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Case Report

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Multifocal Pelvic and Pancreatic Masses Secondary to Granulomatous Polyangiitis Resolved with Pulsed Immunosuppression

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Malignancy often tops the differential diagnosis of multifocal masses in a middle-aged lady. In addition, certain disease conditions like vasculitis, haematomas, and developmental anomalies like hamartomas should be considered a differential apart from ruling out benign and malignant tumours. This patient presented acutely with anaemia, renal impairment and weight loss setting us in the direction of ruling out malignancy; a however high index of suspicion and negative biopsy results with positive serology helped make a prompt diagnosis of ANCA positive GPA. Institution of immunosuppressive treatment resulted in complete resolution of these masses and a positive outcome.

Case Report

A 61years old lady was admitted with a brief history of vomiting and reduced oral intake. The patient was hypotensive on admission with acute kidney injury (urea 35.7, creatinine 477) and anaemia (Hb 56). Inflammatory markers were also raised (CRP 202, WBC 16.9), and renal ultrasound showed bilateral hydronephrosis.

Two months before admission, the patient had undergone an MRI to investigate right-sided quadriceps wasting. The MRI showed a sizeable pre-sacral mass, followed by a CT abdomen in the same month, which showed the mass to be still present and highlighted a pancreatic mass suspected to demonstrate a pancreatic tumour. Other past medical history includes polymyalgia rheumatica, previous small bowel perforation with resection, post-operative DVT with oral anticoagulation with warfarin and esophagitis. Although, before admission, the patient was mobile independently and had some assistance at home to help with activities of daily living, she was a current smoker with minimal alcohol intake.

She was treated initially with fluid resuscitation, intravenous antibiotics for sepsis of unknown origin, and transfused two units of red blood cells to correct the anaemia. The patient was transferred to the critical care unit as renal function did not improve, and the patient became more systemically unwell, developing respiratory failure. The patient underwent hemofiltration on the unit, and bilateral JJ stents were inserted, as well as a right-sided nephrostomy within the first few days of admission to rectify the bilateral hydronephrosis.

Renal screen results three days after admission revealed the ANCA positive (cANCA positive, ANCA negative, GBM negative. Subsequently, a diagnosis of vasculitis was made and commenced on IV steroids and plasma exchange. The patient required invasive ventilation for the initial three days and subsequently had intermittent CPAP.

Once stabilised, the patient was started on intravenous cyclophosphamide chemotherapy every three weeks , stepped down to a medical ward and continued on a tapering dose of oral prednisolone. Unfortunately, chemotherapy was complicated by urosepsis and a bout of hospital- acquired pneumonia. On admission, other stigmata of vasculitis, like Splinter haemorrhages and cutaneous and nail fold infarcts, were all present at the time of diagnosis of vasculitis; all resolved with cyclophosphamide and prednisolone therapy. Repeat MRI scan four months following the pre-admission MRI pelvis and CT abdomen showed a dramatic reduction in the pre-sacral mass and no evidence of the pancreatic lesion seen on the CT scan four months earlier.

The pre-sacral mass was biopsied twice. The pre-admission biopsy showed chronically inflamed connective tissue with necrotic areas, and suspicion of tuberculosis was raised; however multiple sputum samples analysed for acid-fast bacilli were reported as unfavourable and Zeil-Neilson staining of the tissue was also negative. The mass was biopsied again during the admission, and a smooth muscle neoplasm was suspected. Blood results did reveal a mildly raised CA125 76.7 (1-35 kU/L), CA19-9 was 37 (0.1-31 kU/L), and the patient was discussed in the gynaecology and sarcoma MDTs and discharged from both MDTs respectively.

