig. 1. Gingivitis

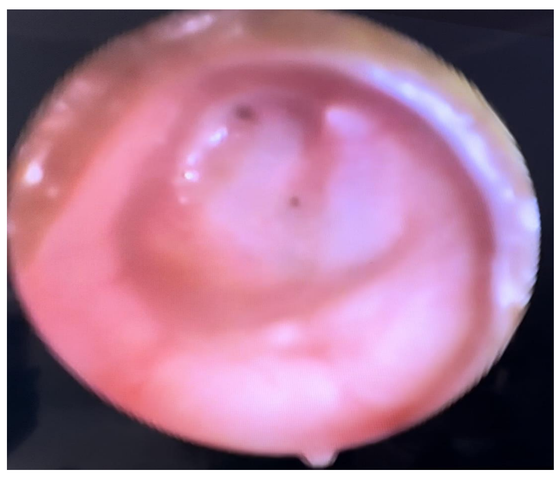

Fig. 2. Left inflamed tympanic membrane

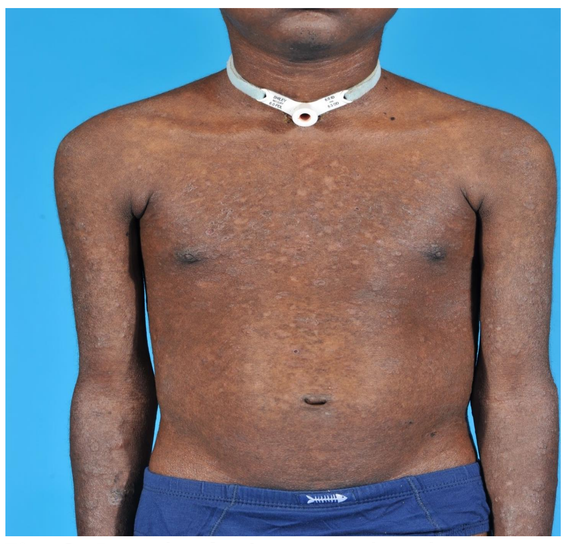

Fig. 3. Papular lesions on the chest and arms

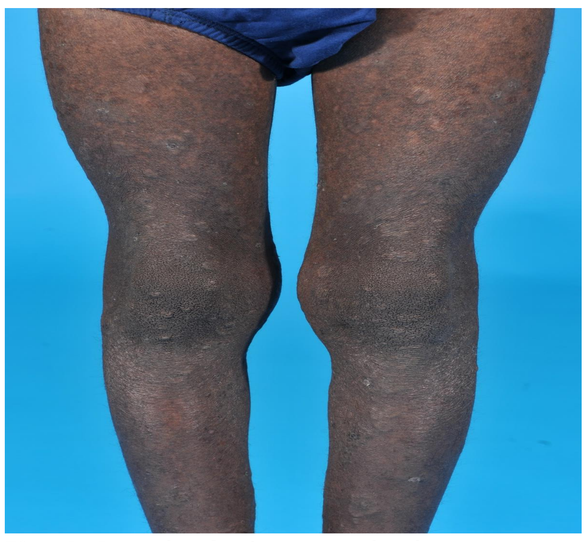

Fig. 4. Papular lesions on the lower limb

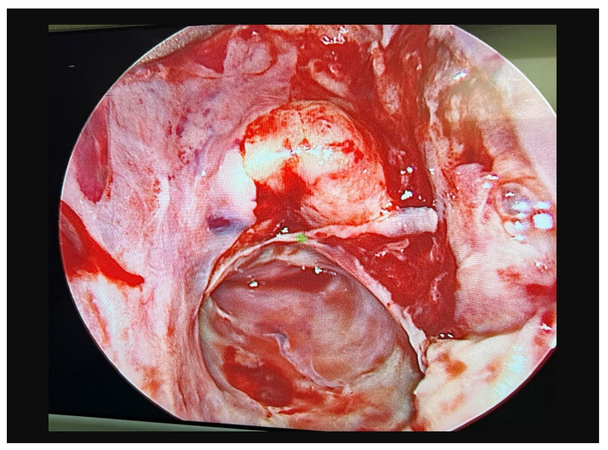

Fig. 5. Oedematous epiglottis, inflamed Mucosa

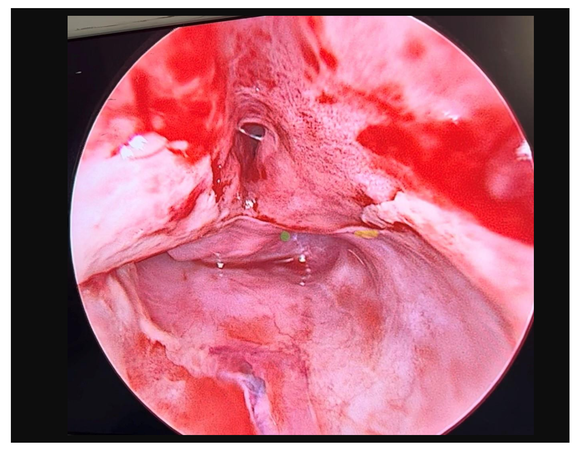

Fig. 6. Supraglottic area

DISCUSSION

- It is a pauci-immune small-vessel vasculitis which in turn decreases blood flow to organs and systems affected. In Europe, the prevalence of GPA is 5/100000 population. GPA can occur in all racial groups but predominantly affects Caucasians. (2)
- There is a slight male predilection and onset is usually at middle age approximately 50 years however, ages of affectation can vary widely and had been seen in children as young as 3 months of age. There are scanty reported cases of GPA in Africans and have been reported to be rare in African Americans. (3) Terrier et al found the black population to be 2.9%. Despite the rarity, disease severity was more in blacks and the age of affectation was lower. (5)
- The classic triad of organ involvement includes lung (involved in 95% of cases), upper respiratory tract/sinuses (involved in 75%-90% of cases), and kidneys (involved in 80% of cases) (1). Patients can present with non-specific symptoms cough, haemoptysis, weight loss, fever, athralgia and rash. Histology shows necrotizing granulomas with an associated vasculitis. 