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Case report/Kazuistyka

Isolated subglottic stenosis by Wegner's granulomatosis in a pediatric patient – A case report

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ABSTRACT

Wegener's granulomatosis (WG) is a systemic autoimmune lesion of unknown etiology characterized by necrotizing granulomatous inflammation and vasculitis affecting small blood vessels. Although WG is a rare condition in children, it is often confused with more common ailments, so there is delay in confirmation of the diagnosis. We present a 13-year-old boy who came with breathing difficulty. A nasopharyngolaryngoscopy revealed narrowing in the subglottic region. This was managed by planned microlaryngoscopy with cold steel instruments. Excision of granulomatous tissue from subglottis along with application of steroid and mitomycin-C. Biopsy from the subglottic area confirmed the diagnosis of WG. During managing subglottic stenosis (SGS), the etiology of WG should be in mind as it is essential to find out the cause for providing the appropriate treatment. An early diagnosis of WG leads to early initiation of treatment which helps in reduction of morbidity and mortality associated with the disease.

Introduction

Wegener's granulomatosis (WG) is a necrotizing granulomatous vasculitis of autoimmune origin having multisystemic involvement. Heinz Klinger first described the Wegener's granulomatosis in 1931 and subsequently described by Frederich Wegener in 1936 [1]. WG has a predilection for the upper and lower respiratory tract and kidneys. Patients without involvement of kidney in WG are called as limited Wegener's. There is no definite cause for WG although it is not infectious or hereditary. The diversity of clinical presentation of WG contributes to difficulties in diagnosis of this disease. The most common ages of presentation of WG are the sixth and seventh decades but it can seen at any age with equal frequency between genders among adult ages [2]. Clinical form of WG can vary from localized to multisystemic involvement. Clinical manifestations of WG range from nasal or oral involvement, microhematuria, characteristic radiological abnormalities and characteristic histopathological pictures. Laryngeal involvement is rare, but if affected, the subglottis and upper trachea are most commonly involved, with characteristic of circumferential scarring and narrowing of the airway. As subglottis is the narrowest part of laryngotracheal airway in children, subglottic stenosis (SGS) becomes a potentially life threatening

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situation in WG. The narrowing of the airway at the level of cricoids cartilage presents a challenging situation for the treating doctors. WG is an uncommon clinical condition with a prevalence of 3 per 100 000 where SGS occurs in around 16% of cases leading to life threatening airway obstruction [3]. The characteristic laboratory findings in WG are elevated ESR, leucocytosis, anemia, mildly elevated rheumatoid factor and positive antiproteinase-3 anti-neutrophil cytoplasmic antibodies (ANCA). The C-ANCA test is not always positive initially and should be repeated if the WG is suspected. Usually SGS in WG does not respond to the medical treatment [3]. We present a case of WG in a pediatric patient in whom isolated SGS was treated by microlaryngeal excision, dilatation with application of mitomycin-C and topical steroids.

Case report

A 13-year-old boy came to Outpatient Department of Otorhinolaryngology for complaints of breathlessness since 5 days. He had no nasal and ear complaints except occasional dry cough. On clinical examination, there is no regional lymph nodes enlargement. Nasopharyngolaryngoscope demonstrated a narrowing at the level of subglottis (Fig. 1). Computed tomography (CT scan) of the neck confirmed the narrowing at the subglottis with normal lower trachea (Fig. 2). He had very high ESR (110 mm/h) and his C-ANCA was positive in previous report. The diagnosis was suspected as WG. Our patient was planned for microlaryngoscopy and dilation made by cold steel instruments. Tissue from the stenotic segments was sent for histopathological examination. After excision of the stenotic segment, the raw area was injected with corticosteroids (methylpredinsolone at the dose of 80 mg) and local application of mitomycin-C (2 mg mitomycin diluted in 1 ml distilled water). The diagnosis of WG was made by demonstration of vasculitis on tissue biopsy. The biopsy of multiple fragments from subglottis revealed granulations infiltrated with lymphocytes, histiocytes, giant cells and a small vessel showing granulomatous vasculitis (Fig. 3). His X-ray paranasal sinus and X-ray chest revealed no abnormality. His serum glucose, urea and creatinine are within normal limit. His three consecutive sputum tests were negative for acid fast bacilli (AFB). The patient is now treated with combination therapy of prednisolone 1 mg/kg and cyclophosphamide 2 mg/kg. He has now no breathing difficulty.

