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ЕЖЕМЕСЯЧНЫЙ НАУЧНЫЙ ЖУРНАЛ

Медицинские новости Грузии

საქართველოს სამედიცინო სიახლენი

GEORGIAN MEDICAL NEWS

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GMN: Georgian Medical News is peer-reviewed, published monthly journal committed to promoting the science and art of medicine and the betterment of public health, published by the GMN Editorial Board since 1994. GMN carries original scientific articles on medicine, biology and pharmacy, which are of experimental, theoretical and practical character; publishes original research, reviews, commentaries, editorials, essays, medical news, and correspondence in English and Russian.

GMN is indexed in MEDLINE, SCOPUS, PubMed and VINITI Russian Academy of Sciences. The full text content is available through EBSCO databases.

GMN: Медицинские новости Грузии - ежемесячный рецензируемый научный журнал, издаётся Редакционной коллегией с 1994 года на русском и английском языках в целях поддержки медицинской науки и улучшения здравоохранения. В журнале публикуются оригинальные научные статьи в области медицины, биологии и фармации, статьи обзорного характера, научные сообщения, новости медицины и здравоохранения. Журнал индексируется в MEDLINE, отражён в базе данных SCOPUS, PubMed и ВИНИТИ РАН. Полнотекстовые статьи журнала доступны через БД EBSCO.

GMN: Georgian Medical News – საქართველოს სამედიცინო სიახლენი – არის ყოველთვიური სამეცნიერო სამედიცინო რეცენზირებადი ჟურნალი, გამოიცემა 1994 წლიდან, წარმოადგენს სარედაქციო კოლეგიისა და აშშ-ის მეცნიერების, განათლების, ინდუსტრიის, ხელოვნებისა და ბუნებისმეტყველების საერთაშორისო აკადემიის ერთობლივ გამოცემას. GMN-ში რუსულ და ინგლისურ ენებზე ქვეყნდება ექსპერიმენტული, თეორიული და პრაქტიკული ხასიათის ორიგინალური სამეცნიერო სტატიები მედიცინის, ბიოლოგიისა და ფარმაციის სფეროში, მიმოხილვითი ხასიათის სტატიები.

ჟურნალი ინდექსირებულია MEDLINE-ის საერთაშორისო სისტემაში, ასახულია SCOPUS-ის, PubMed-ის და ВИНИТИ РАН-ის მონაცემთა ბაზებში. სტატიების სრული ტექსტი ხელმისაწვდომია EBSCO-ს მონაცემთა ბაზებიდან.

WEBSITE

www.geomednews.com

К СВЕДЕНИЮ АВТОРОВ!

При направлении статьи в редакцию необходимо соблюдать следующие правила:

1. Статья должна быть представлена в двух экземплярах, на русском или английском языках, напечатанная через **полтора интервала на одной стороне стандартного листа с шириной левого поля в три сантиметра**. Используемый компьютерный шрифт для текста на русском и английском языках - **Times New Roman** (**Кириллица**), для текста на грузинском языке следует использовать **AcadNusx.** Размер шрифта - **12**. К рукописи, напечатанной на компьютере, должен быть приложен CD со статьей.

2. Размер статьи должен быть не менее десяти и не более двадцати страниц машинописи, включая указатель литературы и резюме на английском, русском и грузинском языках.

3. В статье должны быть освещены актуальность данного материала, методы и результаты исследования и их обсуждение.

При представлении в печать научных экспериментальных работ авторы должны указывать вид и количество экспериментальных животных, применявшиеся методы обезболивания и усыпления (в ходе острых опытов).

4. К статье должны быть приложены краткое (на полстраницы) резюме на английском, русском и грузинском языках (включающее следующие разделы: цель исследования, материал и методы, результаты и заключение) и список ключевых слов (key words).

5. Таблицы необходимо представлять в печатной форме. Фотокопии не принимаются. **Все цифровые, итоговые и процентные данные в таблицах должны соответствовать таковым в тексте статьи**. Таблицы и графики должны быть озаглавлены.

