LIPOMATOUS HYPERTROPHY OF THE INTERATRIAL SEPTUM – A BENIGN HEART ANOMALY CAUSING UNEXPECTED PROBLEM IN ELECTROPHYSIOLOGY (CASE REPORT)

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Lipomatous Hypertrophy of the Interatrial Septum (LHIS) is an unusual condition, usually benign and most often detected as an incidental finding on echocardiography. The classic finding is a homogenous, bi-lobed configuration of the interatrial septum with sparing of the fossa ovalis. This infiltration can also involve the septal tissue and has been associated with various atrial arrhythmias, including multifocal atrial tachycardia, multiple atrial premature contractions, atrial fibrillation and rarely even sudden death [1,d2]. Interatrial septal thickness >2 cm is considered diagnostic of LHIS [3].

The prevalence of LHIS is estimated to be between 1-8%. It usually occurs in older, obese people and there may be a higher incidence in women [4].

Differential diagnoses should be thought of when there is sparing of the fossa ovalis. Fat-containing neoplasms can arise in the atrial septum including cardiac lipoma, cardiac rhabdomyoma, cardiac myxoma, cardiac rhabdomyosarcoma and cardiac liposarcoma . Microscopically, LHIS is characterized by fat infiltration between the myocardial fibers of the atrial septum. [5]. LHIS also can create a mass-like bulge. There is typical sparing of the fossa ovalis.

With this case, we report the incidental clinical presentation and association between atrial arrhythmia and LHIS in an otherwise healthy obese woman.

Case report. A 73 year-old obese woman referred to our cardiology department for planned cryoablation procedure of atrial fibrillation. Past history: episodes of palpitations, dizziness, and fatigue of brief duration. Her family history was unremarkable. Physical examination disclosed obesity (weight - 86 kg, height - 164 cm, BMI - 31,97 kg/m^2). Systemic blood pressure was 132/84 mm Hg; Heart rate (apical) was 85 beats/min, sometimes irregular, and respiratory rate was 18 per min. The cardiac apex was palpable in the fourth intercostal space at the left anterior axillary line. A mild systolic ejection murmur was heard at the apex of the heart. It was associated with normal carotid impulses bilaterally. No diastolic murmurs or sounds were heard. The 12 lead ECG showed sinus rhythm. Past history: episodes of atrial fibrillation with a rapid ventricular response and nonspecific ST-T-wave changes. The patient was treated with intravenous cordaron and a normal sinus rhythm was restored quickly. A chest x-ray was normal. Therefore, the patient was discharged in good health with an indication to further planned cryoablation of atrial fibrillation.

During the procedure in the electrophysiology laboratory arithmologists revealed some difficulties during septal punction. Transseptal catheterization access seemed impossible. This approach is commonly used in electrophysiology and interventional cardiology to treat number of arrhythmias and anatomical defects of the heart. In our case, treatment of left atrial arrhythmia through commonly used transseptal puncture became impossible due to intracardiac unusual mass. A problem related to the anatomy of the interatrial septum made technical difficulties during the transseptal puncture. This difficulties arised in accessing the operated left heart structures, resistance was encountered while inserting catheters.

Subsequently two-dimensional transthoracic echocardiography (TTE) had been performed in electrophysiology laboratory, subcostal four-chamber approach showed a hyperechogenic mass in the interatrial septum. There was no decrease in flow velocities of the superior and inferior vena cava nor a flow disturbance in the pulmonary veins. Ejection fraction was in normal range, mild to moderate dilatation of left atria with mild mitral regurgitation had been revealed. TTE showed thickening and increased echogenicity of the inferior and superior portions of the atrial septum with sparing of the fossa ovalis. These findings were typical for fatty infiltrate and resulted in a “dumbbell-shaped” appearance of interatrial septum on two-dimensional transthoracic echocardiography (2D TTE). The lesion had characteristic an hourglass shape sparing the fossa ovalis. It had pathognomonic imaging features and did not require additional imaging. Based on the performed intraoperatively TTE, a diagnosis of lipomatous hypertrophy of the interatrial septum was made (Fig.).

