Spontaneous Intramuscular Haemorrhage: A Rare Complication of Dermatomyositis

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AUTHORS' CONTRIBUTIONS
All authors contributed to this case report.

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Nil.

CONSENT
Yes.

HUMAN AND ANIMAL RIGHTS
Not applicable.

ABSTRACT

Spontaneous intramuscular haemorrhage is a rare complication that can occur in patients with dermatomyositis. We describe a case which was radiologically diagnosed on computed tomography and the management of which was complicated by her malignancy-associated prothrombotic state. Knowledge of this potentially fatal complication and its radiological features are necessary for timely recognition and management.

CASE REPORT

BACKGROUND

Dermatomyositis is an immune-mediated inflammatory myopathy that presents with characteristic cutaneous manifestations and symmetrical proximal muscle weakness. Spontaneous intramuscular haemorrhage is a rare complication of dermatomyositis which may be under-recognised due to its infrequency and consequent limited familiarity amongst clinicians. This case serves as a reminder of both the clinical and radiological findings of spontaneous intramuscular haemorrhage that can facilitate timely diagnosis, along with the treatment challenges it presents.

CASE REPORT

A 70-year-old female presented to the emergency department describing new right thigh pain. She had no history of trauma or recent falls. On examination, she had tenderness over the right anterior thigh without swelling or bruising.

She had recently been diagnosed with anti-transcriptional intermediary factor-1 gamma (TIF1-γ) antibody positive dermatomyositis just two weeks prior for which she on treatment with 30mg once daily oral prednisolone. She had a previous history of Stage 1b clear cell carcinoma of the ovaries (Figures 1,2), treated with surgical resection and adjuvant chemotherapy, completed three months prior to the current presentation. She was on therapeutic dose low molecular weight heparin (LMWH) for bilateral pulmonary emboli.

Laboratory results showed haemoglobin 9.3g/dL (normal range 11.5-16.4g/dL), platelet count 313 x10ºL (normal range 130-400 x10ºL), INR 0.9, PT 10.5s (normal ratio 9.6-12.7s), APTT 29.9s (normal range 25.0-36.5s) creatine kinase 491 IU/L (normal range 26-192 IU/L), CRP 9mg/L (normal range <5mg/L), ESR 20mm/hr (normal range <20mm/hr), lactate dehydrogenase 310 IU/L (normal range 135-250 IU/L) and Ca-125 level of 11 kU/L (normal range 0-35 kU/L).

Her symptoms were attributed to dermatomyositis and, she was discharged home with simple analgesia and a plan for urgent outpatient Rheumatology follow up. One week later, she underwent computed tomography of the thorax, abdomen and

pelvis (CT-TAP) to screen for malignancy given her diagnosis of dermatomyositis.

Imaging findings

CT-TAP showed no lymphadenopathy or recurrence of malignancy however revealed a 3cm heterogeneous expansion of the right proximal rectus femoris muscle and a 7cm heterogenous expansion of the left pectoralis major muscle consistent with intramuscular haemorrhages (Figures 3,4).

Management and follow up

She was reviewed in the outpatient Rheumatology clinic at which time the decision was made to cease anticoagulation.

One month later, she underwent 18-F Fluorodeoxyglucose (FDG) Positron Emission Tomography (PET) which showed diffuse increased tracer uptake in the skeletal musculature, most prominent in the upper limb girdle, consistent with ongoing dermatomyositis (Figure 5). The spontaneous intramuscular haemorrhage was further visualised as an area of photopenia in the left pectoralis musculature (Figure 6), the volume of which had significantly reduced. There were numerous FDG-avid lymph nodes in the para-aortic, common iliac and inferior mesenteric artery territories which were concerning for malignancy. Following multi-disciplinary team discussion, biopsy of these nodes was deemed too challenging and a plan for interval surveillance imaging was advised.