6. Фотографии должны быть контрастными, фотокопии с рентгенограмм - в позитивном изображении. Рисунки, чертежи и диаграммы следует озаглавить, пронумеровать и вставить в соответствующее место текста **в tiff формате**.

В подписях к микрофотографиям следует указывать степень увеличения через окуляр или объектив и метод окраски или импрегнации срезов.

7. Фамилии отечественных авторов приводятся в оригинальной транскрипции.

8. При оформлении и направлении статей в журнал МНГ просим авторов соблюдать правила, изложенные в «Единых требованиях к рукописям, представляемым в биомедицинские журналы», принятых Международным комитетом редакторов медицинских журналов - http://www.spinesurgery.ru/files/publish.pdf и http://www.nlm.nih.gov/bsd/uniform_requirements.html
В конце каждой оригинальной статьи приводится библиографический список. В список литературы включаются все материалы, на которые имеются ссылки в тексте. Список составляется в алфавитном порядке и нумеруется. Литературный источник приводится на языке оригинала. В списке литературы сначала приводятся работы, написанные знаками грузинского алфавита, затем кириллицей и латиницей. Ссылки на цитируемые работы в тексте статьи даются в квадратных скобках в виде номера, соответствующего номеру данной работы в списке литературы. Большинство цитированных источников должны быть за последние 5-7 лет.

9. Для получения права на публикацию статья должна иметь от руководителя работы или учреждения визу и сопроводительное отношение, написанные или напечатанные на бланке и заверенные подписью и печатью.

10. В конце статьи должны быть подписи всех авторов, полностью приведены их фамилии, имена и отчества, указаны служебный и домашний номера телефонов и адреса или иные координаты. Количество авторов (соавторов) не должно превышать пяти человек.

11. Редакция оставляет за собой право сокращать и исправлять статьи. Корректура авторам не высылается, вся работа и сверка проводится по авторскому оригиналу.

12. Недопустимо направление в редакцию работ, представленных к печати в иных издательствах или опубликованных в других изданиях.

При нарушении указанных правил статьи не рассматриваются.

REQUIREMENTS

Please note, materials submitted to the Editorial Office Staff are supposed to meet the following requirements:

1. Articles must be provided with a double copy, in English or Russian languages and typed or compu-ter-printed on a single side of standard typing paper, with the left margin of **3** centimeters width, and **1.5** spacing between the lines, typeface - **Times New Roman** (**Cyrillic**), print size - **12** (referring to Georgian and Russian materials). With computer-printed texts please enclose a CD carrying the same file titled with Latin symbols.

2. Size of the article, including index and resume in English, Russian and Georgian languages must be at least 10 pages and not exceed the limit of 20 pages of typed or computer-printed text.

3. Submitted material must include a coverage of a topical subject, research methods, results, and review.

Authors of the scientific-research works must indicate the number of experimental biological species drawn in, list the employed methods of anesthetization and soporific means used during acute tests.

4. Articles must have a short (half page) abstract in English, Russian and Georgian (including the following sections: aim of study, material and methods, results and conclusions) and a list of key words.

5. Tables must be presented in an original typed or computer-printed form, instead of a photocopied version. **Numbers, totals, percentile data on the tables must coincide with those in the texts of the articles.** Tables and graphs must be headed.

6. Photographs are required to be contrasted and must be submitted with doubles. Please number each photograph with a pencil on its back, indicate author's name, title of the article (short version), and mark out its top and bottom parts. Drawings must be accurate, drafts and diagrams drawn in Indian ink (or black ink). Photocopies of the X-ray photographs must be presented in a positive image in **tiff format**.

Accurately numbered subtitles for each illustration must be listed on a separate sheet of paper. In the subtitles for the microphotographs please indicate the ocular and objective lens magnification power, method of coloring or impregnation of the microscopic sections (preparations).

7. Please indicate last names, first and middle initials of the native authors, present names and initials of the foreign authors in the transcription of the original language, enclose in parenthesis corresponding number under which the author is listed in the reference materials.