As the MRI scan showed significant improvement in the size of the pre-sacral mass and discussion of relevant results at both the gynaecology and sarcoma Discussion at the MDT's concurred, no evidence of malignancy, backed up with a tissue diagnosis of

inflamed connective tissue, it would appear that this patient had a pelvic mass secondary to Granulomatous polyangiitis which improved with treatment of the underlying condition. ANCA screen -positive, PANCA - negative, CANCA - positive PR3= 44.4u/ml, GBM- Negative Following initiation of immunosuppressive treatment for ANCA -positive vasculitis, the patient's renal function did improve but not to baseline before discharge (Ur 20, Cr 200), and the patient remains under regular review by the renal physicians. In addition, the right nephrostomy was removed, and a repeat renal ultrasound showed the resolution of the bilateral hydronephrosis and average-sized kidneys. Therefore, the urology team planned to remove the JJ stents as an outpatient procedure.

The patient's mobility improved to near pre-admission state, so much so that the initial plan for discharge to a rehabilitation centre was shelved so that the patient could return directly to her own home. The patient was switched from warfarin to LMWH due to inconsistent INR results (which had been a problem prior to admission) and was discharged on LMWH, which she will self-administer. An IVC filter was also inserted during the patients' stay in critical care. The rheumatologists, nephrologist and respiratory team continued to follow up on this patient for six months initially and annually after that. The patient received ten cycles of pulsed IV cyclophosphamide, after which the patient was swapped to azathioprine with a reducing prednisolone regime.

Although renal replacement therapy was discussed, the patient never required dialysis subsequently. Two years later patient developed an osteoporotic lesion affecting the D7 D8 D12 vertebra, and the ANCA became negative after four years. The patient's renal function was maintained with azathioprine 100mg once daily and steroids wholly weaned. The various specialities continue to monitor vasculitis.

Discussion

Granulomatosis with polyangiitis (GPA), previously Wegener's Granulomatosis, is an anti-neutrophilic cytoplasmic autoantibody (ANCA) associated small vessel vasculitis with protean presentations. They can present with skin, pulmonary, renal, or nasopharyngeal involvement. The prevalence is 135 per million, and the 1-year mortality is about 14% [1]. Therefore, a high index of suspicion is imperative to make a prompt diagnosis, and delays in diagnosis can cause significant morbidity.

Our patient presented with nonspecific symptoms of anaemia renal failure, and imaging revealed masses in the pelvis and Pancreas. The initial differential at that time was metastatic malignancy with Pancreas as primary and pelvic metastasis or vice versa. The imaging was discussed both at the Gynae MDT and Upper GI MDT. However, pelvic mass biopsy on two separate occasions confirmed vasculitis, and at the same time, ANCA returned positive. The patient was commenced on immunosuppressive therapy with ten cycles of IV pulsed cyclophosphamide.

There are several examples of GPA mimicking malignant disease in the form of ubiquitous masses. Although commonly presenting as pulmonary or renal masses, pancreatic masses secondary to Wegener's have been well described with or without pancreatitis [2-10]. Reports of pelvic masses secondary to Granulomatous polyangiitis are rare and have also been described as case reports [11-16]. Typical treatment would have been immunosuppressive, although hysterectomy has been described when a clear indication of uterine involvement and pancreatic resection has also been considered in some cases [15.17].

This is the first case presenting as multifocal masses simultaneously in retroperitoneal organs like the Pancreas and pre-sacral mass in literature. Previous reports of "tumour-like" lesions secondary to vasculitis have been reported, most commonly affecting the breast, central nervous system and ovarian tissue.

In the past, patients have often not been diagnosed with vasculitis until after an operation to remove a possible tumour. In contrast, this could have been avoided if clinically suspected and appropriate investigations and treatment had been given. Our patient had complete resolution of the pelvic and pancreatic mass following immunosuppressive therapy with no recurrence.

Our case is unique in that concomitant multifocal masses can be a presenting feature of GPA. We want to highlight that such masses can be resolved entirely with immunosuppressive therapy. Although vasculitis has been reported as a paraneoplastic phenomenon, we have not found any evidence of malignancy in various investigations in this patient and during the seven years follow-ups of the patient [18-20].