While off anticoagulation, she developed acute left lower limb swelling. Doppler ultrasonography showed occlusive thrombosis of the left peroneal vein and posterior tibial vein with non-occlusive thrombus in the femoral vein. She was reviewed by the Coagulation team who considered both her bleeding risk from dermatomyositis and thrombotic risk from malignancy. The decision was made to restart anticoagulation with therapeutic dose LMWH.

Interval CT imaging performed three months later showed resolution of the intramuscular haemorrhages. Unfortunately, it also showed marked progression of her pulmonary metastatic disease with innumerable metastases throughout all lobes and new extensive bilateral hilar and right paratracheal lymphadenopathy. She was deemed too unstable to undergo endobronchial ultrasound guided biopsy at this time and continued to deteriorate despite a trial of empiric chemotherapy. The decision was made to switch to a palliative approach, and she died shortly thereafter.

DISCUSSION

Aetiology and demographics

Spontaneous intramuscular haemorrhage is a rare complication of dermatomyositis with only twenty-four previously reported cases in the literature [1]. The pathophysiology is poorly understood but thought to be due to a possible relationship between active inflammation of vasculature supplying muscles, anti-thrombotic therapy and high-dose glucocorticoid use [2].

Dermatomyositis is an immune-mediated inflammatory myopathy [2] and has a strong association with malignancy, particularly with lymphatic/haematopoietic, lung, ovary, colon, pancreatic and breast cancers [3, 4]. Patients with dermatomyositis have a five-fold increased risk of malignancy compared to the general population [4]. In patients with high-risk features, such as TIF1-γ and nuclear matrix protein 2 (NXP2) antibodies [3], age >40 years old, severe cutaneous disease, resistance to treatment, prior history of malignancy or absence of interstitial lung disease, prompt relevant screening is recommended [5]. Malignancy may predate, occur concurrently with or occur after the onset of dermatomyositis but is usually recognised within three years of the diagnosis [6]. In our case, the patient developed dermatomyositis shortly after completing treatment for ovarian cancer. The oncology team deliberated whether she had a recurrence of her ovarian cancer or developed a new malignancy but could not obtain a tissue diagnosis. No new primary malignancy was identified on imaging to suggest an alternative origin of this paraneoplastic phenomenon.

Clinical and Imaging findings

Presentation of spontaneous intramuscular haemorrhage is heterogeneous, varying from myalgia with localised swelling to severe anaemia with haemodynamic instability. Presentation is most common in the first six months after onset of dermatomyositis. Iliopsoas is the most commonly affected muscle followed by limb girdle muscles, retroperitoneal muscles and rectus sheath muscles [7]. Treatment with high-dose steroids or use of anticoagulants is associated with increased risk [7].

Non-contrast CT is the first-line imaging test for the diagnosis of spontaneous intramuscular haemorrhage [8]. It can be used to confirm diagnosis of the haemorrhage and determine the site, size and presence of any extension [9]. CT angiography can be used to locate the specific source of active bleeding [8]. Of the twenty-four previously reported cases of spontaneous intramuscular haemorrhage in dermatomyositis, 87.5% had CT performed and 29.2% had CT angiography performed for diagnosis [1].

Following diagnosis of dermatomyositis, patients will frequently be referred for further imaging such as CT-TAP and PET (to investigate for associated malignancy) and chest x-ray or high-resolution CT thorax (to investigate for associated interstitial lung disease). This imaging can be used opportunistically to assess for complications of dermatomyositis including spontaneous intramuscular haemorrhage as in this case.

Treatment and prognosis

Recommended management of spontaneous intramuscular haemorrhage includes discontinuation or reversal of

anticoagulation in the first instance [10] along with supportive measures such as vascular filling and transfusions [8]. Arterial embolization by interventional radiology may be required for larger haemorrhage not responding to conservative management [8]. The concurrent high risk of both bleeding from dermatomyositis and thrombosis from its associated malignancy requires careful consideration. Clinicians should consider Haematology consultation when prescribing prophylactic or therapeutic anticoagulation as was done in this case. It is a potentially fatal condition with its reported mortality as high as 50%, secondary to related complications such as haemorrhagic shock and disseminated intravascular coagulation [1]. Deep muscle involvement is associated with poorer outcomes [1].