8. Please follow guidance offered to authors by The International Committee of Medical Journal Editors guidance in its Uniform Requirements for Manuscripts Submitted to Biomedical Journals publication available online at: http://www.nlm.nih.gov/bsd/uniform_requirements.html
http://www.icmje.org/urm_full.pdf
In GMN style for each work cited in the text, a bibliographic reference is given, and this is located at the end of the article under the title "References". All references cited in the text must be listed. The list of references should be arranged alphabetically and then numbered. References are numbered in the text [numbers in square brackets] and in the reference list and numbers are repeated throughout the text as needed. The bibliographic description is given in the language of publication (citations in Georgian script are followed by Cyrillic and Latin).

9. To obtain the rights of publication articles must be accompanied by a visa from the project instructor or the establishment, where the work has been performed, and a reference letter, both written or typed on a special signed form, certified by a stamp or a seal.

10. Articles must be signed by all of the authors at the end, and they must be provided with a list of full names, office and home phone numbers and addresses or other non-office locations where the authors could be reached. The number of the authors (co-authors) must not exceed the limit of 5 people.

11. Editorial Staff reserves the rights to cut down in size and correct the articles. Proof-sheets are not sent out to the authors. The entire editorial and collation work is performed according to the author's original text.

12. Sending in the works that have already been assigned to the press by other Editorial Staffs or have been printed by other publishers is not permissible.

Articles that Fail to Meet the Aforementioned Requirements are not Assigned to be Reviewed.

ავტორთა საყურადღებოდ!

რედაქციაში სტატიის წარმოდგენისას საჭიროა დავიცვათ შემდეგი წესები:

1. სტატია უნდა წარმოადგინოთ 2 ცალად, რუსულ ან ინგლისურ ენებზე,დაბეჭდილი სტანდარტული ფურცლის 1 გვერდზე, 3 სმ სიგანის მარცხენა ველისა და სტრიქონებს შორის 1,5 ინტერვალის დაცვით. გამოყენებული კომპიუტერული შრიფტი რუსულ და ინგლისურენოვან ტექსტებში - **Times New Roman (Кириллица)**, ხოლო ქართულენოვან ტექსტში საჭიროა გამოვიყენოთ **AcadNusx.** შრიფტის ზომა – 12. სტატიას თან უნდა ახლდეს CD სტატიით.

2. სტატიის მოცულობა არ უნდა შეადგენდეს 10 გვერდზე ნაკლებს და 20 გვერდზე მეტს ლიტერატურის სიის და რეზიუმეების (ინგლისურ, რუსულ და ქართულ ენებზე) ჩათვლით.

3. სტატიაში საჭიროა გაშუქდეს: საკითხის აქტუალობა; კვლევის მიზანი; საკვლევი მასალა და გამოყენებული მეთოდები; მიღებული შედეგები და მათი განსჯა. ექსპერიმენტული ხასიათის სტატიების წარმოდგენისას ავტორებმა უნდა მიუთითონ საექსპერიმენტო ცხოველების სახეობა და რაოდენობა; გაუტკივარებისა და დაძინების მეთოდები (მწვავე ცდების პირობებში).

4. სტატიას თან უნდა ახლდეს რეზიუმე ინგლისურ, რუსულ და ქართულ ენებზე არანაკლებ ნახევარი გვერდის მოცულობისა (სათაურის, ავტორების, დაწესებულების მითითებით და უნდა შეიცავდეს შემდეგ განყოფილებებს: მიზანი, მასალა და მეთოდები, შედეგები და დასკვნები; ტექსტუალური ნაწილი არ უნდა იყოს 15 სტრიქონზე ნაკლები) და საკვანძო სიტყვების ჩამონათვალი (key words).

5. ცხრილები საჭიროა წარმოადგინოთ ნაბეჭდი სახით. ყველა ციფრული, შემაჯამებელი და პროცენტული მონაცემები უნდა შეესაბამებოდეს ტექსტში მოყვანილს.

