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Case Report

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Subaortic membrane in a turner syndrome: A case report

Abstract

Cardiovascular abnormalities are common in Turner syndrome. Coarctation of the aorta and aortic bicuspid valve are more commonly found. The presence of a subaortic membrane in patients with Turner syndrome has rarely been reported in the literature. We report a case of an isolated subaortic membrane responsible for severe sub valvular stenosis in a 26-year-old woman. The patient underwent surgical excision phase of the membrane under cardiopulmonary bypass. The post-operative course was uneventful.

Keywords: turner syndrome, subaortic membrane, surgery

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Introduction

Turner syndrome is a clinical syndromic set associated in 50% with an X monosomy (homogeneous or mosaical) and in the remaining 50% with rearrangements in the short arm of an X-chromosome.[1] The existence of cardiovascular abnormalities is well known in association with Turner syndrome. The most common are aortic valve disease and coarctation of the aorta. In our knowledge, the association between Turner syndrome and subaortic membrane is uncommon and rarely reported. We had to manage à patient with Turner syndrome who inderwent a subaortic membrane resection responsible for a very severe subvalvular stenosis.

Case presentation

We report a 26-year-old woman born from a consanguineous marriage. Turner syndrome was diagnosed at the age of 20 due to morphological abnormalities: small size, low implantation of hair and ears, naevi, nipple gap, with absence of mammary gland development and impuberism. The examinations carried out gave the following results: Molecular biology of X: a single band in favor of Turner syndrome, molecular biology of Y was negative. The abdomino-pelvic ultrasound done showed an absence of individualization of the uterus, an absence of individualization of the ovaries without a latero-uterine mass. The hormonal assessment showed hypothyroidism and hypogonadism. The bone age was estimated at 25years. As part of the assessment of her disease, the clinical examination found a systolic murmur in the aortic area. The patient was asymptomatic in cardiovascular terms.

After confirmation of the diagnosis of turner syndrome, the patient did not shown for follow-up, due to the family's precarious economic conditions and the difficulty of accessing healthcare facilities. The cardiac ultrasound was therefore only done 6years later.

The trans thoracic ultrasound showed an isolated subvalvular membrane responsible for a severe stenosis with an estimated mean gradient of 46mmHg, an hypertrophied non-dilated left ventricle for her age, an ejection fraction at 75% without coarctation of the aorta. Thoracic CT-Scan was normal.

Under Cardiopulmonary bypass, the patient underwent surgery for subaortic membrane excision and myomectomy. Postoperative follow-up was uneventful. Postoperative trans thoracic ultrasonography showed à mild gradient (10mmhg). A two years follow up did not found any sign of recurrence.

Discussion

Turner's syndrome has a set of malformations affecting different systems: skeletal malformations (micrognathia, shield thorax with exaggerated nipple spacing, narrowing pelvis, ulna valgus, genu varum, pterygium colli), renal (horseshoe kidney), cutaneous (nevi pigmentary), visceral, including ovarian agenesis, and finally cardio-vascular malformations. Sometimes there is an intellectual deficit but one can observe subjects with a normal or even high mental level.[1] Congenital heart defects are found in 23 to 40% of cases.[2,3] They mainly affect the left heart. The most common abnormality is bicuspid aortic valve[4,5] observed in 20% of subjects.

Aortic coarctation is also frequent, observed in 10 to 15% of cases.[6,7] Our patient didn't have bicuspidia or coartation of the aorta. The subaortic membrane is a thin fibrous diaphragm measuring 1 to 3 mm generally thick, inserted on the walls of the left ventricle outflow tract, 0.5 to 3cm below the aortic annulus.[8] The membrane is inserted forward on the interventricular septum and behind on the anterior mitral cusp.[8] It can be isolated or associated with other cardiac diseases. It is an acquired lesion with predisposing factors such as: a more acute angle than normal between the long axis of the left ventricle and the aorta, an increase in the mitro-aortic distance, a dextroposition of the aorta. These factors lead to an increase in subaortic shear forces with proliferation of tissue leading to the formation of a subaortic membrane. Surgery is indicated when the maximum gradient on cardiac ultrasound exceeds 30mmHg or as soon as aortic regurgitation appears.[8]

In our case the patient had a subaortic membrane without bicuspid of aortic valve; this membrane was responsible for a severe stenosis. During our patient's surgery, no anatomical abnormality was found to explain the existence of the subaortic membrane. Surgery remains the treatment of choice despite a real risk of recurrence in the long term.

Conclusion

Turner syndrome is frequently associated with cardiovascular abnormalities. The cardiological assessment is mandatory in turner syndrome patients. Surgical management is often possible even in young subjects, with good long-term results.

Patient consent

We declare that we received the informed consent of the patient and their family prior to the writing of this manuscript.

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None.

Conflicts of interest

The authors declare no conflict of interest.

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