Estimates suggest that 70,000 people in the UK have symptomatic HH; only about 3,000 cases are recognised [1]. This may be due to onset being later than previously believed [2] and variable phenotypic penetrance in homozygotes; one study has suggested a penetrance of <1% [3]. This raises issues for population screening.

Optimal treatment involves regular venesection, which is not without pitfalls in older people, who often have other co-morbidities and struggle with the logistics of frequent hospital visits [4].

Treatment prevents progression to cirrhosis in pre-cirrhotics and also the onset of non-hepatic complications. Some symptoms improve but there is no effect on diabetes, arthritis or impotence. The presence of cirrhosis dramatically reduces prognosis, and treatment can only slow progression to liver failure; it has no impact on the incidence of hepatocellular carcinoma which causes death in 30% of patients [5].

Since older patients may still only have mild disease at presentation, HH is an important diagnosis to consider. This is not only for screening family members, but also because early intervention can decrease morbidity in a group whose life expectancy has increased over the last 50 years.

Key points

- HH is not commonly diagnosed in the elderly; it may often be missed due to its multifaceted presentation mimicking common geriatric conditions.
- Older people may present with only mild disease and therefore gain significant benefit from treatment.

Conflict of interest

None.

Informed consent

The patient discussed in this report has given written consent for using details of her case for purposes of medical education, including publication in a scientific journal.

References


Severe exfoliative dermatitis caused by strontium ranelate: two cases of a new drug reaction

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Abstract

Strontium ranelate is a relatively new drug used as a second-line treatment for osteoporosis, often targeted at older patients. It is known to cause skin rash and rarely drug reaction with eosinophilia and systemic symptoms, but there are no reports of exfoliative dermatitis as a reaction in the literature. We present the first two cases of this adverse effect of the drug, combined with
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eosinophilia but no systemic symptoms. We illustrate the significant morbidity involved and use of systemic steroid treatment required, highlighting the need for awareness of this reaction in medical and particularly in elderly care communities.

**Keywords:** anti-osteoporotic, drug reaction, elderly, exfoliative dermatitis, osteoporosis, rash, strontium ranelate

We present two cases of exfoliative dermatitis with eosinophilia but no systemic symptoms as a reaction to strontium ranelate, causing significant morbidity in elderly patients. This new anti-osteoporotic medication is recommended by the National Institute of Clinical Excellence as second-line treatment after bisphosphonates for primary and secondary prevention, with marketing often focused at an older population who are more vulnerable to cutaneous drug reactions. By inhibiting bone resorption, the medication increases bone mineral density and reduces fracture incidence [1]. Diarrhoea is the commonest side effect; skin rashes are recognised. However, we are unaware of any previous publications of this particular presentation, although the Medicines and Healthcare products Regulatory Agency (MHRA) yellow card system identified one case.

The first patient was a lady aged 83 admitted by the orthopaedic surgeons with a fall and fractured neck of femur. Her medical history included sarcoidosis, hypothyroidism and lymphoedema. She took only long-term thyroxine prior to this hospitalisation but was started on strontium 2 g at after her operation. Forty-one days later, she noticed itchy, urticated macular erythematous lesions on her back, arms and chest. A skin biopsy demonstrated features typical of a drug eruption; her eosinophil count was raised at 3.4. The rash developed over a few days into a generalised exfoliative dermatitis (Figure 1) requiring potent topical steroids and high-dose oral prednisolone. The strontium was withdrawn. She remained systemically well and over a month the rash slowly improved, eventually resolving.

Case two involved a 75-year-old lady whose medical problems included porphyria cutanea tarda, vasculitis, hypertension, osteoporosis and vitamin B12 deficiency. She was allergic to penicillin, which caused a rash, and normally took calcichew, lansoprazole, strontium and co-amoxiclav. She was admitted with an *Escherichia coli* urinary tract infection that was treated with 11 days of meropenem and 14 days of vancomycin intravenously. She had taken strontium for just 4 days prior to hospitalisation. After 24 days as an inpatient and 10 days after completing her antibiotics, she developed itchy erythematous lesions on her back, buttocks, abdomen and extremities. Over a week, these progressed to a generalised exfoliative dermatitis. Her eosinophils rose to 2.46 but liver function remained normal with no systemic symptoms. Potent topical steroids and oral prednisolone for 7 days gave minimal improvement. She experienced 6 weeks of rash before strontium was identified as the potential cause and stopped. Three weeks after withdrawal of the drug, the symptoms resolved. Despite a recognised potential cross-reactivity with penicillin and meropenem, the antibiotics were stopped 10 days before the rash started making strontium a far more convincing culprit. The onset just a month after this drug was initiated; the prolonged symptomatic period and resolution after removal also strongly support causation.

Exfoliative dermatitis is a rare presentation of a dermatological drug reaction, accounting for just 2.5% in a recent series [2]. We could find no published examples resulting from strontium. The MHRA is aware of one episode. The British National Formulary warns clearly of the risk of drug reaction with eosinophilia and systemic symptoms (DRESS syndrome), which occurs infrequently, with the first two case reports recently published [3]. The patients here do not fit the criteria for DRESS syndrome having some features but not systemic symptoms, defined as fever, lymphadenopathy or internal organ involvement; however, both required oral steroids. We suggest that these cases demonstrate a severe exfoliative reaction causing significant morbidity in a vulnerable elderly patient group more likely to experience drug eruptions [4] to a new and emerging drug, strontium ranelate.

These adverse skin effects are potentially under-recognised and of particular importance in the older population the medication targets.

**Conflicts of interest**

None.

**References**


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