6. ფოტოსურათები უნდა იყოს კონტრასტული; სურათები, ნახაზები, დიაგრამები - დასათაურებული, დანომრილი და სათანადო ადგილას ჩასმული. რენტგენოგრამების ფოტოასლები წარმოადგინეთ პოზიტიური გამოსახულებით **tiff** ფორმატში. მიკროფოტოსურათების წარწერებში საჭიროა მიუთითოთ ოკულარის ან ობიექტივის საშუალებით გადიდების ხარისხი, ანათალების შეღებვის ან იმპრეგნაციის მეთოდი და აღნიშნოთ სურათის ზედა და ქვედა ნაწილები.

7. სამამულო ავტორების გვარები სტატიაში აღინიშნება ინიციალების თანდართვით, უცხოურისა – უცხოური ტრანსკრიპციით.

8. სტატიას თან უნდა ახლდეს ავტორის მიერ გამოყენებული სამამულო და უცხოური შრომების ბიბლიოგრაფიული სია (ბოლო 5-8 წლის სიღრმით). ანბანური წყობით წარმოდგენილ ბიბლიოგრაფიულ სიაში მიუთითეთ ჯერ სამამულო, შემდეგ უცხოელი ავტორები (გვარი, ინიციალები, სტატიის სათაური, ჟურნალის დასახელება, გამოცემის ადგილი, წელი, ჟურნალის №, პირველი და ბოლო გვერდები). მონოგრაფიის შემთხვევაში მიუთითეთ გამოცემის წელი, ადგილი და გვერდების საერთო რაოდენობა. ტექსტში კვადრატულ ფჩხილებში უნდა მიუთითოთ ავტორის შესაბამისი N ლიტერატურის სიის მიხედვით. მიზანშეწონილია, რომ ციტირებული წყაროების უმეტესი ნაწილი იყოს 5-6 წლის სიღრმის.

9. სტატიას თან უნდა ახლდეს: ა) დაწესებულების ან სამეცნიერო ხელმძღვანელის წარდგინება, დამოწმებული ხელმოწერითა და ბეჭდით; ბ) დარგის სპეციალისტის დამოწმებული რეცენზია, რომელშიც მითითებული იქნება საკითხის აქტუალობა, მასალის საკმაობა, მეთოდის სანდოობა, შედეგების სამეცნიერო-პრაქტიკული მნიშვნელობა.

10. სტატიის ბოლოს საჭიროა ყველა ავტორის ხელმოწერა, რომელთა რაოდენობა არ უნდა აღემატებოდეს 5-ს.

11. რედაქცია იტოვებს უფლებას შეასწოროს სტატია. ტექსტზე მუშაობა და შეჯერება ხდება საავტორო ორიგინალის მიხედვით.

12. დაუშვებელია რედაქციაში ისეთი სტატიის წარდგენა, რომელიც დასაბეჭდად წარდგენილი იყო სხვა რედაქციაში ან გამოქვეყნებული იყო სხვა გამოცემებში.

აღნიშნული წესების დარღვევის შემთხვევაში სტატიები არ განიხილება.

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DELAYED SURGERY FOR GIANT SPONTANEOUS RUPTURE OF THE DISTAL THORACIC AORTA CAUSED BY CYSTIC MEDIAL NECROSIS

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Abstract.

Spontaneous (idiopathic) thoracic aortic rupture (STAR) is uncommon and assumes a rupture of the normal-sized thoracic aorta with no visually apparent aortic disease. Since 1961 about fifty reports have been published. STAR is established in cases of thoracic normal-sized aorta rupture with no traumas, aneurysms / dissections, infection, inflammation, connective tissue diseases, aortic and adjacent organs tumors/metastases, previous surgery, and occurring during pregnancy and the peripartum. Atherosclerosis penetrated atherosclerotic ulcer, neurofibromatosis type I, peri- and postpartum estrogen-mediated elastin irregularities of the aortic media, and cystic medial necrosis (CMN) were identified as a cause of STAR when histopathological examinations were performed. A case of a 68-year-old man with giant STAR of the descending aorta in the background of CMN is reported here. The patient in terminal hemodynamic condition was successfully treated by delayed open surgery two weeks after the disease's onset.