Differential Diagnoses

A differential diagnosis of haemorrhagic metastases was also considered in this case. However, given her recent diagnosis of dermatomyositis and treatment with high-dose steroids a diagnosis of spontaneous intramuscular haemorrhage was favoured which was further supported when interval imaging showed significant reduction and later resolution.

CONCLUSION

This case serves as a reminder to remain cognizant of spontaneous intramuscular haemorrhage when reviewing the imaging of patients with dermatomyositis as early diagnosis of this rare but high-risk complication may allow treatment prior to potentially life-threatening progression. This case also highlights the complexity in the management of anticoagulation in these patients with concurrent risk of both haemorrhage related to dermatomyositis and thrombosis associated with their concomitant malignancy.

TEACHING POINT

Spontaneous intramuscular haemorrhage is a rare complication of dermatomyositis which is usually diagnosed on non-contrast CT and CT angiogram. The mainstay of treatment includes supportive measures, withholding anticoagulation and arterial embolization in more severe cases. Although rare, it is important to be mindful of this complication as it is associated with high mortality and managing anticoagulation in these patients can be complex.

QUESTIONS

Question 1: What is the most common site of spontaneous intramuscular haemorrhage

Iliopsoas muscle (applies)
Rectus sheath muscles
Limb girdle muscles
Retroperitoneal muscles
Pectoralis muscle

Explanation: [Iliopsoas is the most commonly affected muscle followed by limb girdle muscles, retroperitoneal muscles and rectus sheath muscles]

Question 2: What is the first line imaging modality for diagnosing spontaneous intramuscular haemorrhage

MRI
Ultrasound
CT (applies)
PET
Xray

Explanation: [Non-contrast CT is the first-line imaging test for diagnosis of spontaneous intramuscular haemorrhage]

Question 3: Which of these are the mainstay of treatment of spontaneous intramuscular haemorrhage

Blood transfusion (applies)
Discontinuation or reversal of anticoagulation (applies)
Arterial embolization (applies)
Antibiotics
Vascular filling (applies)

Explanation: [Recommended management of SIH includes discontinuation or reversal of anticoagulation in the first instance along with supportive measures such as vascular filling and transfusions. Arterial embolization by interventional radiology may be required for larger haemorrhage not responding to conservative management.]

Question 4: Which of these are NOT a high risk feature of malignancy in dermatomyositis?

TIF1-γ or NXP2 antibodies
Age >40 years old
Severe cutaneous disease
Prior history of malignancy
Presence of interstitial lung disease (applies)

Explanation: [In patients with high risk features, such as TIF1-γ and nuclear matrix protein 2 (NXP2) antibodies [4], age >40 years old, severe cutaneous disease, resistance to treatment, prior history of malignancy or absence of interstitial lung disease]

Question 5: Which of these statements regarding spontaneous intramuscular haemorrhage are false

It is a rare complication of dermatomyositis
Risk factors for include treatment with high dose steroids and use of anticoagulants
The highest risk of developing it is within 6 months of diagnosis of dermatomyositis
Deep muscle involvement is associated with better prognosis (applies)
It can be potentially fatal

Explanation: [Deep muscle involvement is associated with poorer outcomes]