This case report highlights an example of a patient with a pancreatic and pre -sacral mass initially thought to be due to a malignant disease process. However, both showed dramatic improvement with immunosuppressive treatment, as confirmed by repeat MRI abdomen/ pelvis.

This case report aims to highlight the differential diagnosis of vasculitis in patients with abdominal -pelvic masses so that appropriate investigations are done early, and management starts once the correct diagnosis has been made.

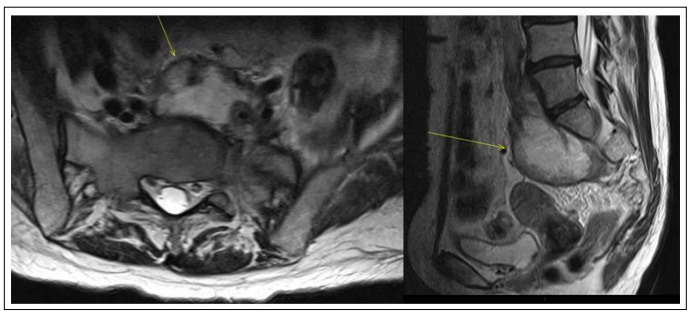
Learning Points

- Granulomatous polyangiitis can mimic malignant disease. Vasculitis can also occur secondary to malignancy.
- Appropriate treatment of Granulomatous polyangiitis can provide an improvement in the size of tumour-like lesions.
- It is crucial to differentiate tumour-like lesions secondary to Granulomatous polyangiitis, and vasculitis secondary to malignancy as the treatment is different.

Images

1. Dated 28/04/2013

Axial and sagittal T2 weighted MR images of the pelvis reveal a well-circumscribed hyper-intense pre-sacral lesion (9.5 cm crania caudal X 4.6 cm Antero posterior X 6.3 cm Trans), encasing the aortic bifurcation and proximal iliac vessels.



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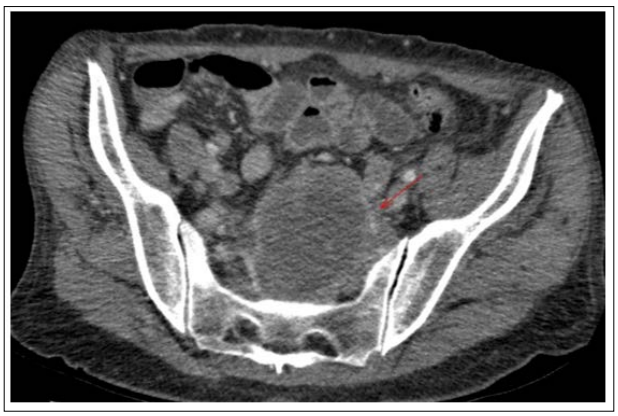
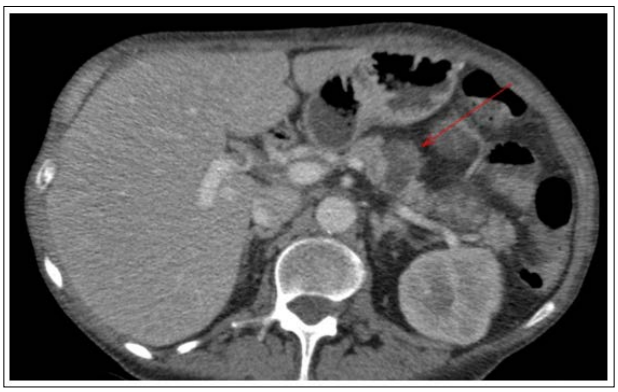
2. Dated 9/5/2013

Axial CT scan at the pelvis level reveals a rim enhancing, predominantly necrotic pre-sacral lesion), encasing the aortic bifurcation and proximal iliac vessels.