Key words. Aortic rupture, spontaneous, thoracic aorta, acute aortic emergencies, Gsell-Erdheim's syndrome, cystic medial necrosis.

Introduction.

Spontaneous or idiopathic rupture of the thoracic aorta is a sudden rupture of the normal-sized aorta with no visually apparent aortic disease [1,2].

Spontaneous thoracic aortic rupture (STAR) is an uncommon event. Since 1961 up to 2006 only 31 cases have been reported [1]. It was found only 19 additional cases on Medline for the period between 2006 – 2023 (Table 1). Due to its high potential to be lethal, STAR demands to be treated urgently. Since only successful cases are usually published, it can be assumed that the real number of cases is an underestimate.

A case of a giant spontaneous rupture of the distal descending thoracic aorta, which occurred in the background of cystic medial necrosis is reported here. The patient in terminal hemodynamic condition was successfully treated by open surgery two weeks after the disease's onset.

Case report.

A 68-year-old man experienced an acute chest pain attack two weeks before admission to our hospital. In the small too remote hospital in the place where the patient lived, CT scans on the day of disease's onset revealed the normal-sized thoracic aorta with local lateral protrusion in the aortic contour of the distal segment of the descendent thoracic aorta (Figure 1A,1B and 1C). Thoracic aorta rupture was recognized and treated by immobilization, the maintenance of systolic blood pressure not above 100 mm Hg, transfusions of fresh frozen plasma / red cells and the drainage of the left chest cavity. There was no equipment and technical opportunities for endovascular or surgical procedural treatment in the hospital. The search for vascular centers and patient transportation took as long as two weeks. As a result, the patient was admitted to our hospital two weeks after disease onset in poor clinical condition with depressed consciousness, a weakened peripheral pulse, borderline hemodynamics, and limited urinary output. There was significant increasing of paraaortic hematoma and bilateral pleural effusion on the CT at the admission (Figure 1D). An endovascular approach was not considered due to the poor condition of the patient demanding circulatory support. Immediately, the left thoracotomy was performed via the 6th intercostal space with evacuation of 1500 ml bloody fluids. The descending thoracic aorta was surrounded by a thick layer of blood-imbibed paraaortic tissues. The aorta was cannulated just after the left subclavian artery branching. The main pulmonary artery was cannulated intrapericardially for venous return. The cardiopulmonary bypass (CPB) was initiated, and the patient's temperature decreased to19°C. The CPB volume was diminished to 0, 5 l/m2. The proximal aortic clamp was placed on the free aorta just distal to the cannulation site. The distal thoracic aorta was opened without clamping. The intima appeared non-atherosclerotic. The X-shaped rapture measured at 35 x 50 mm was identified on the lateral wall in accordance with the protrusion on the CT (Figure 2A). The ruptured segment was replaced with a 120mm-long linear prosthesis (Figure 2B). After aortic unclamping, the CPB was restored to 2.5L/m2. During re-warming, the heartbeat has been spontaneously restored. The aortic cross-clamping period was 53 minutes. The patient was weaned from CPB. The aortic specimen revealed changes coincident with cystic medial necrosis (CMN) (Figure 2C and 2D). The patient's recovery was prolonged due to acute renal failure without replacement therapy and gastroenteropathy demanded of entire parenteral nutrition during the two weeks after the procedure. At 8-months follow-up, the light weakness of legs persists, demanding the use of a walker to be insured.

Discussion.