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FIGURES

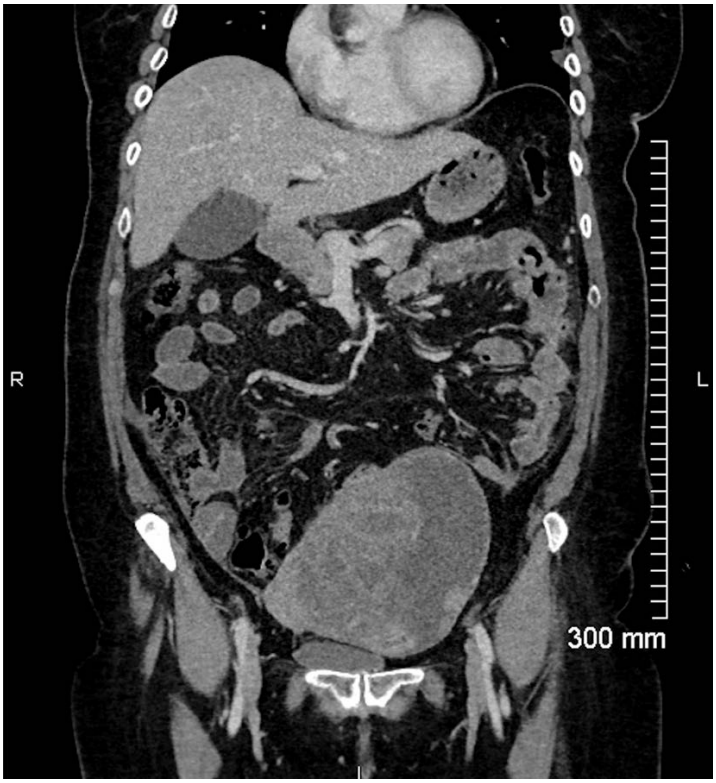

Figure 1: Coronal image from the portal venous phase abdominal CT shows a large pelvic mass measuring 14cm, predominantly solid with some cystic components, and trace free fluid in the pelvis.

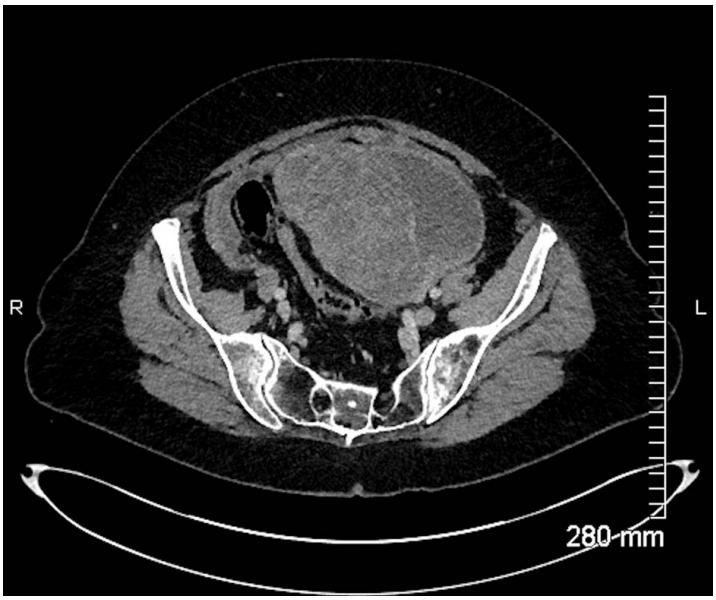

Figure 2: Axial image from the portal venous phase abdominal CT shows large pelvic mass.