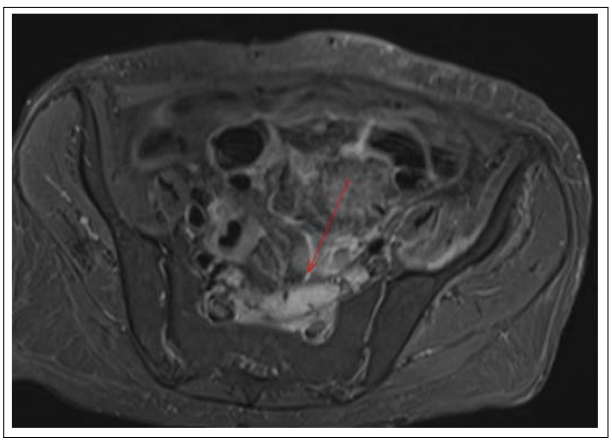
3. Ct dated 9/5/2013

Axial CT of the abdomen reveals a well-defined, predominantly cystic/necrotic lesion in the pancreatic body (measures 3.0 X2.2 cm) with the eccentric solid component.



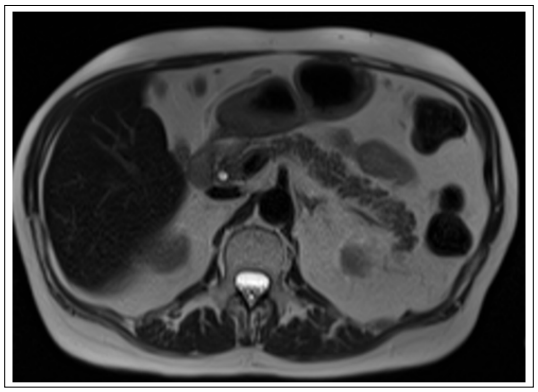
4. MRMR dated 28/1/2014

Axial T1 fat Sat image of the pelvis revealed an interval decrease in size of the pre-sacral mass lesion and the presence of residual oedema.



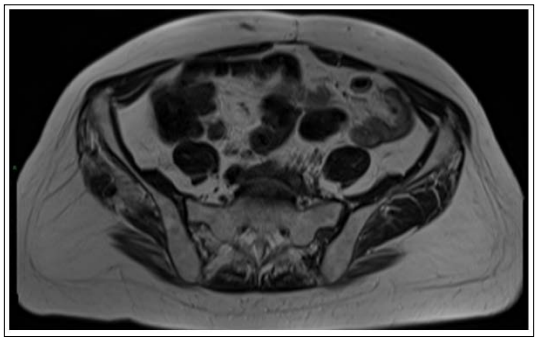
5. MRMR dated 14/12/2014

Axial T2 Weighted MR image of the abdomen reveals complete interval resolution of pancreatic body mass.



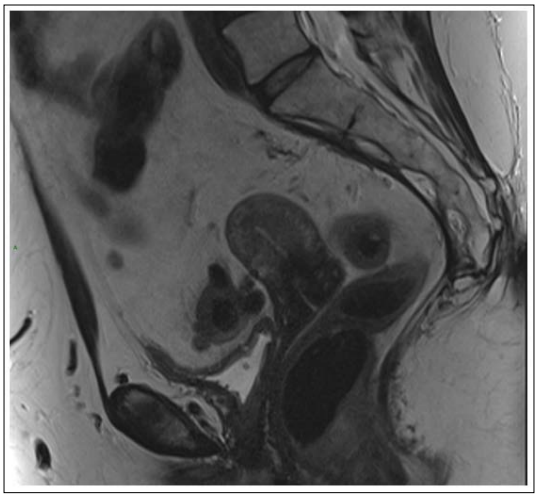
6. MRMR dated 16/02/2015

Axial T2W MR Image of the pelvis reveals complete interval resolution of pre-sacral mass.



7. MRMR dated 12/07/2016

A Sagittal T2W MR image of the pelvis reveals complete resolution of pre-sacral mass and no recurrence.



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