Clinical review

Interactive case report

A 66 year old woman with a rash: case outcome

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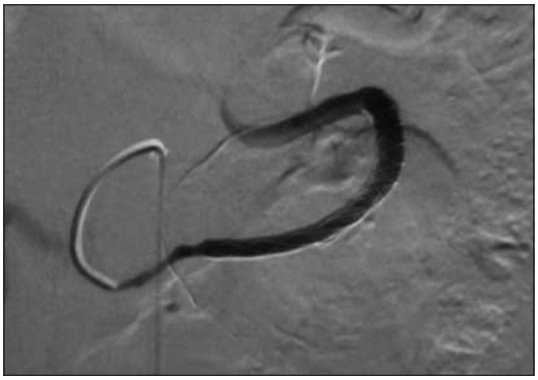
This is the final part of a 3 part case report which describes the outcome and summarises the comments made by readers during the case presentation

Three weeks ago (*BMJ* 2003;326:588) we published the case of Ruth, who presented to her general practitioner with a rash after feeling unwell for some time. She had dermatomyositis diagnosed and, during investigation for possible underlying causes, was found to have an aneurysm of the splenic artery (*BMJ* 2003;326:640). Ruth's case was discussed at a multidisciplinary meeting and a decision was made to treat her aneurysm with a covered stent placed under fluoroscopic guidance by a radiologist. A theatre space was booked for the same time as the procedure in case of complications. She had a repeat angiogram and a wire was passed across the aneurysm.

Surgery and follow up

Two covered stents were placed across the neck of the aneurysm and a repeat contrast injection showed that there was no communication between the splenic artery and the aneurysm sac (figure). Ultrasonography the next day showed the aneurysm filled with thrombus with no flow on colour Doppler. The following day contrast enhanced computed tomography showed no evidence of a leak. Follow up computed tomography continued to show no leak and no flow in the aneurysm sac. Ruth was given pneumococcal and haemophilus influenza vaccines as a prophylactic measure in case of future hyposplenism caused by migration or occlusion of the stent.

As it was envisaged that Ruth would be taking steroids long term, she was referred for bone densitometry (dual *x* ray absorptiometry), which showed no evidence of osteoporosis. Ruth was reassured but advised that it would be sensible to increase her calcium intake to 1500 mg a day. She found this difficult to achieve and so was started on an oral calcium and vitamin D supplement. She was also advised to take as much weight bearing exercise as possible. The steroid dose was kept as low as possible. A further consideration is whether Ruth's relatives should be screened as she and her sister have both had aneurysms.



Angiogram after placement of the splenic artery stent showing good flow through the stent but no leakage of contrast into the aneurysm sac

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We thank D Gould, S Travis, G Maskell, and K Woodburn, who were responsible for Ruth's hospital management.

Commentary: Patient's perspective

Ruth with the support of Richard Farrow

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I have found being involved in this new interactive report very interesting. I wanted to put something back. My doctors have been marvellous and have done everything possible to help me. Right from my first consultation, my general practitioner told me what he thought I had and said that I should see a dermatologist at the hospital. I had only just got home when he telephoned with the appointment date.

It's better to know

How have I felt about my condition being discussed? Well I am happy to have all this discussion about my condition. I would prefer to know what is going on and often talk to my husband about these things. Nevertheless, the comments on the web have reminded me that I have a few unusual complaints—it would be nice to have something common and ordinary.

There have been a few comments about my mammogram. I would like to say that I really believe in screening and am screened regularly. If they made it compulsory, I would be close to the front of the line. Other doctors seem to feel that I should not have been told about the risk of cancer with the dermatomyositis. I think this is wrong; it is better to know so that you understand why you have been referred for all these tests. It is better to be prepared from an early stage. It's not as if I don't have experience of cancer; my mother and grandfather both died of bowel cancer.

Many of the contributors ask about weakness in my muscles. This was not really an issue at the start. It was just my rash. But now I am beginning to have difficulties in walking up the stairs.

There were several comments about the aneurysm. My friends were very worried and felt that I should not go out on my own. My husband even talked about giving up bowls, but what was the point? He could have been mowing the lawn, and if I was inside and collapsed, he still could not have helped.

It has been interesting to see comments from around the world. It has also let me as a patient have insight into what a doctor is thinking. The doctors are talking to you about one thing, perhaps your rash, and thinking something completely different, such as does she have cancer.

