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Case Report

CASE REPORT

Arterio-venous fistula—expanding role for venous access in mastocytosis patients

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Abstract

Long-term central venous access is increasingly common as there are growing number of patients suffering from conditions that require repeated infusions for various indications. However, central venous access has its downside where there is a risk of infection and thrombosis (F Pinelli, E Cecero, D Del'Innocenti, V Selmi, R Giua, G Villa *et al*., 'Infection of totally implantable venous access devices: a review of literature,' *J Vasc Access* 2018;**19**: 230–42.). Arterio-venous (AV) fistula is traditionally used for haemodialysis, however in patients requiring repeated access or long-term central venous access it can be considered as an unconventional solution.

We report a case where 61-year-old male who has a history of systemic mastocytosis. He presents frequently to the Emergency Department with anaphylactic reaction requiring intravenous adrenaline, antihistamine and steroids. He had multiple issues with central lines as well as ports including line sepsis and thrombosis. On further discussion, an arterio-venous fistula was considered and surgically created to allow the AV fistula to be cannulated directly or under ultrasound guidance with its advantage it can be accessed rapidly in emergency setting.

INTRODUCTION

Mastocytosis is one of the eight subcategories of myeloproliferative neoplasms. It results from a clonal, neoplastic proliferation of morphologically and immunophenotypically abnormal mast cell that accumulate in one or more organ systems [1]. Clinical presentation of mastocytosis is heterogenous, ranging from skin-limited disease to a more aggressive variant with extracutaneous involvement (systemic mastocytosis) that may be associated with multiorgan dysfunction/failure and shortened survival [1]. Systemic mastocytosis can often present with pruritus/flushing, abdominal pain, cramps, diarrhoea, nausea and vomiting [2]. Patients often present following systemic mast cell degranulation resulting in anaphylaxis.

CASE STUDY

A 63-year-old man with long history of systemic mastocytosis. Due to his condition, having central venous access is crucial for rapid administration of adrenaline and antihistamines during anaphylaxis. The condition was diagnosed at the age 47 following an anaphylactic reaction that resulted in 3 months stay in intensive care unit (ICU). He initially had insertion of infusaport into his right subclavian vein for central venous access. Unfortunately, the port became colonized with *Staphylococcus aureus* within 18 months which he was admitted for bacteraemia and septic shock. He spent another 3 weeks in ICU battling severe line sepsis which resulted in multi-organ failure. He was discharged home after 4 weeks of intravenous antibio-

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tics and managed to salvage the infusaport. The infusaport was later colonized with *Escherichia coli*, the decision was made to remove the infected port. A peripheral inserted central catheter (PICC) was inserted as a replacement for his central venous access. 3 months later, a replacement infusaport was inserted. This infusaport remained thrombus and infection free for 5 years until it became infected with *Staphylococcus aureus*. After consulting with vascular surgeon, the decision to surgically created arteriovenous fistula (AVF) was made. The principle was to remove any potential form of infection while maintaining vascular access and central venous patency.

The patient began work up for AVF including ultrasound venous mapping of the upper arms, and education in managing and care for AVF. Two weeks later, he underwent left brachio/radiocephalic AVF formation under IV sedation with local anaesthesia. The patient was followed up in clinic, AVF had matured by 8 weeks and PICC line was subsequently removed. For 12 months the patient was cannulated on weekly basis for administration of antihistamines, adrenaline, corticosteroids with no complications reported. He was later commenced on Midostaurin which reduced his incidents of anaphylactic reactions.

DISCUSSION

Anaphylactic reaction from mastocytosis requires urgent access to patient's central venous system from adrenaline infusion. In emergency setting where patient is having an anaphylaxis, rapid access is essential and potentially lifesaving.

Central venous catheters with or without ports are traditionally inserted for long-term intravenous access but are prone to several complications. These include pneumothorax (1.3–1.5%), septicaemia (4–8.6%) and thrombosis (1.2%) [3]. Totally implantable venous access devices have infection rates ranging from 0.018/1000 catheter days to 0.35/1000 catheter days in adults [4–8]. Permanent catheter showed higher infection rates, catheter dysfunction and thrombosis compare to AVF [9]. Estimated cost of implantable device is ranging from Australian $2186–$2274, where AVF cost Australian $1636. This makes AVF more cost effective.

Sure, traditionally AVF is a preferred haemodialysis access where the fistula is accessed usually three times a week and as a result this access is an option that needs to be considered for any patients required long-term central venous access for repetitive administration of drugs e.g. antibiotics, etc.

CONFLICT OF INTEREST STATEMENT

None declared.

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