Case Report

Fistula dysfunction secondary to a subcutaneous myelomatous deposit

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Introduction

Arteriovenous fistula dysfunction is a frequent complication in, and hindrance to, patients with end stage kidney disease undergoing renal replacement therapy with intermittent haemodialysis. We present an unusual complication in a 68-year-old male patient undergoing haemodialysis therapy for end stage kidney failure caused by biopsy-proven multiple myeloma. We explore the occurrence of an extramedullary cutaneous myeloma deposit situated adjacent to an arteriovenous fistula resulting in fistula dysfunction. This is the first reported case in the English literature of a myeloma deposit encasing an arteriovenous fistula, deriving its blood supply from the fistula and thus undergoing rapid growth, eventually leading to fistula dysfunction.

Case

A 68-year-old man presented to his local doctor with a 6 week history of constitutional symptoms including lethargy, anorexia, nausea, slow healing rib injury and generalized progressive malaise. His past medical history was unremarkable except for a previously excised superficial malignant melanoma over his left scapula. Investigations on presentation revealed severe non-oliguric renal failure (serum creatinine 740 μmol/L). Dipstick urinalysis was positive for protein and subsequent urinary protein quantification revealed 1.3 g proteinuria per 24 h, the majority λ light chains. Serum electrophoresis demonstrated free λ light chains and raised β2-microglobulin. A bone marrow aspirate and trephine bone biopsy revealed frequent abnormal multinucleate plasma cells. Skeletal survey identified no significant areas of bone destruction. Renal biopsy confirmed light chain deposition disease and cast nephropathy consistent with a diagnosis of myeloma kidney.

Initial management consisted of pulsed methylprednisolone and intermittent haemodialysis through a temporary non-cuffed internal jugular venous catheter. Chemotherapy was initiated with vincristine, Adriamycin and dexamethasone. A left forearm radiocephalic (Cimino) arteriovenous fistula (AV fistula) was created in anticipation of future haemodialysis requirements. The patient achieved chemotherapy-induced remission and subsequently became dialysis independent.

Nineteen months following the initial diagnosis, a peripheral blood stem cell autograft was successfully performed. Seven months later, however, the patient had worsening renal function due to haematogenous relapse, and again required haemodialysis. Unfortunately, the dormant Cimino AV fistula that had been created the previous year was found to be stenotic at this time requiring surgical revision of the stenosis and ligation of venous tributaries. Intermittent haemodialysis was performed through a second temporary non-cuffed internal jugular venous catheter.

Within 6 months of the previous surgical revision of the AV fistula, the patient developed further fistula dysfunction. Over a period of weeks, the left arm became progressively swollen. The fistula became increasingly difficult to access and painful to use. Pressures in the venous limb of the fistula rose sharply. A fistulogram and ultrasound were performed: the fistulogram revealed numerous ill-defined vessels, stemming from the fistula and leaking into the surrounding soft tissues (Figure 1). The ultrasound demonstrated a solid vascular hyperechoic mass surrounding the AV fistula (Figure 2). Three sites of vascular that flow into the mass from the fistula were...
noted. These findings were consistent with a parasitic circulation, i.e. a mass deriving its blood supply from the AV fistula.

The patient underwent surgical exploration of the left forearm swelling. During the operation, significant amounts of myxomatous-type tissue were identified in close relation to the fistula, and a frozen section was taken. Histopathology revealed extensive plasma cell infiltration within the connective tissue, some of which were atypical, consistent with myeloma. The AV fistula was not surgically revised at this time. Following surgery the patient’s condition continued to deteriorate and he died a few months later.

Discussion

This is the first documented case of an extramedullary subcutaneous myeloma deposit causing angiogenesis from a surgically created AV fistula. It is also the first reported case describing AV fistula dysfunction secondary to an extramedullary subcutaneous myelomatous deposit.

Multiple myeloma is the second most common haematological malignancy. It occurs through malignant proliferation of plasma cells derived from a single clone. It accounts for \(~10\%\) of all haematological malignancies, with an annual incidence of \(4/100\ 000\) per year. The median age at diagnosis is 65 years. Median survival time after diagnosis is 3 years [1]. Myeloma is characterized by bone marrow plasmacytosis, lytic bony lesions and monoclonal protein (M protein) in the blood, urine or both. Clinical presentations can vary considerably, but commonly include bone pain, fractures, susceptibility to bacterial infections, anaemia, hyperviscosity syndromes and renal failure.

