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CLINICAL CASE REPORT

Myocardial infarction in a patient with hereditary haemorrhagic telangiectasia in a remote location

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ABSTRACT

Context: Prompt reperfusion therapy in acute ST-segment elevation myocardial infarction (STEMI) improves outcomes and is therefore a key part of the management of affected patients. Primary percutaneous coronary intervention (PPCI) is preferred over thrombolysis; however, when PPCI is not available within an acceptable time frame, thrombolysis should be administered without delay. When thrombolysis is contraindicated, this can lead to delays in achieving coronary artery reperfusion, and making therapeutic decisions is more challenging.

Issues: A 57-year-old woman with hereditary haemorrhagic telangiectasia (HHT) presented with chest pain to local medical services on a Scottish island. Initial assessment including 12-lead ECG was performed without delay and led to a diagnosis of infero-lateral STEMI. Initial treatment was with antiplatelets, nitrate and opiate analgesia. Immediate reperfusion therapy with thrombolysis was considered; however, this was contraindicated due to the diagnosis of HHT. Following discussion with the regional centre she was treated with heparin and urgently transferred to the nearest catheterisation laboratory for PPCI. Unfortunately delays in transfer were caused by inclement weather conditions, which precluded the use of an air ambulance. The patient reached a cardiac catheterisation laboratory by road. A total occlusion of a distal branch of the circumflex artery was found to be the culprit lesion and despite wiring and ballooning good blood flow was not restored. However, she made a good recovery and was discharged home on secondary prevention therapy, which was modified to take into account her increased bleeding risk associated with her pre-existing HHT.

Lessons learned: In this case, the patient presented early to medical services following the onset of symptoms and was immediately assessed, resulting in a prompt accurate diagnosis at first medical contact. Despite this, the presence of HHT, and the particularities of living in a remote area compounded by unfavourable weather conditions, resulted in a delay in definitive treatment that would have been available to a similar patient presenting in an urban setting. In remote regions where reperfusion therapy for

acute STEMI relies more often on thrombolysis, an increased awareness of the issues around the contraindication of thrombolysis, together with early discussion with a regional cardiology service leads to the most efficient way of establishing the best treatment plan for individual patients. Despite this, rural patients may remain at a disadvantage.

Key words: hereditary haemorrhagic telangiectasia, primary percutaneous coronary intervention, ST elevation myocardial infarction, thrombolysis.

Context

The current preferred treatment of acute ST-segment elevation myocardial infarction (STEMI) is primary percutaneous coronary intervention (PPCI). This is the optimal treatment provided it can be delivered within a reasonable time frame (90–120 min)[1] of diagnosis. However, in remote areas many patients with acute STEMI may not have immediate access to a cardiac catheterisation laboratory (cath lab). One such area is the Highlands of Scotland where the potentially long transfer times to the nearest cath lab might lead to unacceptable delays to achieving coronary artery reperfusion, and thus thrombolysis in this area remains the mainstay of initial reperfusion strategy for many patients with acute STEMI.

We report a case of STEMI in a patient with hereditary haemorrhagic telangiectasia (HHT) and discuss the issues surrounding the immediate and subsequent treatment.

Case

A 57-year-old woman living on a Scottish island with a past medical history of HTT, hyperlipidaemia and hypothyroidism woke at 4.00 am with severe central chest pain associated with sweating and vomiting. Her husband called for an ambulance but unfortunately all three ambulances on the island were unavailable at the time due to other clinical cases, so she was driven to the local hospital and arrived shortly after.

She was assessed promptly and an ECG performed at 4.19 am which revealed ST segment elevation in leads II, III, aVF, V5 and V6, consistent with a diagnosis of an infero-lateral STEMI (Fig1).

Initial treatment with aspirin 300 mg and clopidogrel 300 mg, sublingual glyceryl trinitrate, diamorphine 2.5 mg and metoclopramide 10 mg was promptly given at 4.25 am. After taking a clinical history and assessing the patient as per protocol, the decision had been made to administer thrombolysis (due to potentially long transfer times to the nearest cath lab). As preparations were made for thrombolysis, the patient then revealed her diagnosis of HHT, which only came to light when using the local thrombolysis contraindications check list. The diagnosis of HHT had been made 15 years previously. The patient had previously confirmed pulmonary telangiectasia but no documented cerebral involvement, and the symptoms from her HHT were limited to occasional epistaxis and chronic microcytic anaemia treated with oral iron replacement.

