Cardiac myxoma: an unusual cause of sudden death in childhood

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SUMMARY: Akyıldız EÜ, Tolgay E, Öz B, Yılmaz R, Koç S. Cardiac myxoma: an unusual cause of sudden death in childhood. Turk J Pediatr 2006; 48: 172-174.

Primary tumors of the heart and pericardium are rare in autopsy series. Considering all age groups, the most common cardiac tumor is the myxoma. They may arise in any of the four chambers or, rarely, on the heart valves. About 90% are located in the atria, with the left-to-right ratio of 4:1. The majority of patients are usually in the age group of 30-60 years. Female predominance has been reported. In this report, we describe a six-year-old boy with right atrial myxoma. A 5x4x3 cm, polypoid, smooth-surfaced, brown-colored tumor was found in the right atrium. It was attached by the stalk to the atrial septum. Microscopically, the tumor had the typical appearance of a myxoma with spindle cells widely spaced by abundant myxoid matrix. The diagnosis was based on histopathological and immunohistochemical findings.

Sudden death may occur in patients with atrial myxoma. Death is caused by coronary or systemic embolization or by obstruction of blood flow at the mitral or tricuspid valve.

Key words: myxoma, autopsy, sudden death.

Primary cardiac tumors are rare, occurring in only 0.0017% to 0.28% of autopsies[1]. Considering all age groups, the most common cardiac tumor is the myxoma[2,3]. In adults, almost half of the benign cardiac tumors are cardiac myxomas, but in infants and children, the most common cardiac tumor is the rhabdomyoma[3].

Cardiac myxomas arise from the endocardium as a soft, polypoid, lobulated mass often attached by a stalk to the septum near the foramen ovale, and 90% protrude into the left atrium[4,5]. Right atrial myxomas are three to four times less frequent than those located in the left atrium[2].

The clinical presentation is primarily dependent upon the cardiac chamber involved[3]. The pedunculated myxoma is frequently sufficiently mobile to move into or through the atrioventricular valves during systole[6]. The most common clinical presentation is systemic emboli. The second most common symptom is congestive heart failure due to mitral valve obstruction with murmurs and atrial arrhythmias.

The majority of patients are usually in the age group of 30-60 years. There is an equal sex distribution in most series.

Most myxomas occur as single tumors, but multiple tumors have been reported in families[5].

Case Report

A six-year-old boy was admitted to the hospital with syncope and cyanosis, and he died in the hospital on the same day. The public prosecutor sent the body for autopsy because of sudden unexpected death. The diagnosis was made by autopsy and microscopic examination.

A 5x4x3 cm, polypoid, smooth-surfaced, brown-colored tumor was found in the right atrium. It was attached by the stalk to the atrial septum (Fig. 1). Hemorrhagic areas were seen in the cut surfaces.

The specimen was fixed with 10% formaldehyde and embedded in paraffin. Sections (4 micron thick) were stained with hematoxylin and eosin. Immunohistochemical analysis [smooth muscle

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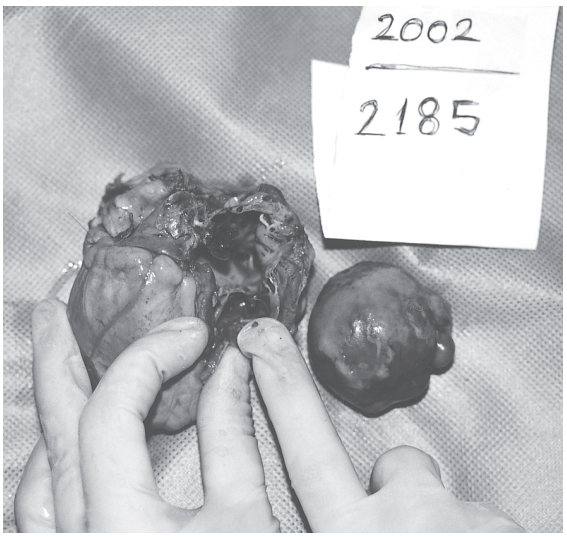

Fig. 1. A photograph showing the tumor attached by the stalk to the atrial septum.

actin (NeoMarker MS113-P, 1/200), desmin (DAKO, D33, N1526, predilue), S 100 (DAKO, N 1573, predilue)] was performed.

There was no other finding except intraalveolar hemorrhage in the lung and stasis in the liver.