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Commentary: Vascular surgeon

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Splenic artery aneurysms are an incidental finding in almost 1% of abdominal angiograms. They are usually less than 2 cm in diameter and often affect the distal splenic artery. Their association with multiple pregnancies partly explains why they are four times commoner in women than men. Other documented associations include pancreatitis, portal hypertension, trauma, and, with possible relevance to Ruth, systemic lupus erythematosus.[1]

The main risk is of rupture, and a lifetime rupture rate of 2-10% is often quoted. Although this is not based on hard prospective data, large splenic artery aneurysms such as Ruth's (which was about 10 cm in diameter) carry at least a 10% risk. Coexistent inflammatory conditions such as dermatomyositis may increase that risk. If Ruth's aneurysm had ruptured, her chance of survival would have been about 75%.[2]

Different treatment options

The rapid responses on bmj.com illustrated the different treatment options for these aneurysms. One option is to sit tight on the assumption that the risk of rupture is low and that of intervention relatively high. The traditional surgical approach is laparotomy and ligation of the arteries proximal and distal to the aneurysm, possibly with splenectomy if the aneurysm is in the hilum of the spleen. Ligation can also be achieved laparoscopically without a large abdominal incision.[3] Endovascular approaches include placing a stent graft across the aneurysm to exclude the expanded lumen[4] or coiling the splenic artery to thrombose the aneurysm.[5]

Sitting tight was not a viable option in Ruth's case because of the high risk of rupture. Open surgery would not have been easy because the aneurysm was large and likely to be stuck to the stomach and colon and difficult to remove from the pancreas. Open surgery may also be associated with poor wound healing, chest infection, and deep vein thrombosis. When the spleen has to be removed, a subphrenic abscess may form on the splenic bed, and there is a long term risk of increased infection. A laparoscopic approach would have been difficult with such a large aneurysm.

Endovascular occlusion of the aneurysm was another option. But instrumentation of the splenic artery and aneurysm may precipitate rupture, dissection, or distal embolisation of the spleen leading to infarction and pain and possibly some of the complications associated with splenectomy. It would also have been difficult, given the large size of the aneurysm, and recanalisation has been shown in

such cases. Stent grafting, however, is successful with large visceral aneurysms,[4] and it is not surprising that this was considered the best treatment for Ruth.

Follow up

In future, Ruth should have annual computed tomography scans to check that the stent remains in the right position and check for the development of further aneurysms.

Competing interests: None declared.

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Commentary: Dermatology

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Although dermatomyositis is rare, its cutaneous signs are characteristic. It was therefore not surprising that two thirds of the responses to the initial presentation of Ruth's case agreed with the general practitioner's provisional diagnosis. In classic cases a purplish-red heliotrope erythema occurs on the eyelids, upper cheeks, forehead, and temples. Small, erythematous or violaceous, flat papules (Gottron's papules) and small plaques occur over the knuckles and on the dorsal surface of the finger joints. This is associated with aching and muscle weakness, which may appear later, and in time results in muscle atrophy.

The diagnosis is confirmed by muscle biopsy, electromyography, and raised serum concentrations of creatine phosphokinase. Glutamic oxalacetic transaminase concentrations are often also raised. The cause is unknown, but there is increasing evidence of early damage to blood vessels, probably humorally mediated.[1]

Neoplasia

The overall incidence of underlying neoplasia in reported series varies from 29% to 40% of patients aged over 40 years. The primary tumour most commonly occurs in the lung, breast, female genital tract, stomach, rectum, kidneys, or testis. In Chinese people, nasopharyngeal carcinoma is the commonest underlying malignant disease. The condition may also be associated with lymphomas.[2] Dermatomyositis precedes the neoplasm in 40% of cases.[3]

Neoplasia seems to be more common in men than women.[4] Skin manifestations tend to worsen in parallel with the growth of the neoplasm but may improve when the cancer is treated. Neoplasia may be missed because of failure to reinvestigate relapse of previously stable dermatomyositis.[5] Ruth has a strong family history of cancer and therefore she should be offered regular screening tests, including chest radiography, mammography, pelvic ultrasonography and a test or tests for tumour markers. The advice that several respondents gave about sun protection is essential, and Ruth should be reassured that she cannot pass the condition on to her children.

Competing interests: None declared.