It was an incidental finding. Access to the operated left atrium was significantly impeded, and the transseptal approach, without disturbing the LHIS structure, could not be possible.

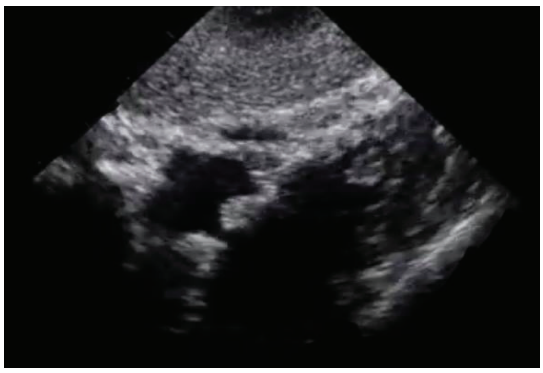

Fig. Echocardiographic Finding

Transthoracic echocardiogram showing lipomatous hypertrophy of the intraatrial septum. Subcostal four- chamber view shows echo dense structure of the interatrial septum. These findings are consistent with lipomatous hypertrophy of the interatrial septum.

A 73 year-old asymptomatic woman was found to have an incidental cardiac mass, TTE findings ware consistent with lipotamous hypertrophy of the interatrial septum. Given the characteristic appearances on TTE, biopsy or surgery was not indicated, the procedure of cryoablation has been stopped and the patient was managed conservatively.

Lipomatous hypertrophy of the interatrial septum is a rare but increasingly recognized non-neoplastic benign abnormality of the heart. LHIS must be included in the differential diagnosis of any right atrial mass, or any fat-containing neoplasm. This condition is more common than true cardiac lipomas, occurring almost exclusively in elderly, obese patients and is usually asymptomatic. Unlike lipomas, the fatty lesions of LHIS are not encapsulated. TTE diagnosis of LHIS is based on the classic morphology of the thickened, echogenic bi-lobed septal mass, which always spares the foramen ovale, and this distinguishes it from other car-

diac lesions, such as lipomas, liposarcomas, metastatic tumors, myxomas and amyloidosis, which can be also present as a septal tumor mass.

The first descriptions of LHIS in vivo were made in 1983 on the basis of echocardiography by Fyke et al. who published the first diagnostic guidelines [6]. Currently, TTE, TEE, CT and MRI are used for diagnostics. Lipomatous lesion derives entirely from the upper and/or lower part of the atrial septum, typically sparing the fossa ovalis. The pathologic mass makes a characteristic, considered by some to be pathognomonic, an hourglass-shaped image of atrial septum. The importance of this diagnosis should not be confused with other lesions that may occur in this part of the heart including, but not limited to, lipoma, liposarcoma, teratoma, myxoma or other benign tumors of the heart and avoid unnecessary investigations and anxiety to the patient and the ordering physician.

Besides being usually benign and asymptomatic, a significant number of patients have been reported to have unexplained arrhythmias such as atrial fibrillation, atrioventricular (AV) block and sudden death.

The differential diagnosis for a fat-containing cardiac tumor includes LHIS, liposarcoma, and lipoma. The presence of a fatty tumor within the interatrial septum, sparing the fossa ovalis and thus creating a dumbbell appearance, is pathognomonic for LHIS.

Asymptomatic LHIS does not require cardiac surgery. Surgical treatment of LHIS should be limited only to cases of patients with marginal obstruction of the superior vena cava (SVC) or the right atrium, which is an indication for a resection of the lesion with simultaneous interatrial septum plasty [7]. In the presented case, a procedure of cryoablation has been stopped and patient continued medical treatment.

