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CASE REPORT

Alcohol septal ablation in obstructive acromegalic hypertrophic cardiomyopathy – a first case report

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Abstract Acromegaly is a rare disease, mostly caused by a growth hormone (GH)-secreting benign pituitary tumor, with an increased production of GH and insulin-like growth factor 1 (IGF-1). Cardiovascular complications are common and are associated with cardiomyocyte apoptosis and concentric cardiac hypertrophy. Suppression of GH and IGF-1 appears to improve cardiac function only in the short term, with little or no decrease in left ventricular mass or improvement in cardiac function after prolonged treatment. Alcohol septal ablation (ASA) has emerged as a minimally invasive alternative to septal myectomy, with significant improvement in symptoms, gradients and left ventricular remodeling. In this report, we describe the case of a 73-year-old woman with acromegaly due to a pituitary adenoma diagnosed and treated surgically at the age of 38 but with recurrence and reoperation at the age of 50. She was referred to our cardiology department due to a three-month history of progressively worsening exercise-induced dyspnea and orthopnea under optimal medical therapy. Echocardiography and magnetic resonance imaging revealed severe basal hypertrophy of the interventricular septum (19 mm), dynamic left ventricular outflow tract obstruction with a gradient of 70 mmHg at rest and 120 mmHg with Valsalva maneuver, and systolic anterior movement (SAM). Genetic testing excluded the most frequent forms of familial hypertrophic cardiomyopathy. ASA was performed with injection of 2 cc of alcohol in the first septal branch of the left coronary artery, without complications. At one-year reassessment, significant clinical and echocardiographic improvement was noted, with disappearance of SAM. To our knowledge, there have been no previously reported cases of ASA in hypertrophic cardiomyopathy due to acromegaly. We report a case of successful ASA in acromegalic cardiomyopathy.

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PALAVRAS-CHAVE
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Ablação septal alcoólica na cardiomiopatia hipertrófica obstrutiva acromegálica – primeiro caso descrito

Resumo A acromegalia é uma doença rara, causada essencialmente por um tumor benigno da glândula pituitária secretor de hormona de crescimento (HC), provocando uma produção excessiva de HC e de fator de crescimento semelhante à insulina tipo 1 (IGF-1). As complicações cardiovasculares são comuns e têm sido associadas à apoptose de cardiomiócitos e à hipertrofia cardíaca concêntrica. A supressão de HC e de IGF-1 parece melhorar a função cardíaca apenas a curto prazo, com pouco ou nenhum efeito na redução da massa ventricular esquerda ou na melhoria da função cardíaca a longo prazo. A ablação septal alcoólica (ASA) surgiu como uma alternativa minimamente invasiva à miectomia septal com melhoria significativa a nível de sintomas, gradientes e remodelagem ventricular esquerda. Neste caso clínico, descrevemos as imagens de uma mulher de 73 anos de idade, com acromegalia devido a um adenoma da glândula pituitária diagnosticado, e submetida a cirurgia aos 38 anos, mas com recorrência e reoperação aos 50 anos. A doente foi referenciada ao nosso departamento de cardiologia por um quadro com três meses de evolução de dispneia de esforço e ortopneia com agravamento progressivo sob terapêutica médica otimizada. O ecocardiograma e ressonância magnética cardíaca revelaram uma hipertrofia grave da porção basal do septo interventricular (19 mm), com um gradiente obstrutivo dinâmico da câmara de saída do ventrículo esquerdo de 70 mmHg em repouso e de 120 mmHg com a manobra de Valsalva e com movimento sistólico anterior (SAM) da válvula mitral. A avaliação genética excluiu as formas mais frequentes de cardiomiopatia hipertrófica (CMH) familiar. A ASA foi realizada com 2 cc de injeção de álcool na primeira artéria coronária septal, sem complicações. Na reavaliação do primeiro ano, houve uma melhora clínica e ecocardiográfica significativa com o desaparecimento de SAM. Tanto quanto é do nosso conhecimento, não existe na literatura a descrição de casos de ASA em doentes com CMH por acromegalia. Relatamos aqui o caso de uma ASA bem-sucedida numa cardiomiopatia acromegálica.

Introduction

Acromegaly is a rare disease that is most often caused by a growth hormone (GH)-secreting benign pituitary tumor and is characterized by progressive skeletal and soft tissue overgrowth, frequently accompanied by cardiovascular, cerebrovascular, and respiratory complications that contribute to the poor prognosis observed in this condition.[1] Cardiovascular disorders are common and are mainly responsible for the two-fold increase in mortality seen in acromegalic patients.[1,2] The increased autonomous production of GH results in increased insulin-like growth factor 1 (IGF-1), and chronic excess secretion of both hormones is associated with various changes in cardiac structure and function due to cardiomyocyte apoptosis, with concentric cardiac hypertrophy being a frequent finding even in young patients with short disease duration. As a consequence, early impairment of left ventricular (LV) diastolic filling has been described, while impaired systolic function with heart failure may develop in later stages if the disease is untreated or unsuccessfully treated.[1–3] Other cardiovascular disorders such as hypertension, coronary artery disease and ventricular arrhythmias may be present and worsen acromegalic cardiomyopathy, defined as any cardiac involvement in acromegalic patients in the absence of other known causes of cardiomyopathy.[1] Lowering of GH and IGF-1 to age-corrected normal limits is mandatory; however, treatment of acromegaly appears to improve cardiac function only in the short term, with little or no decrease in left ventricular mass or improvement in cardiac function after prolonged treatment.[4]

Alcohol septal ablation (ASA) has emerged as a minimally invasive alternative to septal myectomy for patients with obstructive hypertrophic cardiomyopathy (HCM) and symptoms refractory to medical therapy. This procedure alleviates symptoms by producing a targeted, limited infarction of the upper interventricular septum, resulting in an increase in left ventricular outflow tract (LVOT) diameter, a decrease in LVOT gradients, and regression of the component of LV hypertrophy that is due to pressure overload. Clinical success, with significant improvement in symptoms and reduction in gradients, is achieved in the great majority of patients and has been correlated with left ventricular remodeling. Non-randomized comparisons of septal ablation and septal myectomy have shown similar mortality rates and post-procedure New York Heart Association (NYHA) class for the two procedures.[5,6] To our knowledge, there have been no previously reported cases of ASA in hypertrophic cardiomyopathy due to acromegaly. We report a case of successful ASA in acromegalic cardiomyopathy.

Case report

We describe the case of a 73-year-old woman with acromegaly due to a pituitary adenoma diagnosed and