TEMPI Syndrome Refractory to Current Standards of Therapy

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Introduction

• TEMPI syndrome is a multisystem disease, first described in 2011 as
  - Telangiectasias
  - Erythrocytosis with elevated erythropoietin
  - Monoclonal gammopathy
  - Perinephric fluid collections
  - Intrapulmonary shunting
  - 16 cases reported to date
• Often responds to therapy targeting the abnormal plasma clone - classified as a plasma cell dyscrasia with paraneoplastic features

Case Description

• In 2011, a 50 year old male presented with erythrocytosis
• It had been ongoing for 13 years, previously assumed 2/2 polycythemia vera, which had been treated with therapeutic phlebotomies leading to iron deficiency anemia
• A few months after presentation, the patient was found to have:
  - Shortness of breath, O₂ saturation of 90-92% with clubbing
  - Intrapulmonary shunting of 12.3%
  - IgG-kappa monoclonal protein of 0.91 g/dL on SPEP
  - Perinephric fluids collection on ultrasound
  - Telangiectasias on the chest
• He was diagnosed with TEMPI syndrome
• The patient failed treatment with bortezomib and lenalidomide
• Autologous stem cell transplant not employed due to comorbidities
• Patient remained stable for four years until August 2018, when:
  - Found to have increased intrapulmonary shunting from 12.3% to 40.5% and increased telangiectasias on chest, back, and lips
  - Started on daratumumab but ultimately failed treatment
• Regrettably, the patient succumbed within the year

Discussion

• In many cases, progression to intrapulmonary shunting and hypoxemia can be avoided with proper treatment, which results in a more indolent course
• Therefore, early recognition of the symptoms of TEMPI syndrome and establishing the diagnosis is key
• In this regard, we believe that telangiectasias can play an important role, as they represent a prominent cutaneous finding that has been proposed to occur early in disease
• The present case represents the first refractory to daratumumab

Implications

• Telangiectasias represent a prominent manifestation of TEMPI syndrome, and their presence, when concurrent with other cardinal features of the disease, should facilitate earlier diagnosis
• This case highlights the need for additional treatments and further understanding of TEMPI syndrome

Fig 1. Telangiectasias on lips and chest

Fig 2. Telangiectasias on the back

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