Pseudo-angioedema

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Presentation

- 63-year-old lady presented to A & E with facial swelling for 1 year
- Referred to acute medical team as ‘angioedema ? cause’
- Fixed, progressive, non-tender firm facial swelling initially of the forehead spreading to cheeks and chin
- The lips and other cutaneous sites never involved
- No ‘rash’ or urticaria at any site ever noted
- No triggering factors noted including no foods, medications (only taking HRT), injectants including cosmetic procedures
- Systemically unaffected including no abdominal symptoms
- Improved slightly with short courses of oral prednisolone, but had never fully resolved being persistent and progressive
- Not improved with antihistamines

Investigations

- Apyrexial and haemodynamically stable
- Mild normocytic anaemia
- Inflammatory markers, renal and liver function, ferritin, thyroid function - all normal
- IgE, autoantibodies, C3 and C4, serum ACE, HIV, Hepatitis B and C, T-SPOT - all normal/negative
- Chest x-ray - normal heart and lungs

Management

Blood tests and imaging ruled out any infective process so oral prednisolone 0.5mg/kg/day was initiated which brought about improvement after a few days and maintained improvement over the next few weeks.

Clinical Image

Obvious facial swelling mimicking angioedema – so called pseudo-angioedema. Swollen areas felt very firm and indurated (‘woody induration’) which was not however, along with the persistent and non-epidodic history, in keeping with true angioedema.

Causes of Pseudo-angioedema

- Acute Contact Dermatitis
- usually from hair dye
- Dermatomyositis
- Hypothyroidism
- Foreign body reactions – e.g. to injected cosmetics
- Orofacial Granulomatosis (OFG) – granulomatous inflammation usually in association with Crohn’s disease affecting lips
- Melkerson-Rosenthal syndrome – idiopathic granulomatous inflammation of face
- Morbus Morbihan disease – upper half of face often associated with rosacea
- Superior Vena Cava Syndrome,
- Subcutaneous Emphysema
- Cluster Headache

Not an exhaustive list.

Skin Biopsy

A skin biopsy was performed on the day of presentation. This subsequently revealed a dense and florid foreign-body granulomatous reaction of the subcutaneous fat. With this result the patient was questioned about possible cosmetic procedures she may have had, and remembered having collagen fillers (of uncertain type) some 5 years ago.

Conclusion

- Not everything that looks like angioedema is angioedema.
- Awareness of diagnoses mimicking angioedema allows a careful history and examination to make a correct diagnosis, or at least rule out ‘true’ angioedema.
- This will avoid unnecessary anxiety, admissions, investigations and treatments.

KEY LEARNING POINTS

1. Angioedema is characterised by sudden onset recurrent short-lived/episodic swelling of the skin and mucous membranes, especially the face and lips. It is usually acquired IgE mediated but rare cases of hereditary angioedema exist
2. Angioedema is a deep form of urticaria
3. In Hereditary angioedema (C1 esterase inhibitor deficiency) there is no urticaria ( rash of weals), but bowel symptoms/abdominal pain occur which can mimic an ‘acute abdomen’ – normal C4 levels help exclude the diagnosis.
4. An awareness of conditions mimicking angioedema is important.

References