The diagnosis of spontaneous or idiopathic thoracic aortic rupture assumes a sudden rupture of the normal-sized aorta with no apparent disease of the aortic wall and no external impacts [1]. The diagnosis is highly heterogeneous. In the published reports of open surgical treatment, the thoracic aortic ruptures were named as 'spontaneous' if there were no traumas, aneurysms /

Table 1. *Review of literature (2005 – April 2023): Spontaneous Rupture of the thoracic aorta.*

Author(Year)	Age/Sex	Comorbidity	Rupture site	Phatology	Operation	Outcome
Komanapalli CB(2006) [1]	40M	N/A	Distal arch	Fragmentation of elastica in media	Open surgery (GR)	Survived
Hirai S (2006) [17]	72M	N/A	Ascending	N/A	Open surgery (GR)	Survived
Gaspar M (2007) [5]	53M	Hypertension	Aortic arch	Atherosclerosis	Open surgery (GR)	Survived
Yoshida M (2008) [3]	61M	Takayasu's arteritis	Descending	N/A	Endovascular stent	Survived
Brizzio ME (2009) [4]	N/A	N/A	Ascending	N/A	Open surgery (GR)	Survived
Belov YuV (2011) [6]	67F	Diabetes Mellitus, type I	Descending	Penetrated atherosclerotic ulcer	Open surgery (PR)	Survived
Sasaki H (2012) [18]	75M	N/A	Aortic arch	N/A	Open surgery (GR)	Survived
Lu W (2012) [19]	51M	N/A	Descending-abdominal	N/A	Endovascular stent	Survived
Park YJ (2012) [11]	49M	Neurofibromatosis (NF-1)	Descending-abdominal	Fragmentation of elastica in media	Open surgery (GR)	Survived
Menon A (2013) [7]	17F	Postpartum	Descending	Fragmentation of elastica in media	Open surgery (GR)	Survived
Itonaga T (2014) [20]	67M	N/A	N/A	N/A	Endovascular stent	Survived
Akchurin RS (2015) [21]	67M	Hypertension	Distal arch	N/A	Endovascular stent	Survived
Yoshida C (2015) [13]	94F	N/A	Descending	N/A	Open surgery (N/A)	Survived
Furukawa T(2018) [9]	N/A	Postpartum	N/A	N/A	Endovascular stent	Survived
Tateishi A (2019) [12]	N/A	Peripartum, Neurofibromatosis (NF-1)	Ascending	N/A	Open surgery (N/A)	Survived
Guner EG (2020) [8]	22F	Peripartum	Ascending	Fragmentation of elastica in media	Open surgery (ARR)	Survived
Kasahara H(2021) [2]	76M	N/A	Aortic arch	N/A	Endovascular stent	Survived
Yokoyama T (2021) [10]	N/A	N/A	N/A	N/A	Endovascular stent	Survived
Yokoyama T (2021) [10]	N/A	N/A	N/A	N/A	Endovascular stent	Survived

M – male; F – female; N/A. – not available; GR – Graft replacement; PR – Patch repair; ARR - Aortic root repair.