At this stage, advice was sought from the regional centre. After discussion, as the bleeding risk was deemed high, the patient was not given thrombolysis; the standard intravenous heparin (5000 IU bolus) was given instead as a pragmatic compromise, with a plan for immediate transfer of the patient to the nearest cath lab.

An air ambulance transfer was not possible due to inclement weather conditions (fog). The patient was transferred by road to the nearest cath lab where she arrived at 9.50 am. Coronary angiography via the right radial approach revealed a complete occlusion of a small distal coronary artery branch of

the circumflex artery. Following aspiration and ballooning, there was poor flow in the very small distal vessel, so treatment was conservative and no stent was inserted.

The patient was subsequently transferred to the coronary care unit for ongoing treatment. Troponin I was raised at 2.55 µg/L (normal value <0.04 µg/L), renal function was normal, haemoglobin was 99 g/L and mean corpuscular volume was 72.9 fL.

She was commenced on standard treatment with aspirin 75 mg, atorvastatin 80 mg, bisoprolol 2.5 mg and perindopril 2 mg (all once daily). Because of the potential bleeding risk, and the fact that no coronary stent had been deployed, a second antiplatelet was not administered. Transthoracic echocardiogram demonstrated mild to moderate left ventricular systolic dysfunction. The patient was discharged home on day 3 and has remained well.

Issues

This case report describes a difficult clinical situation where the management of a patient with a STEMI in a remote setting was further complicated by a rare pre-existing medical condition (HHT), which precluded the use of thrombolysis, while geographical remoteness and inclement weather conditions impeded immediate transfer of the patient to a cardiac cath lab.

In this case, no ambulances were available when the initial call for help was made. In our area the standard management is for every patient to have an ECG performed at the site of incident and for the administration of thrombolysis in the prehospital setting to be facilitated via telemetry-enabled decision support[2,3]. This was not possible in this case due to lack of ambulances; this is an issue in remote settings where the organisational resilience to multiple calls may be less than in urban centres. Furthermore, not all ambulances in remote areas are staffed with a paramedic crew and thus there may be variation in practice depending on which crew members are on shift.

Nevertheless, this patient was quickly transferred to the local hospital within 20 minutes and prompt assessment occurred. The major issues in this case were the administration of antiplatelet and antithrombotic medications in a patient with HHT. Although this patient with known pulmonary telangiectasia was reported to have no cerebral telangiectasia, this does not confer a low risk of intracerebral bleed, and the risk of life-threatening pulmonary haemorrhage is also recognised[4]. HHT is a rare contraindication to thrombolysis, but other more common contraindications exist (Table 1). The decision not to administer thrombolysis in this case was a pragmatic clinical decision as there were no guidelines or published reports about how to treat patients with HHT who are having a myocardial infaction. The decision to withhold thrombolysis was based on the perceived risks of intra-cranial or intra-pulmonary bleeding, and the potentially catastrophic consequences of both, balanced against the potential risk of delayed treatment of an inferior myocardial infarction. In this case, a non-medical nurse prescriber was on shift who was able to initiate treatment with aspirin and clopidogrel and consider thrombolysis, but this is not always the case in rural hospitals with less than continuous 'doctor cover'.

Previous close work between the remote hospital and regional centre allowed immediate access by telephone to a consultant cardiologist and the case was discussed.

Once the decision had been made not to administer thrombolysis, transfer to a cath lab was found to be problematic due to inclement weather conditions. This illustrates another issue with remote patient care: despite availability of an air ambulance, weather may remain a barrier to prompt access to definitive case and thus remote patients remain at a potential disadvantage when compared with urban dwellers.

In this case, the patient had an occlusion of a relatively small distal vessel and made a good recovery from her myocardial infarction; thus the impact of the above issues on her individual case was less than it may have been had she had a more proximal vessel occlusion.

