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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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DERMATOFIBROSARCOMA PROTUBERANS: WIDE LOCAL EXCISION AS DERMATOSURGICAL APPROACH WITH FAVOURABLE FINAL OUTCOME-CASE PRESENTATION AND SHORT UPDATE ON THERAPEUTIC OPTIONS

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Abstract.

Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade cutaneous sarcoma typically found on the proximal extremities and the trunk, characterized by infiltrative growth and low risk of metastasis. High rates of local recurrence or relatively large tumor sizes can significantly complicate therapeutic management, particularly when 1) surgical intervention is not adequately performed and /or 2 access to newer medications is limited or their high cost imposes a financial burden on patients.

We present the case of a 63-year-old male with a histologically confirmed dermatofibrosarcoma protuberans, measuring 6 cm in diameter, located on the right dorsal region, accompanied by several confluent multifocal nodules situated infralaterally to the primary formation.

Wide surgical excision with 5 mm margins in all directions was performed, achieving clean resection margins in all directions and short term recidive-free outcome.

This report also provides a brief overview of the therapeutic options available for DFSP, emphasizing the surgical approach, which remains the gold standard for treatment.

Key words. Dermatofibrosarcoma protuberans, dermatologic surgery, wide local excision, Mohs surgery, tumour recidive, skin cancer.

Introduction.

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing superficial neoplasm that typically arises on the proximal extremities and the trunk [1]. The condition is characterized by a high incidence of local recurrence and infiltrative behavior, whereas the risk of metastasis is comparatively low [1,2]. Surgical excision achieving negative margins remains the gold standard treatment for managing this condition [2]. However, radiation therapy and systemic treatment with tyrosine kinase inhibitors, such as imatinib mesylate, are considered alternative options when achieving negative surgical margins is not possible [2]. A brief overview will be conducted, focusing on articles available on PubMed, specifically addressing the treatment options for this condition, including their availability, advantages and limitations.

Case report.

A 63-year-old male presented to the dermatology department with a primary complaint of a chronic tumorous lesion on his back, which periodically bleeds upon contact. His medical history includes an appendectomy performed in 1980 and a diagnosis of chronic obstructive pulmonary disease (COPD). The patient is on systemic therapy with beclometasone dipropionate/ formoterol fumarate dihydrate/ glycopyrronium 87 microgram/5microgram/9microgram, taken twice daily, in the morning and evening, for the past year.

Additionally, the patient has been taking colchicine 0.5 mg, administered twice daily, once in the morning and once in the evening, also for the past year, due to osteoarthritis diagnosed about a year ago of both knee joints.

There was no reported family history of skin cancer among any of the patient's relatives.

The patient requested a physical examination and further therapeutic approach to be established.

The dermatological examination revealed a tumor formation in the right dorsal region, measuring approximately 6 cm in diameter (Figure 1a/1b). The lesion displayed a rounded shape with nodular growth, was mobile relative to the underlying subdermal structures, and exhibited a smooth surface. In the peripheral region of the formation, infralaterally, several confluent rounded nodules were observed, characterized by a pink to red color, smooth and regular surface, and palpable dense consistency (Figure 1a/1b). Lymph nodes were not palpable.

The patient was recommended surgical excision of the entire lesion with the simultaneously removal of the satellite lesions under local anesthesia. Routine blood tests were conducted, resulting without abnormalities. A chest X-ray and an abdominal ultrasound were performed, ruling out any metastatic or concurrent neoplastic process. The tumor formation in the right dorsal region was preoperatively marked (Figure 1b) and removed with a wide local excision, with a safety margin of at least 5 mm in 2 directions (Figure 2), but also about 3cm in the opposite directions. The resulting wound defect was subsequently closed using single interrupted sutures (Figure 3a,b). Daily wound dressings with povidone-iodine were applied, and suture removal was performed between the 10^{th}-12^{th} postoperative days. The postoperative period resulted without complications.