Microscopically, the tumor had the typical appearance of a myxoma with spindle cells widely spaced by abundant myxoid matrix (Fig. 2). There were areas of hemorrhage and chronic inflammatory cells. Mitosis, pleomorphism and necrosis were absent.

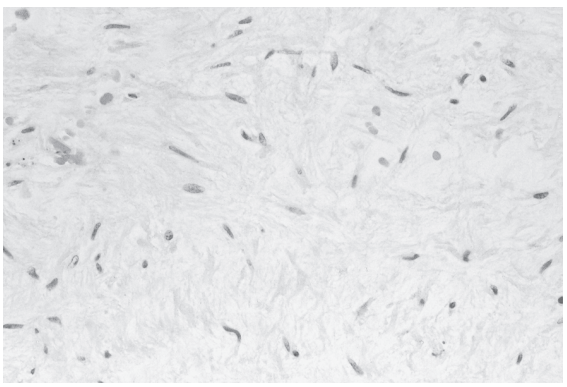

Fig. 2. Microscopic examination of the tumoral tissue showing spindle cells widely spaced by abundant myxoid matrix (H&Ex400).

Immunohistochemically, the tumor cells were positive for smooth muscle actin (SMA), but negative for desmin and S 100.

Discussion

Cardiac myxomas are uncommon, with a reported incidence of 1 in 10,000 autopsies[2]. They occur in all age groups, but more than 50% of patients with cardiac myxoma are in their fourth, fifth, or sixth decade[3]. Eighteen patients (6 males, 12 females) aged 24 to 73 years (mean 55.3) with cardiac myxomas were reported by Durgut et al.[7]. Comparison of clinical features among patients with nonfamilial cardiac myxoma versus those of patients with familial cardiac myxoma shows significant differences. Nonfamilial cardiac myxoma is a disorder of middle-aged (mean age, 51 years) women, while familial cardiac myxoma is a disorder of young (mean age, 24 years) men[8]. Cardiac myxomas are extremely rare in infancy. In the literature, Pasaoğlu et al.[9] reported a case of right atrial myxoma in a 35-day-old male infant. In 2003, Paladini et al.[10] presented right atrial mass in the fetus at 23 weeks' gestation and showed that myxomas can also arise in the fetus. Our case was a six-year-old boy.

Myxoma can occur in any of the chambers of the heart, but about 75% originate within the left atrium and only 18% within the right atrium. But in children, myxomas seem to have a much more variable site than in adults, especially in the right heart[3,11]. Right atrial myxomas are predominant in young children (5 years and younger) in contrast to older children and adults[12]. In our case, the tumor was located in the right atrium.

Although cardiac myxomas are histologically benign, they may be lethal because of their position[13,14]. The left-sided cardiac myxomas may present with signs of mitral stenosis and insufficiency, and right-sided tumors with dyspnea, syncope, and distention of neck veins. They may also lead to multiple emboli in the systemic or pulmonary circulation, depending on their localization. The most common clinical presentation relates to obstruction of the flow of blood. Surgical excision is often curative[4]. Sudden unexpected death was reported in 5% of 130 patients with cardiac myxoma[3]. Right-sided tumors are less often symptomatic[15]. Our case with no previous medical history died shortly after he was admitted to the hospital.

Myxomas were originally believed to be thrombotic in the past but more recently it has been accepted that they are true

neoplasms. Thrombi are frequent in patients with underlying valve disease, while underlying cardiac disease is rare in myxomas. In our case there was no underlying valve disease[5].

The cell of origin has been in dispute. Cytokeratin is negative unless there is the glandular pattern on differentiation. Vimentin is usually positive in the tumor cells. Desmin is usually described as negative, but in some reports SMA shows positive reaction in the myxoma cells[5]. Johansson[16] performed an immunohistochemical study of 19 cardiac myxomas and the results indicated that myxomas arise from mesenchymal pluripotential cells, which are capable of many types of differentiation. In our case the tumor cells were positive for SMA and negative for desmin and S 100, so myofibroblastic cells are suggested as the cell of origin.

Sporadic and familial cases have been identified. The sporadic tumor occurs more frequently in middle-aged women, and usually in the left atrium (86%). The family variety is a disorder of young people, slightly more frequent in men and less commonly located in the left atrium (62%)[4]. Although our case was a six-year-old boy with right atrial myxoma, there was no medical history in family members.

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