CASE REPORT

Adrenocortical carcinoma presenting as bilateral pitting leg oedema

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SUMMARY

We report a case of a 54-year-old woman presented with bilateral pitting leg oedema. Initial workup for common aetiologies was unrevealing and diuretic therapy was ineffective. A CT scan of the abdomen demonstrated left adrenal mass with direct invasion of the adrenal vein and inferior vena cava with direct extension to the right atrium. Adrenocortical carcinoma was confirmed in biopsy and the patient was operated within several days. Fifteen months postoperation, the patient is doing well with good performance status and still in oncological treatment and follow-up. When the common causes of bilateral oedema have been ruled out, no delay should be experienced seeking abdominal mass with vascular invasion potential, as early diagnosis and treatment may be lifesaving.

BACKGROUND

Adrenocortical carcinoma (ACC) is a rare disease with intravascular invasion potential. As early diagnosis and treatment can be lifesaving, it is paramount to identify symptoms that may represent a clue for the diagnosis.

CASE PRESENTATION

A 54-year-old woman was admitted for progressive bilateral pitting leg oedema. Her medical history was notable for diabetes mellitus, hypertension, hyperlipidaemia and rheumatoid arthritis. Medical treatment included ramipril, metformin, simvastatin, hydroxychloroquine, methotrexate and folic acid.

Three weeks before admission, she referred to her family physician with a new onset bilateral pitting leg oedema. Medical history and physical examination were not suggestive of heart failure, hepatic cirrhosis or nephrotic syndrome and she denied taking calcium channel blockers.

INVESTIGATIONS

Laboratory workup revealed normal kidney, thyroid and liver function tests, normal serum albumin level and coagulation profile, negative urine stick for protein and 24 h urine collection for protein were within normal limits. Chest X-ray and trans-thoracic echocardiography showed no pathology.

CT scan of the abdomen and pelvis was performed and demonstrated left adrenal mass with direct invasion of the adrenal vein ([figure 1](#)), with extension of the mass to the right atrium through the inferior vena cava (IVC; [figure 2](#)). Twenty-four-hour urine catecholamines and metanephrines were within normal limits.

Figure 1 Chest CT scan post-intravenous contrast media: left adrenal mass with direct invasion of the adrenal vein is shown. The vein crosses anterior to the aorta from left to right. The upper pole of the right kidney is also shown.

Transoesophageal echocardiography showed a vascular mass in the right atrium, suggestive of a tumour, 30 mm in diameter protruding 50 mm into the right atrium from the IVC ([figure 3](#)). Severe dilation of the inferior and superior vena cava was noted.

CT-guided fine-needle biopsy of the left adrenal was performed confirming the diagnosis of invasive ACC.

TREATMENT

Five days later the patient was operated by a multi-disciplinary team with the use of cardiopulmonary bypass. The tumour was extracted from the heart and the vena cava and the left adrenal gland was removed. The patient was referred to an expert endo-oncologist and began mitotane treatment.

OUTCOME AND FOLLOW-UP

Fifteen months postoperation, the patient is doing well with good performance status and still in oncological treatment and follow-up.

DISCUSSION

ACC is a rare malignancy with an estimated incidence of 0.5–2 new cases per million per year.¹ Patients may present either with a hormonally active or inactive tumour. In non-functioning tumours, which are seen in 30–60% of the cases, the tumour size often exceeds 10 cm at the time of diagnosis causing early satiety, weight loss or abdominal pain.^{2 3} This may be associated with tumour thrombus in the IVC with direct extension to the right atrium.^{4 5} Chiche *et al* have reported a 15-patient series of ACC extending to the IVC, and suggested that surgical treatment can be effective for management of ACC with extension into the

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Figure 2 Chest CT scan post-intravenous contrast media: the right ventricle is filled with opacified blood. Within the atrium there is a hypodense mass delineated by the opacified blood. Small amount of pleural effusion is shown on the right hemithorax.

IVC. They also stated that long-term prognosis is poor owing to delay in diagnosis, frequent-associated metastatic disease and lack of effective adjuvant treatment.⁶ Hence, early identification of intracaval thrombus is paramount to the successful treatment of ACC, since complete local excision is the best therapy for advanced local disease.⁷

Oedema is defined as a palpable swelling caused by an increase in interstitial fluid volume. The most likely cause of leg oedema in patients over 50 years of age is venous insufficiency.⁸ Common causes of bilateral leg oedema include venous insufficiency, pulmonary hypertension, heart failure, lymphoedema, drugs, obesity and pregnancy.⁸ Less common aetiologies include nephrotic syndrome, liver disease, pelvic tumour or lymphoma causing external pressure and rare causes include protein-losing enteropathy, malnutrition, malabsorption, restrictive pericarditis and cardiomyopathy, Beri Beri and myxoedema.⁸

Initial workup for bilateral leg oedema consists of a well-detailed medical history and physical examination, looking for symptoms and signs of specific causes such as congestive heart failure, nephrotic syndrome and cirrhosis. Additional information can be obtained from laboratory parameters such as liver and kidney function tests, serum albumin level, thyroid function

tests, coagulation profile and 24 h urine collection for protein. If the previous workup returns unrevealing, echocardiography should be performed to rule out congestive heart failure and pulmonary hypertension. Abdominal imaging should be the next diagnostic step looking for pelvic tumour or lymphoma causing external pressure.⁸

ACC is a very rare cause of isolated bilateral leg pitting oedema, as the non-functioning tumours usually grow locally and produce symptoms related to mass effect.

We report a unique case of advanced ACC of the left adrenal gland that extended to the IVC and the right atrium through direct vascular invasion of left adrenal vein and presented as an isolated bilateral leg pitting oedema. It is valuable to note that no local extravascular extension or external compression was demonstrated in imaging studies or intraoperatively.

The patient was operated within a short period of time owing to the rapid progression of the disease as reflected by the discrepancy between the findings in the transthoracic and transoesophageal echocardiographic studies performed 5 days apart.

As ACC can be fatal when it invades the heart, early diagnosis and treatment is crucial. Its death may be related to rapid disease progression via the vascular route, as the case of our patient. Hence, when ACC is suspected, urine collection for catecholamines and metanephrines should be performed as soon as possible to rule out pheochromocytoma prior to biopsy or surgery and the patient should be operated without delay.

In conclusion, bilateral leg oedema can be the presenting symptom of highly malignant disease with a high death potential owing to cardiac involvement. When the common causes of bilateral oedema have been ruled out, no delay should be experienced seeking abdominal mass with vascular invasion potential, as early diagnosis and treatment may be lifesaving.

Learning points

- ▶ Bilateral pitting oedema may be the only presenting symptom of adrenocortical carcinoma (ACC).
- ▶ ACC can be rapidly progressive because of its unique vascular invasion.
- ▶ Pheochromocytoma should be ruled out before adrenal biopsy or surgery.
- ▶ Complete local excision is the best therapy for advanced local ACC.
- ▶ Early diagnosis and treatment may be lifesaving.

Figure 3 Four-chamber transoesophageal echocardiography view: vascular mass in the right atrium, 30 mm in diameter protruding 50 mm into the right atrium from the inferior vena cava.

Contributors MEN was involved in the conception, defining the aims, writing and revising the final manuscript. AI was involved in choosing and interpreting the imaging studies and in reviewing the manuscript. EB was involved in the conception, defining the aims and revising the final manuscript.

Competing interests None.

Patient consent None.

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