Arteriovenous fistulae used for haemodialysis have a number of recognized complications, which result in problems with their use for renal replacement therapy. One angiographic study demonstrated that the most common causes of fistula dysfunction resulted from significant stenosis (40%), total occlusion from thrombus (9%), aneurysm formation (7%), incorrect needle placement or anatomical abnormalities (20%) as well as local cellulitis and sepsis [2].

Extramedullary manifestations may occur in the latter stages of multiple myeloma as a reflection of increased tumour cell burden. Cutaneous involvement usually arises secondary to direct extension to the skin from an underlying bone lesion, although it may rarely be a primary metastatic lesion [3]. Previously recorded cutaneous lesions have been found most commonly in the trunk and abdomen (65% reported cases), followed by scalp, face and neck (27%), lower limbs (23%) and upper limbs (16%) [3]. Development of cutaneous lesions can occur in the early stages of multiple myeloma; however, it is usually a late development in the disease process, and generally reflects a poor prognosis, with death usually occurring within 12 months of the appearance of the lesion.

A few unusual clinical manifestations of extramedullary multiple myeloma deposits causing mass effect are described below. Vertebral extramedullary plasmacytomas causing spinal cord compression via pathological fracture or direct mass effect have been well documented [4]. Multiple myeloma presenting as a rapidly enlarging thyroid mass has been documented
Fistula dysfunction secondary to a subcutaneous myelomatous deposit

[5], as has a case of airway obstruction due to an extramedullary plasmacytoma in the neck [6]. Another case with superior vena cava syndrome due to a huge obstructing sternal plasmacytoma has been described [7]. The mass effect of extramedullary myeloma deposits in other areas of the body, however, is much rarer, and interesting clinical manifestations of this have been sparsely documented throughout the medical literature.

In the case described, systemic haematogenous spread to the site as a primary subcutaneous metastasis is plausible, because there is no radiological evidence to suggest the secondary growth of this deposit from a bone lesion. The venous limb of the fistula and surrounding subcutaneous tissues were subject to the repeated trauma of multiple needle cannulations for vascular access. The combination of repeated trauma to the site along with a high flow circulation of haemodialysis is likely to have created an environment conducive to tumour seeding. This is highly plausible, given the well documented local recurrence of tumours in biopsy sites [8], access ports [9] and wound scars [10] in the surgical oncology literature.

We employed a variety of imaging modalities to investigate the AV fistula dysfunction by the mass lesion overlying it. An ultrasound and a fistulogram demonstrated the presence of new vasculature arising from the AV fistula supplying the tumour mass. It could be argued that these vessels may represent venous collaterals, a known complication of AV fistulae from which the patient had previously suffered. However, the appearance of small distal leaks from the vessels suggests that these vessels represent new vessels, i.e. angiogenesis from the fistula supplying a subcutaneous myeloma deposit. It is our opinion that the clinically rapid enlargement of this deposit was a direct consequence of the fact that it derived its nutrient supply from such a haemodynamically significant circulation. Biopsy of the lesion demonstrated no evidence of haematoma and thus the leak demonstrated on fistulogram from the new vessels (Figure 1) was not a cause for the rapid enlargement of the deposit. As such, it is likely that had the AV fistula not been present, the speed at which the deposit enlarged would have been significantly slower.

Fistula dysfunction in this case was caused through a combination of factors that altered flow within the AV fistula. Multiple, smaller calibre, neogenic vessels arising from the venous limb of the fistula, with a far greater amount of wall relative to their cross sectional surface area, would have greatly increased the resistance to flow. The pressure increase required to maintain the same amount of flow from a large vein is proportionate to the fourth power of the radius of each tributary vessel, as described by the Poiseuille equation. This factor would no doubt have contributed to the reported sharp increase in venous access pressures of the fistula immediately preceding the discovery of the myelomatous deposit. Secondly, a rapidly expanding mass lesion surrounding and encroaching upon the fistula would eventually result in external compression of the main conduit, especially in the venous limb, with a similar effect on resistance and venous pressures as the smaller neogenic vessels described above. The distortion of the overall fistula anatomy due to a large subcutaneous mass would have increased the difficulty of cannulation.

Conclusion

This unusual presentation of extramedullary subcutaneous myeloma deposit surrounding an AV fistula demonstrates the interesting effect that an artificially augmented circulation can have, being a preferential site for tumour seeding, and causing accelerated progression of an adjacent, localized neoplastic disease process. This in turn will lead to AV fistula dysfunction with altered haemodynamics, access difficulty and ultimately extrinsic compression of the fistula due to local mass effect.

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Conflict of interest statement. None declared.

References

5. De Schrijver I, Sneets P. Thyroid enlargement due to extramedullary plasmacytoma. Organe de la Societe Royale Belge de Radiologie 2004; 87: 73–75

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