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Commentary: View from primary care

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As Ruth's case has unfolded her general practitioner has had several issues to discuss with her. Her initial presentation was unusual, and as responders to bmj.com underlined, her general practitioner did well to make a provisional diagnosis of dermatomyositis. But even in the absence of a working diagnosis, it was evident that Ruth needed urgent investigation. Many respondents reflected on the appropriate extent of investigation before referral to a specialist. Some mentioned the possibility of diagnostic support via the web. Pictures are available at http://dermis.multimedica.de (a commercially sponsored site) and www.gpnotebook.co.uk offers accessible information, although not its evidence base.

What to tell Ruth

Once the diagnosis of dermatomyositis had been raised, there was the difficult question of what to tell Ruth about the link with malignancy. General practitioners do not carry figures for this in their heads, and although Medline is accessible, it takes time to appraise. Many patients (though not all) want us to discuss unpleasant possibilities openly.[1] The general practitioner had to judge what Ruth wanted to know,

calling for good communication skills including possible admission of uncertainty. Sharing the referral letter to secondary care may help transparency.[2]

While Ruth was having extensive investigation in hospital, it will have been important to ensure that the information she was given from general practice was consistent with what the hospital was telling her and that her perceptions and concerns were being addressed. Although Ruth's stenting was planned at a case conference, her informed consent to the intervention required sharing decision making. The best way to do this is open to debate,[3] and many patients turn to their general practitioners before making their mind up. Prompt and clear communication between specialists and general practitioners and delineation about who is doing what—for example, regarding vaccination[4]—is vital.

Ruth experienced adverse effects from azathioprine and risks more while she continues taking steroids. The aims, risks, and benefits of treatment need to be discussed with her, because her views will ultimately determine whether she takes prescribed drugs.[5]

Because Ruth is receiving ongoing treatment with steroids her general practitioner must monitor her for glucose intolerance and hypertension and consider treatment to prevent osteoporosis. Two recent reviews are relevant for steroid induced osteoporosis,[6 7] and Ruth may find the support of the National Osteoporosis Society (www.nos.org.uk) helpful. Ruth's perceptions and those of her general practitioner about new symptoms will be influenced by this illness: although her condition is unusual, many of the issues it has raised in general practice are common.

Competing interests: None declared.

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Commentary: Learning from interactive case reports

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Ten days after Ruth's case was published, over 60 doctors had engaged in the web discussion around diagnosis, investigation, and patient communication. A wide range of specialties has been represented, and just over a third of the contributors work outside the United Kingdom. One of the aims of publishing interactive case reports is to encourage interdisciplinary learning, and it has been good to see the process in action.

Communication with the patient has been a major focus. "To know what I would tell Ruth at this stage, I need to know what she wants to know…What are her concerns and expectations?" asked Judith Harvey. We then heard from Ruth, and her perspectives were illuminating. Whereas doctors were expressing concern about her risk of developing malignancy, she was worrying about steroid induced cataracts.

John McCormack observed that hospital specialists seemed much keener to share their worries with the patient at an early stage than general practitioners. Only one out of 19 general practitioners favoured telling Ruth about the possibility of malignancy when she first presented with dermatomyositis, compared with seven out of 11 specialists.

Subtle differences in approach

A web forum where primary and secondary care physicians can discover subtle differences in their approaches is, for me, an important feature of this new learning exercise. I was therefore interested in the dialogue on how many investigations the general practitioner should do. Liam Farrell favoured prompt referral to secondary care. Jan Van Hollebeke, by contrast, suggested that the general practitioner needed to take simple blood tests straight away so that Ruth could make "an evidence based informed decision" about how she should be investigated further.

Stuart Skeates, a general practitioner, and Colin White, a physician, considered what investigations are helpful for dermatomyositis. After reading the wide diversity of opinions on this on the web, I found Natasha Kapur's expert commentary helpful. Similarly, Linda Hand's synthesis has helped me see more clearly after the technical debate on the web around vascular surgery and interventional radiology.

Can we learn from rare conditions? There were pleas that "Trafalgar Square pigeons" make for better learning than "lesser spotted, greater crested grebes"—and that was before the discovery of Ruth's splenic artery aneurysm. It is a fair point, but others found this case challenging and interesting precisely because it has been a bit out of the ordinary. We certainly need plenty of case material around bread and butter medicine. What this case has managed to do, however, is to provoke a rich response from a wide range doctors, and I look forward to the next one.

Competing interests: None declared.

We welcome contributions of interactive case reports. Cases should raise interesting clinical, investigative, diagnostic, and management issues but not be so rare that they appeal to only a minority of readers. Full details of criteria are available at bmj.com/cgi/content/full/326/7389/564/DC1