90% of the cases demonstrates positive cANCA (PR3), which also correlate with disease activity. A high index of suspicion is needed to diagnose it because of its rarity and nonspecific symptoms at presentation. (1)
- Diagnosis is achieved through clinical assessment, serological tests for anti-neutrophil cytoplasmic antibodies (ANCA) and histological analysis. The 10-year survival rate is estimated to be 40% when the kidneys are involved and 60–70% when there is no kidney involvement. (2)
- Management includes immunosuppressants (cyclophosphamide, methotrexate and steroids) (1), cyclophosphamide is switched to azathioprine in young patients in maintenance phase (2). Without treatment, granulomatosis with polyangiitis is rapidly progressive with 10% two-year survival. Appropriate medical therapy has increased long term survival. (1)

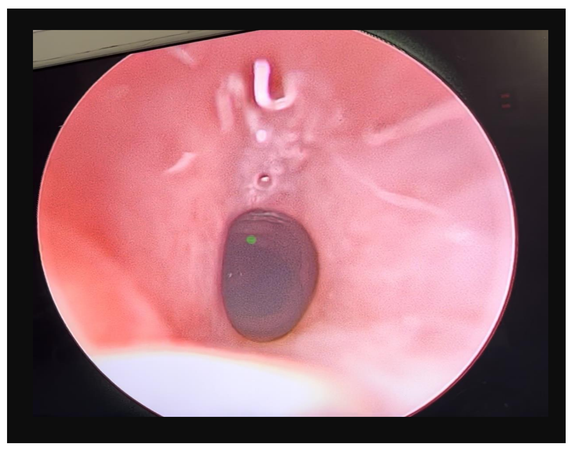

Fig. 7. Subglottic stenosis

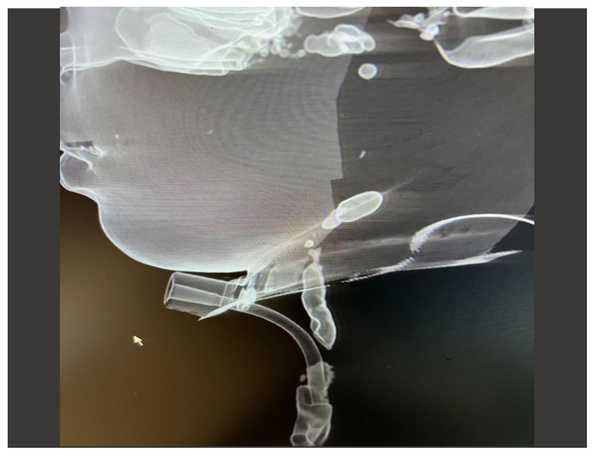

Fig. 8. CT scan showing narrowing of supraglottic area

CONCLUSION

GPA is a rare multisystem autoimmune disease in childhood, without high index of suspicion , diagnosis can be delayed or missed. Treatment strategies and clinical approach are mostly derived from adult GPA studies. Relapses are frequent and patients should be evaluated frequently.

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