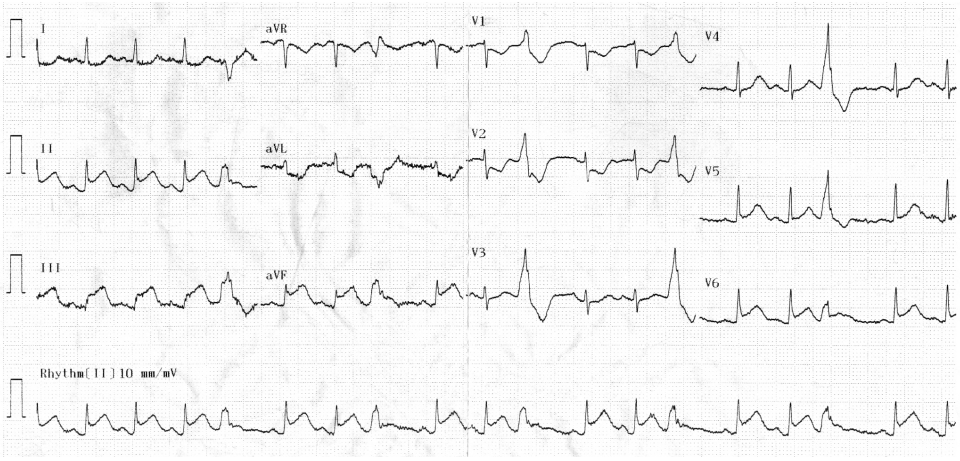

Figure 1: First ECG recorded after initial medical contact showing infero-lateral ST segment elevation and reciprocal anterior ST segment depression, as well as ventricular ectopy.

Table 1: Contraindications to the administration of thrombolysis in acute ST segment elevation myocardial infarction[1]

Absolute contraindications
Previous intracranial haemorrhage or stroke of unknown origin at any time
Ischaemic stroke in the preceding 6 months
Central nervous system damage or neoplasms or atrioventricular malformation
Recent major trauma/surgery/head injury (within the preceding 3 weeks)
Gastrointestinal bleeding within the past month
Known bleeding disorder (excluding menses)
Aortic dissection
Non-compressible punctures in the past 24 hours (eg liver biopsy, lumbar puncture)
Relative contraindications
Transient ischaemic attack in the preceding 6 months
Oral anticoagulant therapy
Pregnancy or within 1 week postpartum
Refractory hypertension (systolic blood pressure >180 mmHg and/or diastolic blood pressure >110 mmHg)
Advanced liver disease
Infective endocarditis
Active peptic ulcer
Prolonged or traumatic resuscitation

With respect to opportunities to avoid the myocardial infarction, despite hypercholesterolaemia, this relatively young, non-smoking, normotensive, female patient had few risk factors and would have been considered at low risk of a cardiovascular event. Following local guidelines, her low overall risk would not have warranted treatment of hypercholesterolaemia in the absence of other risk factors.

Lessons learned

This case illustrates some of the issues encountered when providing health care in remote areas and the importance of close communication between remote hospitals and regional centres.

Where thrombolysis is frequently the chosen reperfusion strategy due to accessibility, it is important to have an increased awareness of pre-existing conditions that could constitute contraindications for thrombolysis and could lead to life-threatening complications. In this case, use of an established safety check list and early discussion resulted in appropriate withholding of thrombolysis.

In such situations, early discussion with a cardiac specialist centre is vital to provide support and help make the most appropriate individualised treatment decisions. This is especially important with rare conditions such as HHT, when antiplatelets and antithrombotic treatment rely mostly on expert opinion and anecdotal evidence[5] in the absence of a sufficient evidence base to inform standardised therapies.

However, despite good network support, remote services are more likely to be variable – in our area not all ambulances are staffed with paramedics and not all community hospitals that receive patients with myocardial infarction can immediately prescribe appropriate drugs. Thus, potentially life-saving treatment with thrombolysis could be delayed. One could argue that the ability to deliver such therapies is even more critical in remote areas, where transfer to a cath lab for PPCI is not always possible.

This case illustrates that despite distances involved and potential delays, direct transfer to a cath lab may still remain a safer option compared to thrombolysis for some remote patients with STEMI.

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