CASE THREE

Short case number: 3_28_03
Category: Renal & Urinary Systems
Discipline: Paediatrics
Setting: Emergency Department_rural
Topic: Renal disease_Nephrotic Syndrome

Case

You are the intern in a large rural hospital emergency department. Ellie Croft is an 8-year-old girl who presents with her parents. They are concerned because Ellie awoke this morning with a swollen face and hands. They wonder if she has been bitten by something during the night.
You observe that Ellie has oedema present in her face, hands and feet. She appears pale, but is not acutely unwell.
Vital signs: Afebrile, BP 100/75, HR 90 bpm; RR 20/min

Questions

1. You are concerned that Ellie may have nephrotic syndrome. What are the key clinical features of nephrotic syndrome?
2. Urinanalysis confirms the presence of proteinuria and her serum albumin is low. Describe the pathophysiology of the features of nephrotic syndrome and describe the clinical features seen in minimal change nephrotic syndrome.
3. Outline the principles of management of nephrotic syndrome.
4. Ellie is admitted and commenced on a course of oral prednisolone; she does not develop any complications and is discharged home after one week. What are the possible complications of minimal change nephrotic syndrome and why?
5. Ellie is followed up a few weeks later by the paediatrician, unfortunately she has persistent proteinuria and a renal biopsy is recommended. Why is a renal biopsy undertaken in this context?
6. The diagnosis of minimal change nephrotic syndrome is confirmed and the prednisolone is continued for 6 months. Outline the presentation of relapses of minimal change nephrotic syndrome and the indications for cyclophosphamide use.

Suggested reading:

- South M, Isaacs D editors. Practical Paediatrics. 7th edition. Edinburgh: Churchill Livingstone;2012.

ANSWERS

1. You are concerned that Ellie may have nephrotic syndrome. What are the key clinical features of nephrotic syndrome?

Nephrotic syndrome is defined as oedema, proteinuria, hypoalbuminaemia and hyperlipidaemia. Clinical features – facial/periorbital oedema particularly on waking/scrotal vulval, leg and ankle oedema/breathless due to pleural effusion and abdominal distension.

2. Urinanalysis confirms the presence of proteinuria and her serum albumin is low. Describe the pathophysiology of the features of nephrotic syndrome and describe the clinical features seen in minimal change nephrotic syndrome.

Aetiology: There is alteration in the glomerular anionic status. Sensitised lymphocytes secrete a number of lymphokines that alter the normal, negatively changed sialoproteins on the glomerular basement membrane – loss of the membrane negative charge allows anionic proteins to leak across the basement membrane into Bowman's space (develop proteinuria/hypoalbuminaemia and hyperlipidaemia).

Clinical features of minimal change nephrotic syndrome: Generalized oedema is the usual presenting system with facial/periorbital oedema particularly on waking/scrotal vulval, leg and ankle oedema breathless due to pleural effusion and abdominal distension (diagnosis by heavy proteinuria and low plasma albumin).

3. Outline the principles of management of nephrotic syndrome.

Low salt diet is encouraged. Fluid intake generally not restricted because of risk of hypovolaemia but mild restriction may be of benefit with significant oedema. In 90% prednisone induces a remission.

4. Ellie is admitted and commenced on a course of oral prednisolone; she does not develop any complications and is discharged home after one week. What are the possible complications of minimal change nephrotic syndrome and why?

Possible complications are infection, hypovolaemia and a hypercoagulable state.
Infections → related to loss of opsonins and immunoglobulins in the urine.
Hypovolaemia → due to the loss of plasma water into the tissues with consequent fall in the circulating blood volume (occurs in 5% of cases, suspected if oligura<100ml/day/poor peripheral perfusion/ abdominal pain/tachycardia or postural hypotension and confirmed by high haematocrit and low urine sodium).
Hypercoagulable state → due to haemoconcentration and loss of antithrombin III in urine.

5. Ellie is followed up a few weeks later by the paediatrician, unfortunately she has persistent proteinuria and a renal biopsy is recommended. Why is a renal biopsy undertaken in this context?

Biopsy recommended if unresponsive to steroids or if atypical features to exclude a more sinister diagnosis.

6. The diagnosis of minimal change nephrotic syndrome is confirmed and the prednisolone is continued for 6 months. Outline the presentation of relapses of minimal change nephrotic syndrome and the indications for cyclophosphamide use.

Approximately 70% of children have relapses often associated with URTI, frequent relapses can be prevented by regular prednisone (5 mg – 15 mg on alternate days). Cyclophosphamide is indicated when steroid side effects become significant (such as cushingoid appearance, reduced growth).