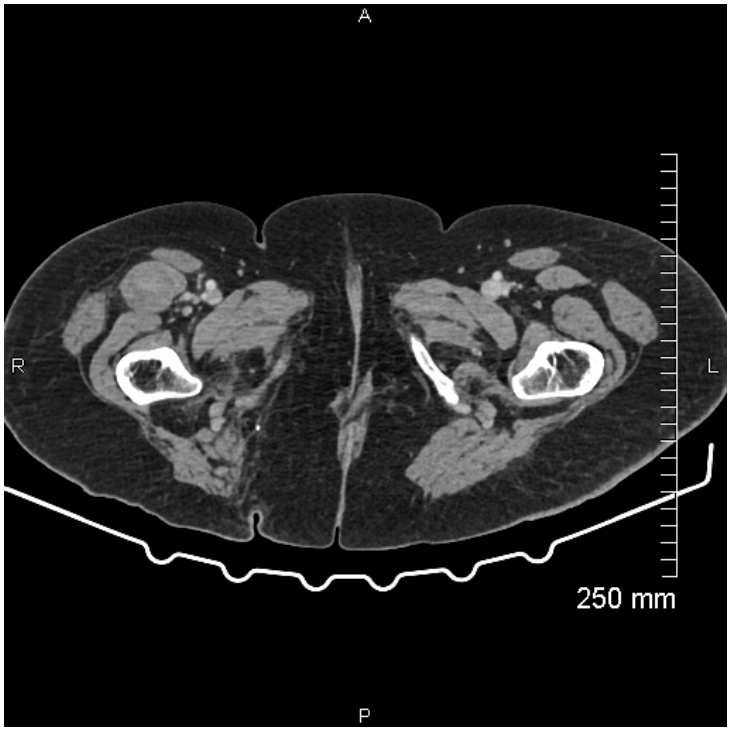

Figure 3: Axial image from the contrast enhanced portal venous phase CT of the thorax, abdomen and pelvis demonstrates a 3cm heterogenous expansion of the right proximal rectus femoris muscle consistent with an intramuscular haematoma.

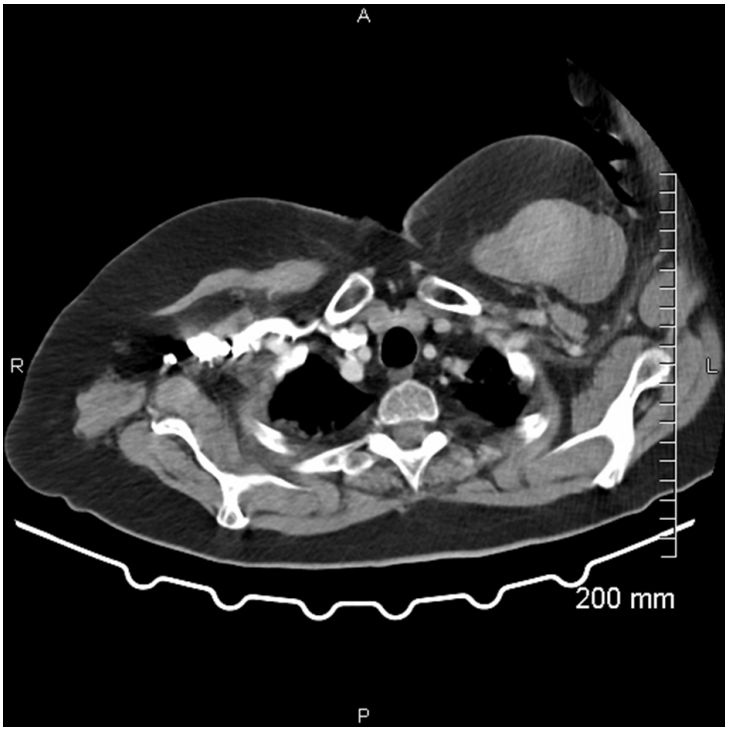

Figure 4: Axial image from the contrast enhanced portal venous phase CT of the thorax, abdomen and pelvis shows heterogenous expansion of the left pectoralis major muscle, measuring 7cm, consistent with intramuscular haematoma.

