Cerebral Embolism Associated With Left Atrial Myxoma

Introduction

Primary cardiac tumors are uncommon in the pediatric population. The vast majority of pediatric cardiac tumors are benign and only 10% are malignant.

Cardiac tumors are rare causes of cerebrovascular disease in children. Most of these tumors are non-malignant and primarily associated with left atrial myxoma — a fibromyxomatous lesion of the left atrial wall.

Abstract

Primary cardiac tumors are very rare in the pediatric population, with an incidence of 0.2% to 0.5%. The majority of pediatric cardiac tumors are almost 95% benign and 5% malignant.

Rhabdomyoma is the most common cardiac tumor in children 80-90% associated with tuberous sclerosis. Myxomas can occur in all regions of the heart, and may result in compression of cardiac structures, valvular insufficiency, and outflow obstruction.

Symptoms range from nonspecific and constitutional to sudden cardiac death. In about 20% of cases, myxomas may be asymptomatic and discovered as an incidental finding. Because of nonspecific symptoms, early diagnosis may be a challenge.

Tumors in the left side of the heart can cause cerebral embolization and occasionally sudden death. The neurologic manifestations are mainly focal deficits from the emboli; and it’s the most common complication of a cardiac tumor.

Echocardiogram is a non-invasive test that will provide an immediate and accurate diagnosis at the bedside. The definitive treatment is resection of the tumor.

Case Presentation

A 12-year-old Hispanic female, presented to the emergency room with a 3-week history of left-sided weakness. She stated that while taking a shower, she felt dizzy, collapsed, and hit the right side of her head against the wall. She subsequently developed left-sided weakness and headache which she described as 5/10, generated, throbbing in type. She also developed nausea and vomiting. Past medical history is unremarkable.

PHYSICAL EXAMINATION: Vital signs are within normal limits. Her weight, height, and BMI are also within normal range. She was oriented x 3 with periods of drowsiness during examination and difficulty following commands.

Significant findings include the presence of vertical and horizontal nystagmus, muscle strength of 4/5 on the left upper and left lower extremity and hyporeflexia in all 4 extremities; failed point-to-point movements and rapid alternating movements, positive Romberg and presence of wale gait. The rest of the physical examination findings are normal.

Results

RADIOGRAPHIC FINDINGS:
- BRAIN MRI: multiple acute and chronic foci of infarct. (Figure 1, 2)
- BRAIN MRA: no abnormalities of intracranial arteries.

ECHOCARDIOGRAM:
- Pedunculated mass on top of mid mitral 5.6 cm x 2.87 cm. No-flow obstruction. (Figure 3, 4)

HISTOPATHOLOGY:
- Gross description: tan, pink, rubbery, gelatinous soft tissue 4.6 mm x 2.1 cm consistent with myxoma, also present white soft-tissue possible valve or wall atrial 1.9 x 1.5 x 0.3 cm
- Microscopic description: polygonal and lipoid cells surrounded by abundant loose stroma. The cells are arranged singly, in small nests forming vascular channels. (Figure 5)

Figure 1

Figure 2

Figure 3

Figure 4

Figure 5

Conclusions

The heart is the most common source of cerebrovascular events in the pediatric population. The most common presentation of cerebral vascular accident in children is acute hemiplegia. Embolic infarction is the most common neurologic complication of a left atrial myxoma. The absence of other clinical evidence for cardiac disease should not dissuade one from considering a cardiac embolic cause. Our patient did not have a cardiac murmur at the time of presentation.

Myxomas are very friable, and the emboli consist mainly of myxomatous material. The tumor can occur in all regions of the heart, and may result in compression of cardiac structures, valvular insufficiency, and outflow obstruction. Myxomas in the left side of the heart are responsible for cerebral embolization and occasionally sudden death. Two-dimensional echocardiography is a non-invasive and highly accurate for the diagnosis.

The definitive treatment is resection of the tumor, which will alleviate the cardiac symptoms and prevent any further cerebral emboli. Regular assessment by the pediatrician, cardiologist, neurologist, and physical therapist will provide the most comprehensive treatment.

References