Acromegaly with Dilated Cardiomyopathy

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Abstract
Acromegaly is a rare condition with an approximate incidence of 3-4 new cases per million per year and occurs as a result of excess secretion of growth hormone (GH). It is associated with several cardiovascular manifestations of which dilated cardiomyopathy with systolic and diastolic dysfunction is relatively rare but associated with increased mortality. There are very few documented cases of acromegaly with dilated cardiomyopathy in Indian female patients in literature, thus justifying the uniqueness of our case.

We report a case of acromegaly in a 41 year old female patient who remained undiagnosed for 6 years and presented to us for the first time with symptoms of heart failure. The symptoms were attributed to dilated cardiomyopathy resulting from a prolonged and excessive exposure of the myocardium to a GH secreting pituitary tumor. Subsequently she underwent trans-sphenoidal resection of the pituitary macroadenoma.

Introduction
Acromegaly is a syndrome caused by GH-hyper-secretion as a result of somatotroph adenoma but can rarely due to extra-pituitary ectopic lesions. Because of the extreme rarity of acromegaly in Indian female patients and in general, accompanied by a slow progression of the disease activity, acromegaly can be missed in early stages and for years (as in our patient) until changes in external features, especially of the face, become noticeable. In patients presenting with cardiac failure symptoms, it is important to recognize this condition as the underlying etiology as it is potentially reversible especially if the patient is young and the condition is diagnosed and treated at an early stage.

Here, we report a case of acromegaly presenting as dilated cardiomyopathy in a 41 year old female patient.

Case Report
A 41 year old female patient was admitted to the medical ward with complaints of breathlessness on exertion since the past 10 days accompanied by anorexia, constipation and abdominal pain. She also had history of swelling on the joints of both the hands and feet and progressive enlargement of the face since the last 6 yrs, intermittent headache for the past 5 yrs, blurring of vision and postprandial vomiting since the last 1 month. There was no past history of ischemic heart disease, diabetes mellitus or hypertension in the patient. Family history was not significant. She was being treated in her hometown with hydroxychloroquine and methotrexate for suspected rheumatoid arthritis for the past 6 years. Her blood pressure was 110/70 mm Hg, pulse was 94/min and RR was 18/min. General examination revealed edema of the lower extremities, coarse facial features and swelling of joints of both hands (Figure 1) and feet. Tachycardia, S5 gallop and bibasal crepitations were noted. EKG revealed only sinus tachycardia.

2D echo revealed a picture of dilated cardiomyopathy: [Left ventricle-dilated in size with global hypokinesia and LVEF of 15%. Left atrium-dilated in size. Right atrium and ventricle-sizes were upper limit of normal with depressed function. Pericardium-mild pericardial effusion noted over LV posterior wall measuring 6 mm].

The patient was started on ACE inhibitors, diuretics, beta- blockers and statins for the management of congestive cardiac failure. There was slight improvement in the cardiac symptoms but headache, blurring of vision and swelling of the joints persisted.

Other laboratory tests done as part of the routine workup included free T3 and TSH in the normal range with free T4: 0.71(normal range 0.8-2), cortisol level: 35.70 ng/ml (Normal value 50-150ng/ml), blood sugar fasting-116.7 mg/dl, PPBS-176 mg/dl, HbA1c-7.22 gm%, negative RA, HLA B27, anti-CCP and normal uric acid levels thus excluding common causes of joint swelling. Because of the presence of coarse facial features, there was a high suspicion of acromegaly in this patient. The routine workup for the evaluation of acromegaly was done, which revealed:

- GH level of 63 (1 hr after 100ml glucose) (reference value 0-3 ng/
ml)
• IGF-1 level: 358 ng/ml (Normal value: 68-225 ng/ml)
• Pituitary hormone profiling showed an increase in prolactin level: 64.36 (Normal value 1.2-29.3 ng/ml) with others in the normal range.

Thus, a diagnosis of acromegalic dilated cardiomyopathy was made and the patient was referred for MRI brain for confirmatory testing. MRI of the pituitary gland with contrast revealed evidence of a large oblong sellar and suprasellar mass lesion giving ‘figure of eight’ appearance thus confirming pituitary macroadenoma. The size was approximately 2.1×3.1×4.0 cm (Figure 2).

Although surgery is considered as a definitive treatment for acromegaly, our patient was given a trial of medical therapy first in view of cardiac problems. She was started on medical treatment with somatostatin, thyroid hormone and prednisolone and was discharged home along with the cardiac medications. She continued to have symptoms even after 2 months of treatment. Hence she underwent transsphenoidal resection of the pituitary tumor. Post-surgery, she showed a significant improvement in headache and field of vision (Figure 3) but the cardiomyopathy did not show any significant improvement on a repeat 2D-Echo done recently.

**Discussion**

Acromegaly is associated with several cardiovascular manifestations like hyperdynamic state, hypertension, valvular dysfunction, systolic and diastolic dysfunction, arrhythmias, aortic root dilation and heart failure. Sustained increased levels of GH and IGF-1 (insulin-like growth factor) lead to derangement of cardiomyocytes, thus causing structural changes in cardiac muscles. Early phase is characterized by a hyperdynamic state. If this is not addressed in time, there is biventricular hypertrophy followed by systolic and diastolic dysfunction. Histopathologic findings in the myocardium due to longstanding acromegalic dilated cardiomyopathy are that of myofibrillosis, interfascicular fibrosis and areas of lymphocytic infiltration gradually impairing the whole architecture. 1,2

Our patient had a history of longstanding acromegaly (about 6 years) resulting in dilated cardiomyopathy with severe systolic and diastolic dysfunction. Very advanced cardiomyopathy with severe systolic dysfunction (as noted in our patient has low likelihood of reversal. The reversibility is higher in young population and decreases as age advances. Also, the time required for these manifestations to resolve is about 1 year. 3 In order to prevent the development of irreversible acromegalic cardiomyopathy, early diagnosis is important. In situations where even tumor resection and anti-failure medications don’t help, orthotopic heart transplant is the only option available. 3

Since it has been only about 3 months since the surgical removal of the pituitary macroadenoma, it is a little early to comment about the reversibility of dilated cardiomyopathy in our patient. However, according to the literature and the follow up Echo done recently showing almost no improvement in the ejection fraction, there is a very high possibility that the cardiomyopathy in our patient might persist.

In conclusion, the highlights of this unique case are:

- Acromegaly with dilated cardiomyopathy is extremely rare in Indian female patients and can be easily missed.
- The patient may remain undiagnosed for years and can directly present for the first time with symptoms of cardiac failure.
- Early diagnosis and prompt treatment is required to increase the likelihood of reversibility in a patient having dilated cardiomyopathy due to acromegaly.

**References**