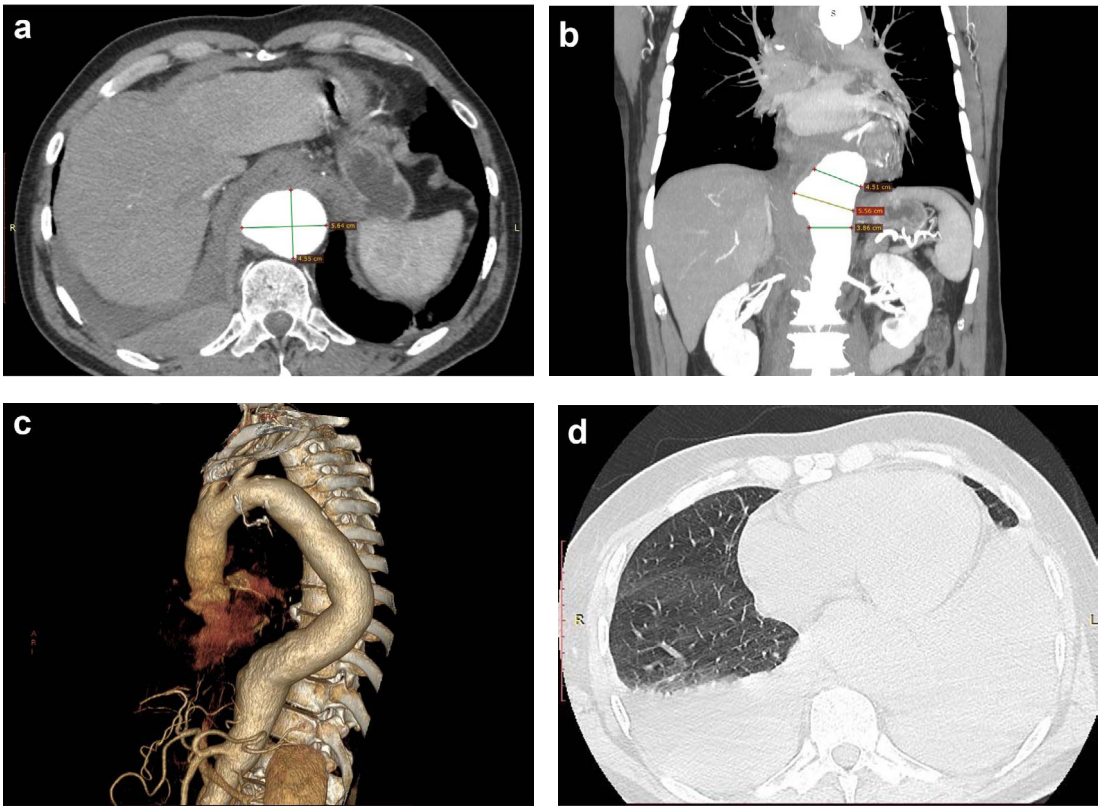

Figure 1. *(A) Axial, (B) Coronal contrast-enhanced CT at the onset of the disease; (C) 3D CT reconstruction; (D) Deferred axial CT scan after two weeks from disease onset.*

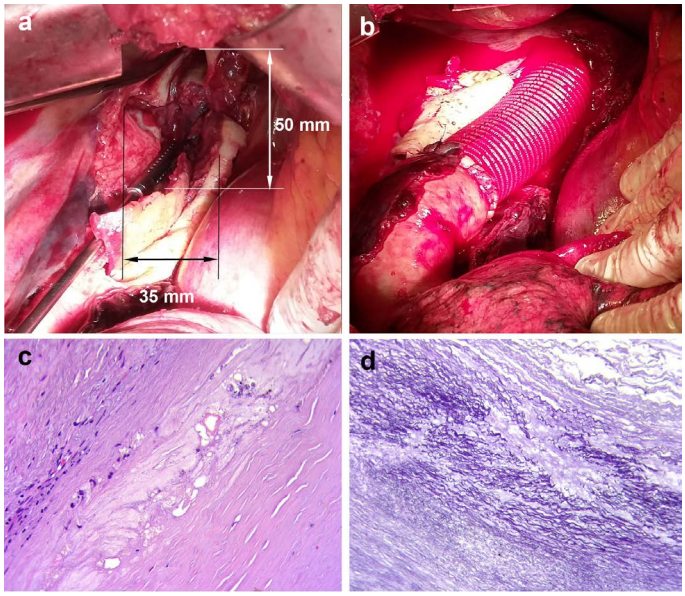

Figure 2. *(A) View of the rupture; (B) cyst-like lesions in the media, H&E×200; (C), elastic fibers disruptions, Weigert staining×100.*

dissections, infection, inflammation, connective tissue diseases, aortic and adjacent organs tumors/metastases [2,3], previous surgery [4] along a normal aortic size. Simultaneously, the penetrative atherosclerotic ulcer was indicated as a cause of aortic rupture also named as 'spontaneous' [5,6]. Aortic ruptures during pregnancy and the peripartum period because of progressive, estrogen-mediated elastin irregularities of the aortic media have also been reported as cases of STAR [7-9].