The histopathological examination revealed extensive mesenchymal neoplasia characterized by orthohyperkeratosis, interspersed with horizontally alternating parakeratotic squamous crusts overlaying areas of epidermal atrophy. These alternated with reticular acanthotic projections and ecstatically dilated capillary loops in the papillary dermis. The neoplasm exhibited a compact proliferation of collagen fibers of varying calibers, accompanied by fusiform atypical fibroblastic cells with centrally located nuclei containing two to three nucleoli. These cells were embedded in a well-vascularized stroma with foci of lymphoplasmocytic infiltration densely occupying the lower dermal layers and diffusely invading the hypodermis. Clean resection lines were achieved. The histopathological findings were consistent with dermatofibrosarcoma protuberans (Figures 4-6).

Registration for further monitoring at the regional oncology clinic was advised, along with a recommendation for outpatient PET-CT imaging.

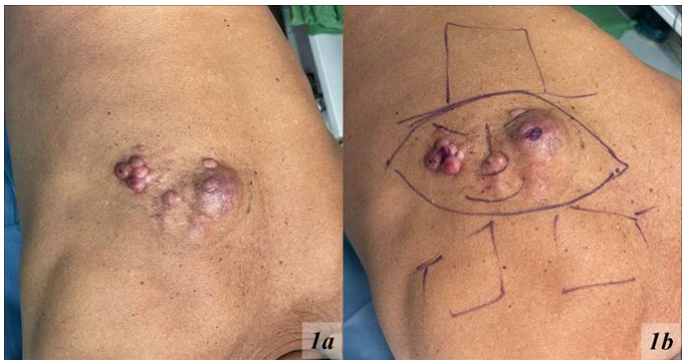

Figure 1a,b. *Preoperative panel:*

1a: *A tumor formation in the right dorsal region, measuring approximately 6 cm in diameter. The lesion has a rounded shape with nodular growth, relatively mobile to the underlying subdermal structures, and with a smooth surface. In the peripheral region of the formation, infralaterally, several confluent rounded nodules can be observed, characterized by a pink to red color, smooth and regular surface, and palpable dense consistency.*

1b: *Creative preoperative marking of the lesion that will be surgically excised.*

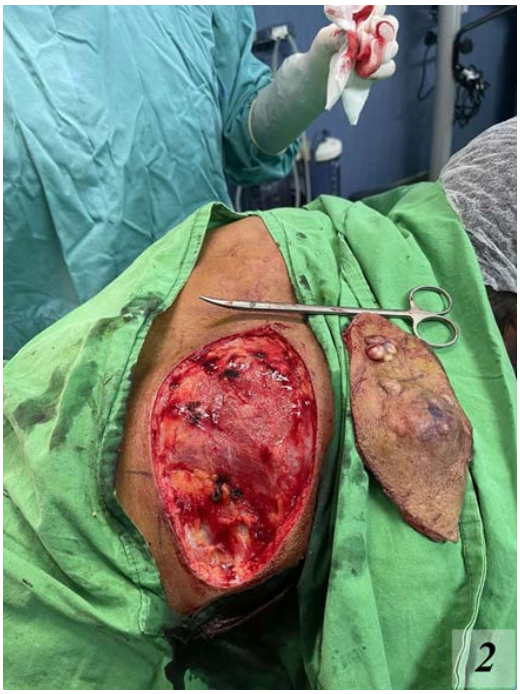

Figure 2. *Intraoperative view: Primary wound defect resulting from elliptical excision of the tumor formation with a 5 mm safety surgical margin in all directions.*

Figure 3a,b. *Postoperative view: The primary wound defect is closed with single interrupted sutures.*

