Lipoblastoma: an unusual tumour of the left ventricle

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Abstract

Lipoblastoma is a rare benign tumour of infancy originating from white foetal adipose tissue. Most commonly located in the extremities, intrathoracic and mediastinal involvement of this tumour is rare, and an intracardiac location is even rarer, with only one reported case. Herein, we present a 2-month old asymptomatic boy diagnosed with an echogenic mass in the left ventricular outflow tract. The patient underwent surgical excision and histopathological evaluation revealed a lipoblastoma.

Keywords: Lipoblastoma • Intracardiac • Histopathology

INTRODUCTION

Lipoblastoma is a neoplasm of embryonic adipose tissue. Although rare in adults, it is known to occur in children of less than 3 years of age [1]. Histologically, this exclusively benign tumour with a very good prognosis consists of fat lobules of variable maturity, multivacuolated lipoblasts, fibrocapillary networks and myxoid stroma [2]. Less than 200 cases have been reported in the literature, occurring mostly in the extremities. Very few have involved the intrathoracic and mediastinal area, with only one reported case of an intracardiac lipoblastoma [3]. We thus, report this unusual presentation of intracardiac lipoblastoma in a 2-month old infant.

CASE REPORT

An apparently asymptomatic 2-month old boy was referred to our clinic, with presentation of coryza and cardiomegaly on chest roentgenogram. On examination, the child had normal vital and growth parameters. Cardiovascular examination revealed a soft systolic murmur of 2/6 in intensity at the base. Transthoracic echocardiography (Fig. 1A) revealed a large echogenic mass at the left ventricular outflow tract (LVOT) causing mild obstruction with a peak and mean gradient of 27 mm Hg and 15 mm of Hg, respectively.

Due to the risk for complete LVOT obstruction and possibility of embolization, the patient underwent excision of the tumour. Surgery was approached through a median sternotomy, with aortic and bicaval cannulation. Following antegrade cardiopulmonary arrest, transverse aortotomy was done. A 10 x 6 x 5 mm, well-encapsulated, grey white mass was located just below the non-coronary and the left coronary cusp and attached to the LVOT by a stalk; the aortic valve was tricuspid with normal morphology and was found to be competent. The mass was resected in total and the specimen sent for histopathological evaluation.

Histopathologically, the tumour was composed of lobules of mature and immature adipocytes separated by fibrovascular septa. There were foci of areas showing lipoblasts with centrally placed hyperchromatic nuclei, myxoid areas with spindle cells and few hibernoma-like cells in the peripheral areas (Fig. 2). The patient had an uneventful postoperative period and was discharged on the 11th postoperative day. At follow-up after 3 months, the child was asymptomatic with a good weight gain of 3 kg. On echocardiogram, there was absence of any echogenic mass with a widely patent LVOT without any obstruction (Fig. 1B).

DISCUSSION

We present an extremely rare case of an intracardiac lipoblastoma in an infant causing LVOT obstruction. Suggested to have a close relationship with foetal white adipose tissue, the morphological criterion of this benign embryonal tumour was established by Chung and Enzinger in 1973 [2]. Two types of pathological lipoblastoma have been identified: a localized, well-circumscribed lipoblastoma and irregularly confined, non-capsulating, multicentric lipoblastomatosis [1]. In our patient, the former histopathological type was present, which is more common and has a better prognosis [2].

Despite being rare, lipoblastoma accounts for more than 15% of benign tissue neoplasms in children. It has no gender predilection and is reported to occur in areas with primitive adipose tissue such as axilla, neck and prevertebral soft tissue, most commonly occurring in lower extremities (50%) followed by upper...
extremities and trunk [1]. Very few cases of lipoblastoma involving the mediastinal cavity have been reported with intracardiac involvement being even rarer. To our knowledge, only one such case of primary cardiac lipoblastoma has been reported, arising from the posterior interatrial septum located within the intrapericardial cavity [3]. This case probably is the first reported intracardiac lipoblastoma.

Usually asymptomatic, the severity of the symptoms depends on the size and location of the lipoblastoma. Although well-visualized ultrasonographically, showing a well-defined highly echogenic mass, computerized tomographic images may demonstrate a low-grade density of fat attenuation suggestive of lipoblastoma [4]. Confirmatory diagnosis is made by histopathological evaluation [1]. Although surgical excision of the tumour is considered to be curative, incomplete resection of the diffuse form may cause recurrence, which has been reported in 14–25% of cases [5]. Mognato et al. have reported spontaneous resolution of lipoblastoma of the left thigh in a 2-day old child [4]. However, in our patient surgical excision was decided based on the size of the tumour causing LVOT obstruction and risk for embolization.

CONCLUSION

We present a rare case of lipoblastoma of the left ventricle in an infant who underwent surgical excision. We advocate that it may be prudent to excise such intracardiac tumours, as recurrence is rare.

Conflict of interest: none declared.

REFERENCES