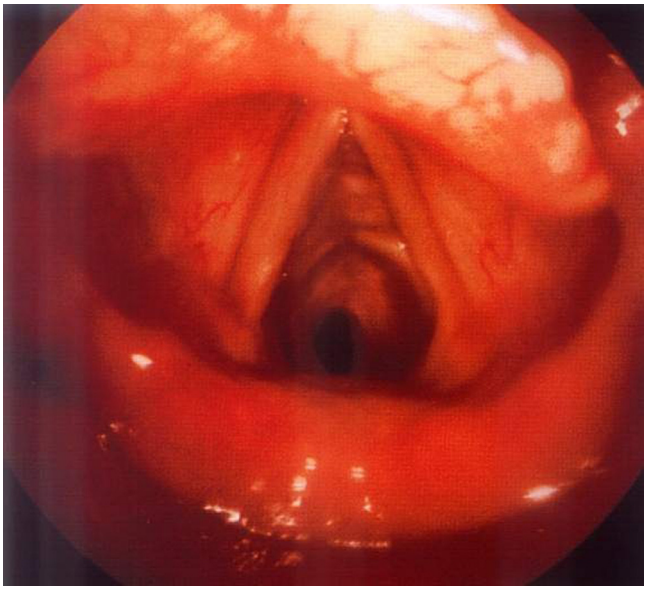

Fig. 1 – Nasopharyngolaryngoscopy showing subglottic stenosis

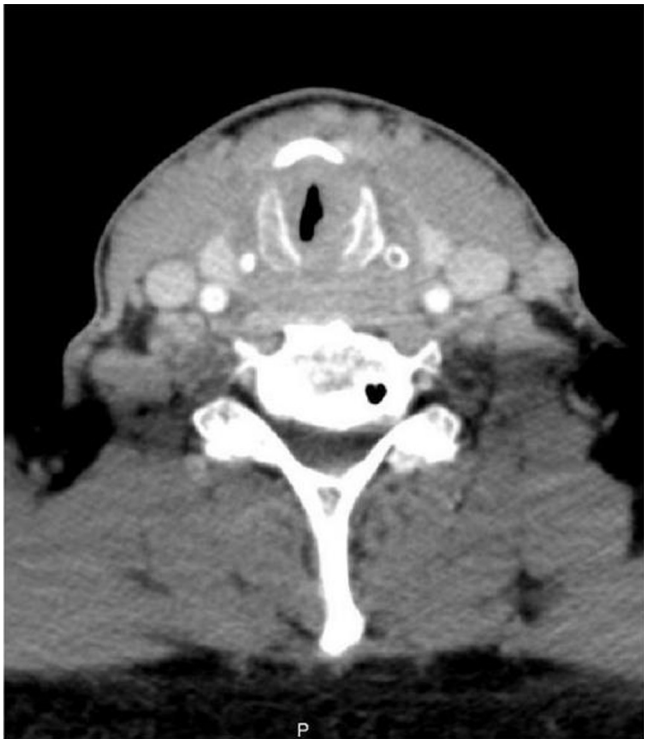

Fig. 2 – CT scan of the neck showing subglottic stenosis

Discussion

WG is a granulomatous multisystemic disorder characterized by granulomatous inflammation of the respiratory tract, necrotizing vasculitis affect small to medium sized vessels and necrotizing glomerulonephritis. WG is also known as granulomatosis with polyangitis (GPA) [4]. There are three variety of WG: type 1, 2 and 3. The type 1 is the limited form of WG where patient presents with symptoms of upper respiratory infection for weeks which is not responding to antibiotics and associated with serosanguinous type of nasal discharge, pain and crust formation in nasal cavities. In Type 2, along with nasal involvement other organs also are involved. In Type 3 WG, there is wide spread involvement of the multiple organs including airway, pulmonary, renal and sometimes associated dermatological lesions [5].

Laryngotracheal manifestations in WG are rare, with subglottic involvement being the most common. These are