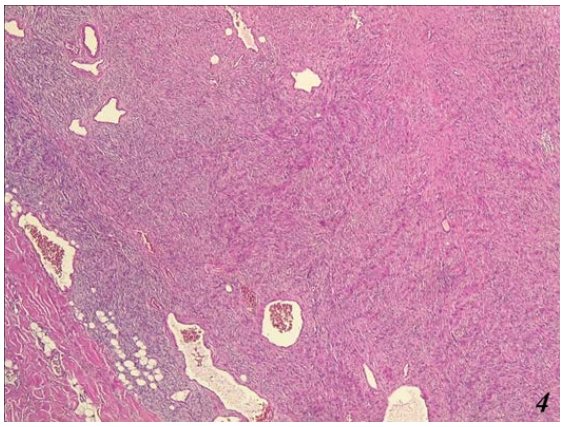

Figure 4. *DFS – prominent involvement in subcutis x HE x 40. Dermatofibrosarcoma protuberans: Extensive mesenchymal neoplasia characterized by orthohyperkeratosis, interspersed with horizontally alternating parakeratotic squamous crusts overlaying areas of epidermal atrophy. These alternated with reticular acanthotic projections and ecstatically dilated capillary loops in the papillary dermis. The neoplasm exhibit a compact proliferation of collagen fibers of varying calibers, accompanied by fusiform atypical fibroblastic cells with centrally located nuclei containing two to three nucleoli. These cells are embedded in a well-vascularized stroma with foci of lymphoplasmocytic infiltration densely occupying the lower dermal layers and diffusely invading the hypodermis.*

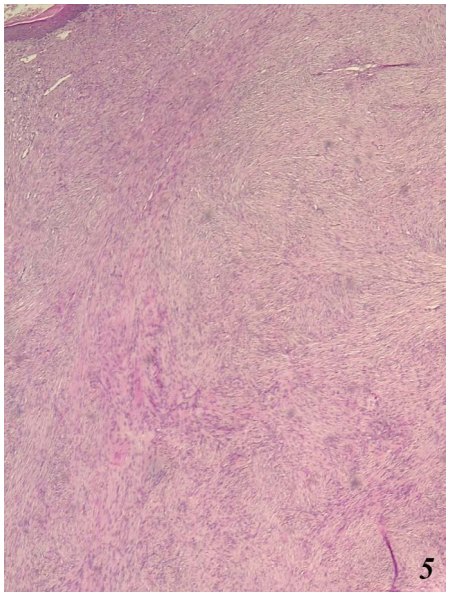

Figure 5. *DFSP x HE x 40.*

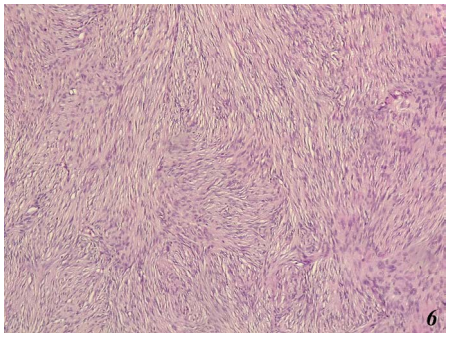

Figure 6. *DFSP x HE x 100.*

Discussion.

Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade cutaneous sarcoma that predominantly affects the trunk and extremities, with an annual incidence of 0.8 to 5 cases per million [3].

It is locally aggressive, exhibiting a significant tendency for local recurrence, yet metastasis is rare [4].

Tumors located in the head, face, and neck regions are associated with greater likelihood of recurrence compared to those on the extremities [3].

A retrospective analysis of 65 patients with 67 primary cutaneous sarcomas revealed that the head and neck region was the most common site of localization, with 44 patients exhibiting various types of sarcomas, including dermatofibrosarcoma protuberans [5].

Criscito et al. [6] conducted an extensive analysis of prognostic factors and the influence of treatment modalities on overall survival in patients with DFSP, examining 3686 individuals with histologically confirmed diagnoses of DFSP. Poorer overall survival was estimated in association with older age (hazard ratio [HR], 1.08; 95% CI, 1.06-1.10; P< .001), male sex (HR, 1.97; 95% CI, 1.09-3.55; P= .03), and tumor size (HR, 1.09; 95% CI, 1.01-1.18; P= .04) [6]. Interestingly, these same factors – older age (odds ratio [OR], 1.01; 95% CI, 1.00-1.02; P= .01) and male sex (OR, 1.95; 95% CI, 1.57-2.42; P < .001) - were also linked to larger tumors (>= 3.0 cm) at presentation [6]. Furthermore, receiving surgery combined with radiation, instead of surgery alone, was significantly associated with older age at presentation (OR, 1.02; 95% CI, 1.01-1.03; P = .01), large tumor size (OR, 1.15; 95% CI, 1.09-1.21; P < .001), and head and neck location (OR, 4.63; 95% CI, 2.66-8.07; P < .001) [6].

