

which was why the studies needed to rule out the disease were performed at birth. The negative result and early clinical symptoms led to the diagnosis. Knowledge of this association may facilitate an early diagnosis of the disease, thus establishing an appropriate treatment.

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### Acute renal failure as a presentation of an aortocaval fistula associated with abdominal aortic aneurism

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#### To the Editor,

Aortocaval fistula (ACF) is an uncommon complication of abdominal aortic aneurysms (AAA)

that requires urgent action. However, they are diagnosed in only half of the cases, which increases postoperative mortality. Indicative of this condition are a continuous abdominal murmur and the presence of high-output heart failure (HF) or regional venous hypertension. Acute renal failure (ARF) may be the presentation of the ACF, as in the case described.

This is the case of a 71-year-old patient with a history of dyslipidaemia, hypertension, truncal obesity and coronary artery bypass. The patient was transferred to our hospital from the emergency department of another hospital due to anuric ARF unresponsive to dopamine or furosemide. The patient complained of self-limited sweating episodes, and had malaise, agitation, disorientation, and tachypnoea, blood pressure at 108/62mm Hg, a pulse of 91 beats/min and central venous pressure (CVP) of 30cm H<sub>2</sub>O. An examination revealed bibasilar crackles and painful, enlarged liver, without a pulsatile abdominal mass or ankle oedema. Laboratory tests revealed the following data: urea 108mg/dl, creatinine 4.05mg/dl, pH 7.19 and bicarbonate 10.2mmol/l, with blood count and coagulation normal.

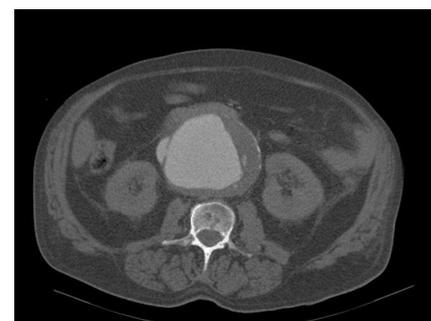
The chest x-ray showed a vascular redistribution pattern with small bilateral pleural effusion. A renal Doppler ultrasound showed normal-sized kidneys without signs of obstruction, a dilated right renal vein, and a large AAA. The CT scan with contrast showed an infrarenal, 10cm AAA with atherosclerosis and thrombosis, without retroperitoneal haematoma, as well as the flow of contrast to the vena cava and right renal vein during the arterial phase and the ACF (Figure 1).

With the diagnosis of ARF and CHF secondary to ACF, haemodialysis and surgery were performed, with control of the aneurysm neck, which

required the ligation of left renal vein, the opening the aneurysmal sac (with the loss of 2 litres of blood), the closure of the ACF and placement of aorto-aortic bypass. The immediate postoperative diuresis was 70ml/h, but the patient developed refractory multiorgan failure and died three days after surgery.

The rupture of an aortic aneurysm and into other organs is uncommon. The inferior vena cava is the most common, followed by the iliac veins, the left renal vein and intestine.<sup>1,2-4</sup> Over 80% of ACF cases are due to the rupture of an atherosclerotic aortic aneurysm (average size 11cm),<sup>2-5</sup> with other causes being penetrating abdominal trauma; iatrogenia, mainly lumbar disc surgery or catheterisation; and occasionally others.<sup>1,3</sup>

Between 31% and 76% of the ACFs are detected during surgery after evacuating the clot from the aneurysm sac, which causes massive



**Figure 1.** CT scan with contrast. Abdominal aorta aneurism and aortocaval fistula

bleeding and/or a paradoxical pulmonary embolism.<sup>3,5-8</sup> They are difficult to identify because 20%-70% are associated with abdominal or retroperitoneal rupture where the characteristic signs (pulsatile abdominal mass, pain, signs of bleeding, abdominal murmur) are absent or may be easily misinterpreted. An aneurysmal thrombus in the fistula, arterial hypotension or compression of the inferior vena cava may mask the clinical signs of an ACF.<sup>1,3-5,7-9</sup>