LHIS can cause undesirable consequences, may render some percutaneous and surgical interventions particularly challenging, as it happened in our case. Pre-interventional recognition of LHIS is very important for invasive cardiological interventions involving transseptal catheterization access. This approach is commonly used in electrophysiology and interventional cardiology to treat number of arrhythmias and anatomical defects of the heart, such as closure of patent foramen ovales (PFO), atrial septal defects (ASDs), or correction of the functional mitral regurgitation through percutaneous "edge-to-edge" mitral valve repair. In our case, interventional electrophysiology to treat left atrial arrhythmias through commonly used transseptal puncture became impossible due to intracardiac mass - LHIS. A very rare problem related to the size of the mass and the anatomy of the interatrial septum are technical difficulties, as described above, which arise during the transseptal puncture when accessing the operated heart structures. In the presented case, resistance was encountered while inserting catheters. Access to the operated left atrium was significantly impeded. It was apparent, that the transseptal approach, without damaging interatrial septum, would be impossible. Therefore, the procedure was stopped. Based on the intraoperatively performed TTE, a diagnosis of LHIS was made.

In conclusion, this case confirms that LHIS is an uncommon anomaly, often diagnosed incidentally and usually not requiring intervention. However, it can be associated with the paroxysmal atrial fibrillation in otherwise healthy, but obese persons.

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SUMMARY

LIPOMATOUS HYPERTROPHY OF THE INTERATRIAL SEPTUM – A BENIGN HEART ANOMALY CAUSING UNEXPECTED PROBLEM IN ELECTROPHYSIOLOGY (CASE REPORT)

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Lipomatous Hypertrophy of the Interatrial Septum (LHIS) is an unusual and benign condition characterized by the excessive deposition of adipose tissue in the interatrial septum, which is most often detected as an incidental finding on echocardiography. The classic finding is a homogenous, bi-lobed configuration of the interatrial septum with sparing of the fossa ovalis. LHIS has been associated with various atrial arrhythmias, including multifocal atrial tachycardia, multiple premature atrial contractions, atrial fibrillation and rarely sudden death.

The prevalence of LHIS is estimated to be between 1-8%. The incidence increases with age, body mass and chronic corticosteroid therapy. There may be a higher incidence in women.

Here the authors describe a case report of a 73 year-old obese female who visited the cardiology department for planned cryoablation of paroxysmal atrial fibrillation. Difficulties raised during transseptal punction, a bidimensional tranthoracic echocardiography (TTE) showed the typical findings of LHIS.

A 73 year-old asymptomatic woman was found to have an incidental cardiac mass, TTE findings were consistent with lipomatous hypertrophy of the interatrial septum. Given the characteristic appearance on TTE, biopsy or surgery was not indicated, the procedure of cryoablation has been stopped and the patient was managed conservatively.

Keywords: hypertrophy, lipomatous, septum, echocardiography, obese woman. atrial fibrillation, electrophysiology.

РЕЗЮМЕ

ЛИПОМАТОЗНАЯ ГИПЕРТРОФИЯ МЕЖПРЕДСЕРДНОЙ ПЕРЕГОРОДКИ – ДОБРОКАЧЕСТВЕННАЯ АНОМАЛИЯ СЕРДЦА, ВЫЗЫВАЮЩАЯ НЕОЖИДАННУЮ ПРОБЛЕМУ В ЭЛЕКТРОФИЗИОЛОГИИ (СЛУЧАЙ ИЗ ПРАКТИКИ)

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Липоматозная гипертрофия межпредсердной перегородки (LHIS) - необычное и доброкачественное состояние, характеризуется чрезмерным отложением жировой ткани в межпредсердной перегородке и случайно обнаруживается при эхокардиографии. Классическим проявлением является однородная билобальная конфигурация межпредсердной перегородки с сохранением овальной ямки. LHIS ассоциируется с различными предсердными аритмиями, включая мультифокальную предсердную тахикардию, множественные преждевременные сокращения предсердий, фибрилляцию предсердий и, в редких случаях, внезапную смерть. Распространенность LHIS колеблется в пределах от 1% до 8%, число случаев увеличивается с возрастом, массой тела и продолжительной кортикостероидной терапией, чаще обнаруживается у женщин.