Fibrosarcomatous differentiation (DFSP-FS) is recognized as a significant risk factor associated with increased rates of local recurrence [3], metastasis, and mortality when compared to classic DFSP without fibrosarcomatous changes [7].

Liang et al. [7] conducted a comprehensive analysis of 1422 patients with DFSP and 225 with DFSP-FS. The study demonstrated substantially higher risks in the DFSP-FS group: local recurrence was observed in 29.8% of DFSP-FS cases compared to 13.7% in DFSP, with a risk ratio 2.2 [95% CI, 1.7-2.9]; metastasis occurred in 14.4% of DFSP-FS cases compared to 1.1% in DFSP, with a risk ratio of 5.5 [95% CI, 4.3-7.0]; and death from disease was found at 14.7% for DFSP-FS compared to 0.8% for DFSP, with a risk ratio of 6.2 [95% CI, 5.0-7.8]) [7].

Wide surgical excision with three-dimensional histologic margin control has been demonstrated to be an effective treatment option for DFSP, achieving outcomes without recurrence or metastatic spread [8].

However, recurrence rates following wide local excision can range from 0% to 41% [9]. Due to this variability, some authors are suggesting Mohs micrographic surgery as a treatment option [3]. A complete response was achieved with Mohs surgery in a 10-year-old child presenting with a DSFP plaque localized to the chest area [10].

Foroozan et al. [11] reported a significantly lower recurrence rate for DFSP with Mohs micrographic surgery, at 1.11% (95% CI, 0.025-6.03%), compared to a recurrence rate of 6.32% (95% CI, 3.19%-11.02%) following wide local excision. Despite these findings, Mohs micrographic surgery or similar surgical techniques were not recommended as first-line therapies for DFSP, especially in regions prone to recurrence [11].

A combination of conservative surgery and radiation therapy can achieve excellent local control in patients with dermatofibrosarcoma protuberans [12]. A retrospective analysis by Castle et al. [12] conducted 53 patients with DFSP, with a median tumor size of 4 cm (range: 1-25 cm), who were treated with surgery and either pre- or postoperative radiation therapy. The study reported an overall survival rate of 98% at both 5 and 10 years [12]. Local disease control and disease-free survival rates were reported as 98% and 93% at 5 and 10 years, respectively [12].

Understanding the molecular mechanisms underlying DFSP has led to the development of target therapies, such as imatinib mesylate [3].

This first systemic-line therapy can be useful in cases of recurrent, metastatic or unresectable tumors [3].

Wang et al. [13] conducted a retrospective analysis of 22 patients, including 10 with locally advanced and 12 with metastatic DFSP, who were treated with imatinib therapy.

The study found that 15 patients achieved partial remission, 6 achieved stable disease, and one developed resistance to the drug [13].

Both classic and fibrosarcomatous DFSP demonstrated similar responses to imatinib [13]. Additionally, 4 out of 10 patients with primarily unresectable DFSP underwent neoadjuvant therapy with imatinib before surgical resection [13].

However, tumor relapse may occur once systemic therapy is discontinued [3].

Sunitinib, a multi-targeted receptor tyrosine kinase inhibitor, presents a promising new approach for the treatment of metastatic and recurrent DFSP [3] (ClnicalTrials.gov identifier NCT00474994).

Pazopanib has shown a partial response in a patient with metastatic fibrosarcomatous dermatofibrosarcoma protuberans [14].

Regorafenib, a multikinase inhibitor, has demonstrated antitumor effect in non-adipocytic soft tissue sarcomas and has been shown to improve progression-free survival [15].

In cases of recurrent and infiltrative DSFP that are unresponsive to conventional therapeutic options - including postoperative radiation therapy - and refractory to treatment with 800 mg of imatinib, sorafenib may serve as a promising alternative treatment option [16].

Conclusion.

Although new therapies are emerging for the treatment of dermatofibrosarcoma protuberans, the gold standard for managing the condition remains the Mohs micrographic surgery or the so called wide surgical excision.

The key takeaway is that when diagnosed early, DFSP can often be effectively eliminated through early surgery alone (wide local surgery), without the need for additional treatment approaches.

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