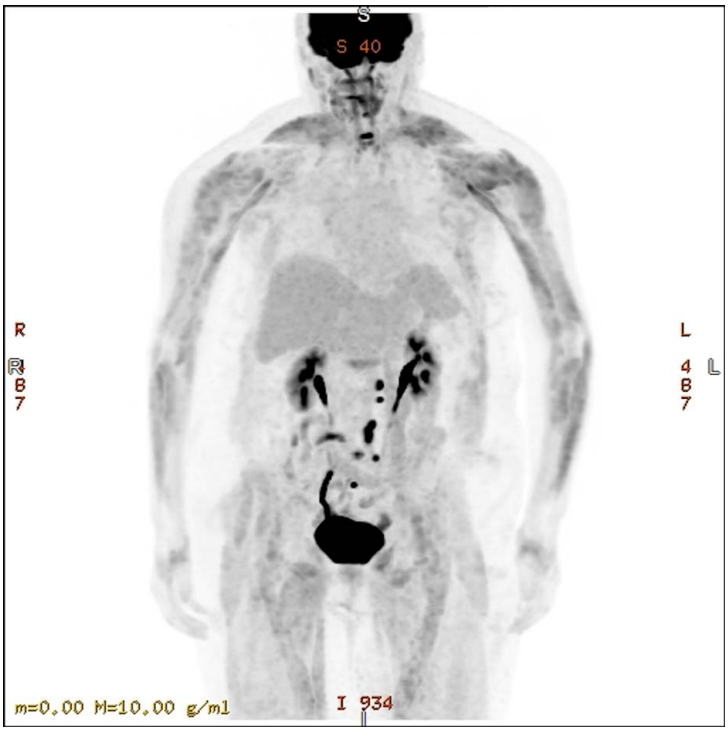

Figure 5: Maximum intensity projection image from the PET CT demonstrates diffuse radio tracer uptake in the skeletal musculature most prominent in the proximal upper limbs.

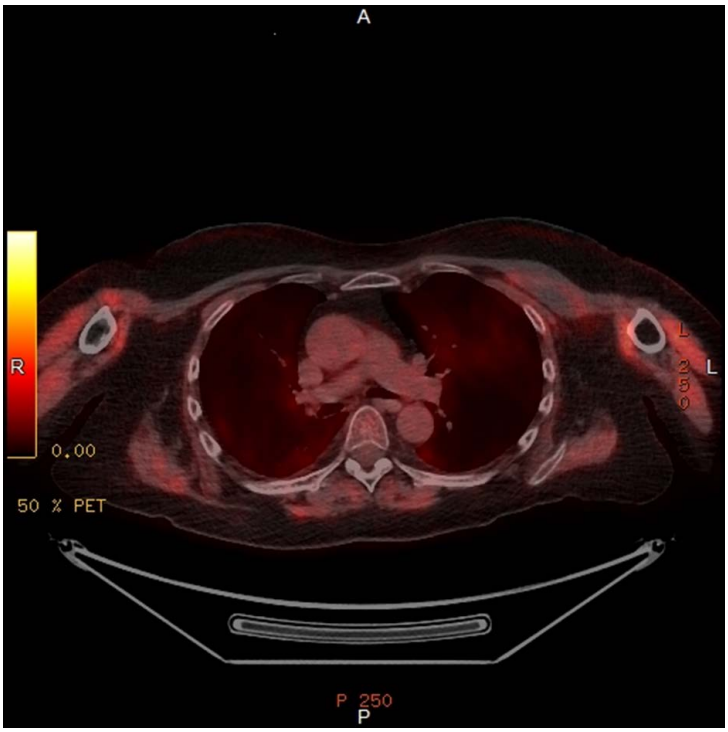

Figure 6: Fused axial image through the upper thorax from the PET CT demonstrates an area of photopaenia in the left pectoralis major muscle corresponding to the intramuscular haematoma, showing significant reduction in size compared to the previous CT. Diffuse increased tracer uptake in the skeletal musculature consistent with dermatomyositis.

KEYWORDS

dermatomyositis; spontaneous intramuscular haemorrhage; spontaneous intramuscular haematoma; TIF1-γ; diagnostic imaging

ABBREVIATIONS

TIF1-γ = Transcriptional Intermediary Factor-1 Gamma (TIF1-γ)

LMWH = Low Molecular Weight Heparin

CT-TAP = Computed Tomography Of The Thorax, Abdomen And Pelvis

FDG = 18-F Fluorodeoxyglucose

PET = Positron Emission Tomography

CT = Computed Tomography

NXP2 = Nuclear Matrix Protein 2

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