The appearance of an ACF involves that aortic arterial blood is flowing to the vena cava, and therefore, arterial resistance is reduced, the renin-angiotensin-aldosterone and sympathetic systems are activated, and a hyperdynamic circulatory state is seen in 30%-50% of cases. If the ACF is large or the heart is unable to increase cardiac output, heart failure and refractory shock appear.<sup>1,6,8</sup> In addition, ACF causes regional venous hypertension due to volume overload that manifests as oedema in the extremities or scrotum, haematuria, priapism, bleeding or dilated abdominal wall veins. The CVP may be low, normal or high, even very high, as in our patient, which should lead to suspecting the possible existence of ACF.<sup>3,7</sup>

ACF-associated renal manifestations are macroscopic or microscopic haematuria due to renal and/or bladder venous congestion (which is suggestive of ACF in AAA patients)<sup>3-5,7,10</sup> and ARF, present in 7%-77% of cases and related to functional haemodynamic changes, renal venous hypertension and perfusion deficit.<sup>3,5,6</sup> The immediate recovery of diuresis after surgical closure of the ACF and even the absence of renal lesions in the renal biopsy have been reported on many occasions.<sup>11,12</sup>

The diagnostic method of choice for ACF is CT, which shows early enhancement of the vena cava,

isodense with the aneurysm and even the arteriovenous communication. Other findings include the inferior vena cava dilated or compressed by the aneurysm, retrograde opacification of the renal veins during the arterial phase, poor renal enhancement or an increase in size, and pelvic and retroperitoneal congestion.<sup>13,14</sup>

Post-operative-associated ACF mortality is 16%-72%, and is mainly related to incorrect diagnosis, hypotension, shock with anuria or ruptured aortic aneurysm.<sup>2-4,7-9</sup>

Specific treatment for ACF is transaortic closure and placement of a prosthetic graft, which improves heart and renal failure.<sup>3,5,7</sup> A proposed alternative treatment to surgery is aortic stenting in the rupture of aortic aneurysms, with good results at centres with extensive experience.<sup>15</sup>

In summary, ACF is a rare complication of an aortic aneurysm, which requires high diagnostic suspicion because of its high mortality. The ACF may present with haematuria and/or oligoanuric acute renal failure requiring dialysis. Decreased renal perfusion and venous hypertension are the major pathophysiological mechanisms involved in the renal failure of these patients.

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## Cinacalcet in the treatment of hypercalcaemia and control of hyperparathyroidism due to ectopic parathyroid glands

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### To the Editor,

Cinacalcet is an allosteric modulator that stimulates the calcium-sensing receptor on the surface of parathyroid cells when activated by the extracellular calcium ion and inhibits parathyroid hormone (PTH) release. At the same time it decreases levels of serum phosphorus and calcium.<sup>1</sup> This calcimimetic agent has established itself as a mainstay in the treatment of secondary hyperparathyroidism, and is also useful in persistent hyperparathyroidism after renal transplantation, in primary hyperparathyroidism and hyperparathyroidism due to parathyroid carcinoma.<sup>1-3</sup>

We report our experience in treatment with cinacalcet in a case of hyperparathyroidism and hypercalcaemia due to an ectopic mediastinal parathyroid gland in a patient with chronic renal failure (CRF). We found no cases of hyperparathyroidism due to ectopic parathyroid glands treated with cinacalcet in the current literature. However, there were two cases of haemodialysis patients successfully treated with this drug<sup>4,5</sup> who were affected by parathyromatosis (hyperfunctioning parathyroid tissue distributed throughout the neck, which is a rare cause of recurrent

hyperparathyroidism after total or subtotal parathyroidectomy).

