

neurosensory deafness and growth hormone deficiency.<sup>7,8</sup> The patients lack tonsils and palpable lymphoid tissue, and the immunoglobulin levels and B lymphocyte counts are either reduced or absent.<sup>6</sup> Cellular immunity is normal. There may be neutropenia in 15–25% of the cases.<sup>6</sup>

The diagnosis of XLA is easy to establish in the presence of a male with intense hypogammaglobulinaemia, a reduced or absent B cell count (<2% of CD19+ or CD20+ cells) and normal T lymphocytes, in the context of other similarly affected males in the family – although in one third of all cases there is no positive family history.<sup>1,2</sup> The study of the Btk gene based on molecular biological techniques allows confirmation of the diagnosis if the causal mutation is found. Such techniques also allow us to establish a differential diagnosis, study carriers, provide genetic counselling, and establish a prenatal diagnosis of the disease.<sup>1,2</sup>

Treatment consists of the administration of gammaglobulin via the intravenous route at high doses which must be individualised ( $\geq 400$  mg/kg every 3–4 weeks). In this context, higher doses are needed in the presence of bronchiectasis and enteroviral infections.<sup>6</sup> The early introduction of gammaglobulin via the intravenous route allows patients to lead a normal life, and generally without organ disease. The prognosis is favoured when an early diagnosis is established, with the prompt introduction of therapy.<sup>2,5</sup> At present, thanks to advances in the diagnostic techniques and treatments, the disease is detected early (mean age 3 years), and the affected children survive and reach adulthood. Although XLA can affect adult daily life (with more frequent hospital admissions and sick leave), quality of life is similar to that of the rest of the population, and these patients can become productive individuals for society (with increased educational and income levels).<sup>9</sup> The development of gene therapy involving Btk gene transfer in haematopoietic progenitor cells could represent a definitive solution for management of the disease.<sup>10</sup>

Our patient involved a typical presentation of XLA. In effect, the disease was diagnosed in a three-year-old boy with recurrent infections from the first year of life, intense hypogammaglobulinaemia and the absence of B lymphocytes. Of note is the important positive family history in this case, with three male siblings who had died of infectious diseases at an early age, which proved to be of great diagnostic utility. Indeed, based on the above antecedents, the diagnosis could have been established in our setting much sooner, and even in the prenatal period – with subsequent early treatment and counselling of the mother after the death of her first child.

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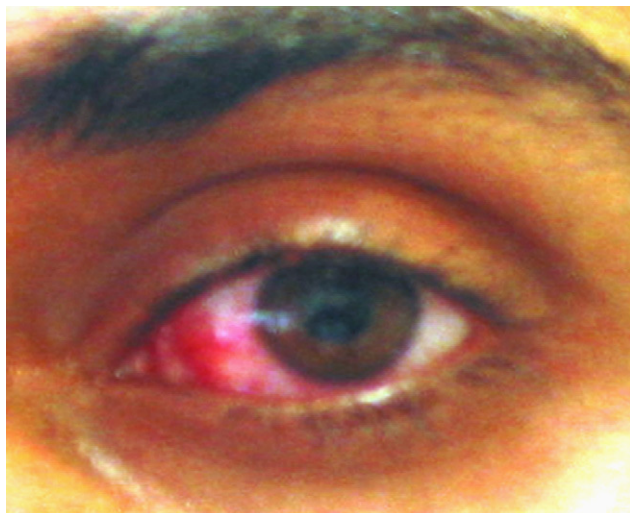
## Rhinoconjunctivitis elicited by skin prick test

*To the Editor:*

Antunes J et al. wrote a nice review about skin prick test (SPT).<sup>1</sup> We congratulate them and we also report a case of adverse reaction to SPT elicited by an iatrogenic procedure.

Skin prick test is an essential diagnostic tool in allergy practice. The simplicity, rapidity of performance, improvement of patient adherence, high sensitivity and low cost make it preferable to in vitro testing for determining the presence of specific IgE antibodies.

Systemic reactions with SPT for inhalant extracts are rare and have decreased dramatically to an overall risk below 0.02% for anaphylactic reactions.<sup>1,2</sup> Studies have identified some risk factors for systemic reactions: SPT with fresh food,



**Figure 1** Conjunctivitis with oedema and hyperaemia of conjunctiva.

latex, and non-standardised extract; SPT in duplicate; SPT in young children, pregnant women, and patients with extreme eczema.<sup>1,2</sup>

Another main cause of adverse reactions to SPT is iatrogenic procedures. SPT should always follow the pre-established recommendations, and safety procedures should be adopted as a precaution in the event of adverse reactions during or after the test. The correct technique, the extract quality and its concentrations are crucial to obtain a reliable and safe SPT.

We report a 27-year-old patient with mild persistent allergic rhinoconjunctivitis and asthma; the diseases were well controlled with low doses of nasal and inhaled corticosteroids. The patient was being evaluated for immunotherapy, during a clinical trial, when he underwent a SPT with a standardised *Dermatophagoides pteronyssinus* (Der p) extract. After 10 min, our patient started to complain of ocular and nasal itching and developed a rhinoconjunctivitis crisis (Figure 1). Mean wheal diameter, calculated as the sum of the largest diameter (25 mm) and its largest orthogonal diameter

(15 mm) divided by 2, was 20 mm. He was promptly treated with intravenous anti-histamine and presented clinical improvement.

Reviewing the extract preparation, we realized that our extract was twenty times more concentrated than a SPT extract should be. By mistake, its dilution had not been made appropriately, and it was not double checked, a procedure that could have prevented the systemic adverse reaction observed.

A month later, the patient was resubmitted to the SPT and performed a specific bronchial challenge, both tests using an appropriate Der p extract. At this time, the SPT elicited a mean wheal diameter of 6 mm. The specific bronchial challenge was positive, showing an early and a late reaction.

This case illustrates that an allergen can be absorbed through the skin, depending on its concentration, and elicits distant reactions in primed tissues. We have also shown that an allergen can be absorbed by the airway mucosa during a specific bronchial challenge and cause delayed urticaria.<sup>3</sup> Skin prick test, a hallmark of allergy practice, is a safe procedure, but pre-established recommendations have to be followed.

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