Представлен клинический случай 73-летней женщины с ожирением, которая обратилась в кардиологическое отделение для плановой криоаблации пароксизмальной фибрилляции предсердий. Трудности возникли при транссептальной пункции, двумерная трансторакальная эхокардиография (ТТЕ) показала типичные характерные признаки LHIS. У женщины случайно обнаружилась внутрисердечная масса, результаты ТТЕ соответствовали липоматозной гипертрофии межпредсердной перегородки. Учитывая характерный вид на ТТЕ, биопсия или хирургическое вмешательство не показаны, процедура криоаблации была прервана, и лечение пациентки продолжилось консервативно.

Представленный случай подтверждает, что LHIS является необычной аномалией, часто диагностируется случайно и обычно не требует вмешательств, однако может быть связана с пароксизмальной фибрилляцией предсердий у здоровых, страдающих ожирением лиц.

რეზიუმე

წინაგულთაშუა ძგიდის ლიპომატოზური ჰიპერტროფია - გულის კეთილთვისებიანი ანომალია, მოულოდნელი პრობლემების გამომწვევი ელექტროფიზიოლოგიაში (კლინიკური შემთხვევა)

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თბილისის სახელმწიფო სამედიცინო უნივერსიტეტი

წინაგულთაშუა ძგიდის ლიპომატოზური ჰიპერტროფია (LHIS) უჩვეულო და კეთილთვისებიანი მდგომარეობაა, რომელიც ხასიათდება ცხიმოვანი ქსოვილის ჭარბი ჩალაგებით წინაგულთაშუა ძგიდეში და შემთხვევითი ექოკარდიოგრაფიული აღმოჩენაა. მის კლასიკურ გამოვლინებას წარმოადგენს ჰომოგენური (ოვალური ფოსოს გარდა), ბილობალური კონფიგურაციის წინაგულთშუა ძგიდე. LHIS ასოცირდება სხვადასხვა წინაგულოვან არითმიებთან, მათ შორის მულტიფოკალურ წინაგულოვან ტაქიკარდიას, წინაგულოვან ექსტრასისტოლიას, წინაგულთა ფიბრილაციას და იშვიათად უეცარ სიკვდილთან. LHIS-ის გავრცელება მერყეობს 1-8% შორის. შემთხვევები მატულობს ასაკთან, სხეულის მასასთან და ქრონიკულ კორტიკოსტეროიდულ თერაპიასთან ერთად. უფრო ხშირად ვლინდება ქალბატონებში.

აღწერილი კლინიკური შემთხვევა ეხება 73 წლის ჭარბწონიან ქალბატონს, რომელმაც კარდიოლოგიურ განყოფილებას მიმართა პაროქსიზმული წინაგულთა ფიბრილაციის გეგმიური კრიოაბლაციის მიზნით. სირთულეები წარმოიშვა ტრანსსეპტალური პუნქციის განხორციელებისას, ორგანზომილებიანი ექოკარდიოგრაფით (ТТЕ) გამოვლინდა LHIS-თვის დამახასიათებელი ტიპური ნიშნები.

ასიმპტომურ ავადმყოფს შემთხვევით აღმოაჩნდა გულშიდა მასა, რომელიც ТТЕ ნიშნებით შეესაბამებოდა წინაგულთაშუა ძგიდის ლიპომატოზურ ჰიპერტროფიას. დამახასიათებელი ТТЕ გამოვლინების გამო, ბიოფსიის ან ქირურგიის ჩვენება არ იყო, კრიოაბლაციის პროცედურა შეწყდა და პაციენტის მკურნალობა წარიმართა კონსერვატულად.