Pulmonary Renal syndrome in Henoch-Schönlein Purpura: a rare but serious complication

1. Katherine Clarke, Department of Paediatric Rheumatology, Great Ormond Street Hospital, London, United Kingdom
2. Elena Kurteva, Department of Paediatric Gastroenterology, Great Ormond Street Hospital
3. Neil Sebire, Department of Paediatric Histopathology, Great Ormond Street Hospital
1. Muthana Al Obaidi, Department of Paediatric Rheumatology, Great Ormond Street Hospital

Clinical Case

A five year old male presented with fever, abdominal pain and rectal bleeding. He had a three week history of rash and arthralgia which had been diagnosed as Henoch-Schönlein Purpura(HSP).He had a widespread purpuric rash and swollen extremities with necrotic lesions noted on his hands and his feet (Figure 1).

Figure 1: Widespread purpuric lesions with necrotic areas.

Investigations showed:

- Anaemia
- Proteinuria and haematuria
- Renal biopsy: evidence of a diffuse glomerulonephritis with strong deposition of IgA (Figure 2).
- Negative ANA, ANCA, DsDNA
- Normal complement

Figure 2: Photomicrograph of the renal biopsy demonstrating diffuse mesangial proliferative and endocapillary proliferative glomerulonephritis

Clinical Course

He was pulsed with IV methylprednisolone. He continued to deteriorate with falling haemoglobin, progressive renal impairment and respiratory deterioration. Chest X Ray showed bilateral focal abnormalities (Figure 3). During intubation fresh blood was noted in his trachea. He eventually required high frequency oscillation ventilation.

Figure 3: Chest X Ray following acute deterioration showing widespread air space shadowing

He received further intravenous steroids followed by cyclophosphamide and plasmapheresis. He made slow improvement and was extubated. He was commenced on oral prednisolone and enalapril. He continued to make a steady recovery and within two weeks he was mobile, with no oxygen requirement, no cutaneous scars and no gastrointestinal symptoms.

Discussion

HSP is the most common type of IgA vasculitis in childhood. It usually runs a benign and self limiting course(1). Pulmonary and glomerular involvement is a rare but life threatening complication requiring early recognition and aggressive management.

References

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