While endovascularly treated cases were reported with no histopathological examinations, the causes of the rupture were recognized in all surgically treated cases after specimen examination. Aortic wall abnormalities such as atherosclerosis [5,10], neurofibromatosis type I [11,12], and cystic medial necrosis were identified as a background in cases of STAR. There are no case reports on STAR with healthy aortic wall in histopathological examinations.

STAR is a rare event. Thirty-one cases treated surgically were founded to be reported from 1961 to 2004 [1,5]. The development of endovascular aortic repair has led to a growing number of reported cases of successful treatment within the last 15 years. Nineteen both surgically and endovascular cases have been discovered on Medline between 2005 and the present (Table 1). Since predominantly successful outcomes are usually published, the real number of cases may be underestimated.

Because of hemodynamic shock, prompt diagnosis and urgent intervention are necessary in the case of STAR [2]. CT is useful for prompt diagnosis, but it is sometimes difficult to detect the exact site of the lesion [2,10]. Any irregularity, ulcer-like projection, extravasation of contrast medium, prominence of aortic contour on CT may help to find the site of lesion. 3D reconstruction may also be useful [13] as in the case described here. The localization and extent of periaortic hematoma also helps to define the location of aortic rupture.

Treatment modalities include open surgical and endovascular techniques. Endovascular treatment has obvious advantages in comparison to open surgery due to the possibility of prompt use, less trauma, and no need for cardiopulmonary bypass. In conditions of extended rupture and absence of its exact localization, endovascular treatment demands a lengthened stent position with potential for spinal cord ischemia and associated paralysis of the lower extremities [10].

Because only singular or short serial cases with no long follow-up were published, it is not possible to describe STAR prognosis, so it is left obscured.

Cystic medial necrosis as well as initial lysell-Erdheim syndrome is a pathology of the aorta and great arteries characterized by degeneration of elastin fibers with loss of smooth muscle cells in the vascular medium that produces areas of glycosaminoglycans accumulation initially described as cysts. The term CMN is historic, since, in fact, no cysts or necrosis exist. The etiology is not well understood. There are two theories of CMN etiology: 1. CMN is a result of affecting on aortic wall of xenobiotic agents having excessive and growing environmental spread. 2. CMN is caused by high activity of tissue proteinases. The activity of tissue proteinases is regulated by numerous complex mechanisms including unrecognized. The result of both etiological pathways is degeneration of elastin fibers with loss of smooth muscle cells in the vascular medium with glycosaminoglycans accumulation [14]. This disease accounts for 0,09% of all sectional cases, occurs in 8.0–22.2% of patients with aortic dissection without Marfan syndrome and in 40–82.0% of patients with combined aortic dissection with Marfan syndrome [15]. The average age of patients ranges from 32 to 50 years, but cystic medionecrosis can be even in newborns and young children [15]. The correct prevalence of CMN is difficult to determine but is quite high as a background in connective tissue diseases such as Marfan syndrome, Ehlers-Danlos syndrome, annul aortic ectasia, and congenital heart defects [15]. In recent time, CMN is documented in unaffected individuals also [14].

The described case demonstrates that the strength of the mediastinal tissues restrained exsanguination in the pleural cavity for two weeks, making performing the delayed open surgery possible.

Open surgery demands the application of CPB. There is no distal aortic clamp in the case because of the vast paraaortic hematoma. Deep hypothermic perfusion was used to reduce blood flow. It may be supposed that the light weakness of the legs persisting for 8 months is caused by compression of the muscle innervation for a long time period in the borderline hemodynamic conditions.

Conclusion.

In conclusion, STAR is a sudden rupture of the normal-sized aorta with no apparent aortic disease. STAR occurs in patients with aortic wall background abnormalities. Only about fifty reports have been published in the literature up to the present. STAR is a life-threatening condition demanding urgent open or endovascular treatment. Giant aortic rupture here described has been presented with a long enough survival period due to the strength of the mediastinal tissues preventing exsanguination.

The delayed open surgery appeared successful despite poor initial clinical conditions of the patient.

The patient's consent was obtained to present the case and to provide images.

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