We describe the case of an 80-year-old woman referred to our department two years before due to stage 4 CRF of unknown origin. She had a history of obesity, poorly controlled hypertension (HT), gout, post-thyroidectomy hypothyroidism, generalised osteoarthritis and a gastric ulcer. The analytical tests revealed the following data: creatinine 3.8mg/dl, serum calcium 10.7mg/dl, phosphorus 4.5mg/dl and iPTH 725ng/ml. Tc-Sestamibi parathyroid scintigraphy showed ectopic hyperfunctioning parathyroid tissue in the mediastinum (Figure 1). Cinacalcet treatment was started (under a compassionate use programme) at a dose of 30µg every other day. The frequency of administration had to be increased and, subsequently, resolution of the hypercalcaemia was observed and the treatment was withdrawn due to gastrointestinal intolerance. However, hypercalcaemia of 11.2mg/dl returned and creatinine had increased to 4.13mg/dl with clinical deterioration (fatigue, walking difficulty, and headache) treatment was resumed at 30µg every 48 hours. With good tolerance and evident clinical improvement, normalisation of calcaemia was achieved after a month.

The cinacalcet dose was progressively increased to optimise iPTH figures and an arteriovenous fistula was subsequently performed to prepare the patient for renal replacement therapy, being the estimated glomerular filtration rate 13ml/min. One year after starting the calcimimetic treatment at a dose of 90µg and 60µg on alternate days, hypocalcaemia of 7.8mg/dl was found, therefore, oral calcitriol was added to the therapy. Two years after diagnosis, the patient had a good quality of life with creatinine levels of 4.17mg/dl, serum calcium of 8.4mg/dl and iPTH of 404ng/ml. The cinacalcet dose was 90µg and 60µg on alternate days and calcitriol 0.25µg per day. Concomitantly, a proper control of blood pressure was reached.

When a patient with advanced CKF presents hypercalcaemia, in addition to secondary hyperparathyroidism, autonomous hyperparathyroidism should also be considered, for example, hyperparathyroidism caused by an ectopic parathyroid adenoma. Hence the usefulness of Tc-Sestamibi scintigraphy in differential diagnosis.<sup>6</sup> Cinacalcet may be useful in the treatment of hypercalcaemia and control of hyperparathyroidism due to hyperfunctioning ectopic parathyroid gland, thereby avoiding surgery in

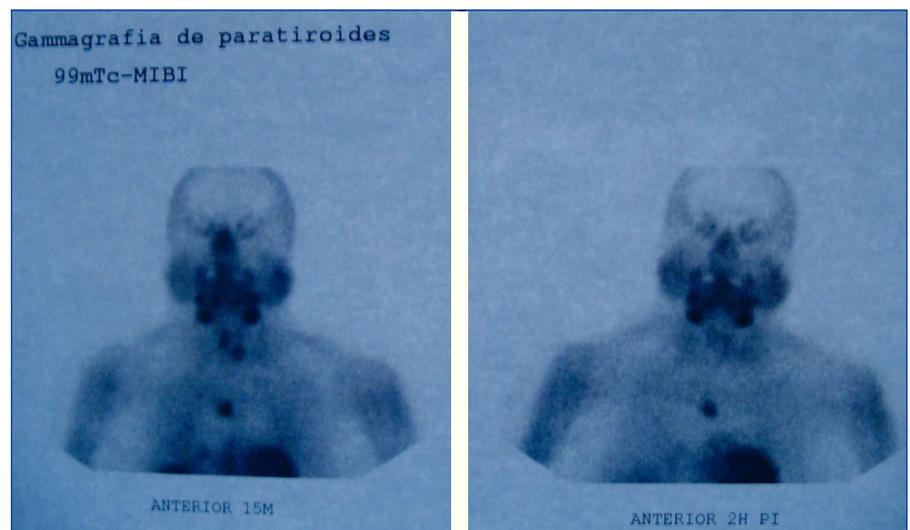


Figure 1. Tc-sestamibi parathyroid scintigraphy.