Two-Dimensional Speckle Strain Imaging in the Management of Paraganglioma, Acute Junctional Tachycardia, and Myocardial Dysfunction in a Child

Two-dimensional speckle-tracking strain imaging (speckle strain imaging) is useful for evaluating left ventricular myocardial function in patients with ischemic heart disease and cardiomyopathy, including hypertrophic and dilated phenotypes. The usefulness of speckle strain imaging in patients with pheochromocytoma who are undergoing adrenal surgery has been described, but we found no reports of the use of this method to evaluate ventricular dysfunction longitudinally in children. Herein, we describe the case of a 10-year-old girl with a paraganglioma, acute junctional tachycardia, and myocardial dysfunction. After control of the tachycardia and partial resection of the tumor, speckle strain imaging enabled clinical management that led to substantial improvement in the patient’s initially diffuse myocardial dysfunction. Because conventional echocardiographic methods alone may be inadequate to guide the management of pediatric patients with partially resected neuroendocrine tumors, we recommend speckle strain imaging as an additional noninvasive option for treatment guidance and monitoring of cardiac tissue response. (Tex Heart Inst J 2012;39(1):119-21)

Paraganglioma and pheochromocytoma are neuroendocrine tumors that produce excessive levels of catecholamines. Cardiac dysfunction, which is well documented in association with these tumors, is thought to be secondary to subendocardial ischemia, hypertension, or catecholamine toxicity. The treatment of paraganglioma and pheochromocytoma involves the preoperative control of hypertension and subsequent resection of the tumor.

Currently, the standard analysis of regional myocardial function consists of visual evaluation of inward motion and wall thickening from 2-dimensional (2D) echocardiographic images. This method is subjective and substantially limited by reader variability and experience. Strain and strain-rate imaging has been used to evaluate early myocardial dysfunction and regional wall-motion abnormalities. Strain analysis measures the deformation of cardiac muscle during the cardiac cycle. Furthermore, improvements in 2D echocardiographic image resolution have enabled the detection of tissue pixels and the tracking of these acoustic markers from frame to frame. The evaluation of longitudinal, radial, and circumferential strain and strain rate from a tissue pixel-tracking system on 2D echocardiographic images has recently been described. Whereas the standard imaging method can show preserved ventricular function, the alternative methods can reveal depressed myocardial contractility.

We found no published reports about the usefulness of 2D speckle-tracking strain imaging—speckle strain imaging (SSI)—in children who have catecholamine-secreting tumors. Herein, we describe the case of a child with a paraganglioma, severe hypertension, acute junctional tachycardia, and myocardial dysfunction whose case management and immediate follow-up evaluation were aided by the use of SSI echocardiography.

Case Report

A 10-year-old girl presented emergently with the chief symptom of a “fast heartbeat” that awakened her from sleep. On examination, her heart rate varied from 120 to 170 beats/min, and her blood pressure was 160/90 mmHg. Her medical history was notable for schizophrenia, and her medications included quetiapine, escitalopram, valproic...
acid, and clonidine. Electrocardiography revealed acute junctional tachycardia (heart rate, 202 beats/min) with no ST-segment changes.

The patient was admitted to the hospital and underwent extensive testing that showed normal thyroid function and elevated plasma catecholamine levels: noradrenaline, 9,166 pg/mL (normal, 150–400 pg/mL); and normetanephrine, 39.3 nmol/L (normal, ≤0.89 nmol/L). The epinephrine level of 25 pg/mL was within normal limits. Pheochromocytoma was suspected, and abdominal magnetic resonance imaging (MRI) revealed a para-aortic mass with involvement of the L4 vertebral body. In addition, 123I-metaiodobenzylguanidine scintigraphy was performed, and the uptake of the contrast medium was consistent with the MRI findings.

Routine echocardiographic evaluation showed moderately depressed myocardial function (left ventricular [LV] ejection fraction, 0.32), and SSI revealed diffuse LV myocardial dysfunction, particularly in the basal myocardial segments (Fig. 1). The global peak systolic longitudinal strain was –9% (normal, greater than –18%).

For the next 2 weeks, the patient was given verapamil for tachycardia and enalapril, clonidine, and phenoxybenzamine for hypertension. After the tachycardia was controlled and the patient’s blood pressure had fallen to 90/50 mmHg, the paraganglioma was resected; however, the neuroendocrine tissue around vertebral L4 was not treated during this intervention. Global ventricular function improved postoperatively (LV ejection fraction, 0.55). The global peak systolic longitudinal strain was –19%, with improvements in previous areas of localized regional wall motion (Fig. 2).

**Fig. 1** Depicted in a bulls-eye format, 2-dimensional echocardiographic speckle strain imaging shows the peak systolic strain of longitudinal myocardial fibers shortly after the patient’s hospital admission. Several areas show different degrees of systolic strain. The apical segments have a preserved deformation when compared with the basal segments.

**Fig. 2** Speckle strain imaging after partial tumor resection shows overall improvement in peak systolic strain; some basal segments show regional wall-motion abnormalities.

**Fig. 3** Speckle strain imaging a few weeks postoperatively shows improvement in the overall pattern of deformation, although the septal and anterolateral basal segments have decreased peak systolic strain values.
The patient was weaned from the cardiac medications and was discharged from the hospital 2 weeks after surgery. A few weeks later, echocardiography showed improved cardiac function and LV ejection fraction. Normetanephrine levels were still slightly elevated (1.12 nmol/L). On SSI analysis, LV myocardial function was substantially improved; however, persistent areas of basal myocardial-segment dysfunction were noted (global peak systolic longitudinal strain, –18.5%) (Fig. 3).

Discussion

Catecholamine-secreting tumors have been associated with depressed myocardial contractility. This patient presented with diffuse regional wall-motion abnormalities, as detected by SSI and standard 2D echocardiography. After the paraganglioma was partially resected, systolic function appeared to be normal on 2D echocardiography, but regional wall-motion abnormalities persisted on SSI. These abnormalities may have been due to a myocardial band of necrosis associated with inflammation and resulting in fibrosis. The global myocardial dysfunction could have been due to focal ischemia, a possibly increased influx of intracellular calcium, and increased production of toxic free radicals. Because the affected basal segments showed some residual deterioration, anti-remodeling cardiac therapy could have been considered.

Strain-rate imaging can be used preoperatively to evaluate morbidity: adults with pheochromocytoma who have a strain rate of less than 2 s⁻¹ are at high risk of circulatory collapse.

In conclusion, conventional echocardiographic methods alone may be inadequate to aid the management of pediatric patients with partially resected neuroendocrine tumors. In such patients, additional noninvasive options such as SSI can help to guide treatment and improve the monitoring of cardiac tissue response. In our patient, SSI was beneficial during management and follow-up, especially in the evaluation of elevated catecholamine levels and their effect on the myocardial tissue.