

Synchronous Superficial Angiomyxoma without Carney-Complex- A Particularly Entity

Vidu Viorica^{1*}, Herlea Vlad², Mihai Dana³, Ceck Cristina⁴ and Batca Severina Andreea⁵

¹Senior Researcher, Surgeon, Department of General Surgery and Liver Transplantation, Fundeni Clinical Institute, Bucharest, Romania

²Pathologist, Department of Pathology, Fundeni Clinical Institute, Bucharest, Romania

³Dermatologist, Fundeni Clinical Institute, Bucharest, Romania

⁴Cardiologist, C.C. Iliescu Institute of Cardio-Vascular Diseases, Bucharest, Romania

⁵Resident of Surgery, Department of General surgery and liver transplantation, Fundeni Clinical Institute, Bucharest, Romania

***Corresponding Author:** Vidu Viorica, Senior Researcher, Surgeon, Department of General Surgery and Liver Transplantation, Fundeni Clinical Institute, Bucharest, Romania.

Received: January 20, 2020; **Published:** February 17, 2020

DOI: 10.31080/eccmc.2020.03.00170

Abstract

Superficial cutaneous angiomyxoma is a very rare, frequently noninvasive, benign mesenchymal neoplasia, which is part of a heterogeneous group of myxomatous soft tissue neoplasms, which share the ability to produce myeloid stroma, concomitantly with the proliferation of tumor cells of different origins.

Keywords: Superficial Angiomyxoma; Carney-Complex

Introduction

It is a relatively recent entity described in the medical literature, the first reporting belongs to Allen PW., *et al.* in 1988, of 28 cases of superficial angiomyxomas without the Carney complex [1].

Common locations are on the head, neck, trunk, and less frequently on the limbs [1,2].

It can be solitary or multiple, it is frequently associated with the Carney complex [2], synchronous peripheral (cutaneous) forms without the Carney complex being extremely rare.

Our study present the case of a patient with synchronous superficial (cutaneous) angiomyxomas of different sizes, the largest being located on the right arm and without associated Carney complex.

Case Report

Patient B.I. 58 years old, diagnosed with chronic hepatitis B, with antiviral treatment, is admitted to the Fundeni Clinical Institute in Bucharest with multiple pedicled skin tumors, the largest is around 4/3 cm on the back of the right arm (Figure 1), which has grown considerably in recent months, with polylobate nodular appearance, with tumor buds with erosive tendency on the surface and elastic consistency; other polypoid, synchronous pedicled tumors located: the largest is about 1/1 cm anterior thoracic (Figure 2) and others of millimetric dimensions, in the cervical (Figure 3) and axillary (Figure 4) regions.



Figure 1: Cutaneous superficial angiomyxoma of the arm posterior face: clinical aspect.



Figure 2: Synchronous cutaneous superficial angiomyxomas of the thorax: clinical aspect.

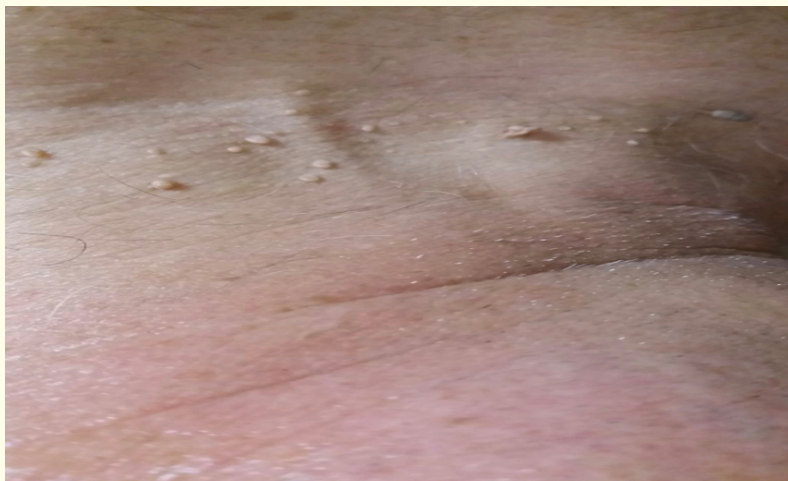


Figure 3: Synchronous cutaneous superficial angiomyxomas pediculated of the cervix area: clinical aspect.



Figure 4: Synchronous cutaneous superficial angiomyxomas pediculated of the axilla.clinical aspect.

The patient does not have the clinical components characteristic of Carney syndrome.

Serological tests and tumor markers (CEA, AFP, CA19.9) in normal limits.

Cardiological and echocardiographic examination were normal, without cardiac myxoma.

Under general anesthesia, extensive excision of the right arm skin tumor was performed (Figure 5), in depth to the superficial fascia of the arm and with resection margins within oncological safety limits, multiple excisions of the thoracic and cervical tumors below 1 cm and electrocauterisation. of the axillary and cervical for small lesions.



Figure 5: Cutaneous superficial angiomyxoma of the arm. Piece of tumoral resection.

Pathological examination of the excised pieces (two cutaneous tumors with the largest dimensions were analyzed, given the multitude of excised operative pieces, macroscopically identical), identifying polypoid vascular proliferation with small vessels and abundant intra-dermal and subepidermal myeloid stroma, without cell atypia, with diffuse inflammatory infiltrates in the myxoid stroma in both tumor pieces examined, the histopathological diagnosis being superficial cutaneous angiomyxoma in the arm (Figure 6). and anterior thorax (Figure 7).

Resection edges with preserved architecture. Immunohistochemistry: vimentin-positive and rare actinic and negative muscle fibers for CD34, desmin and S 100 protein.

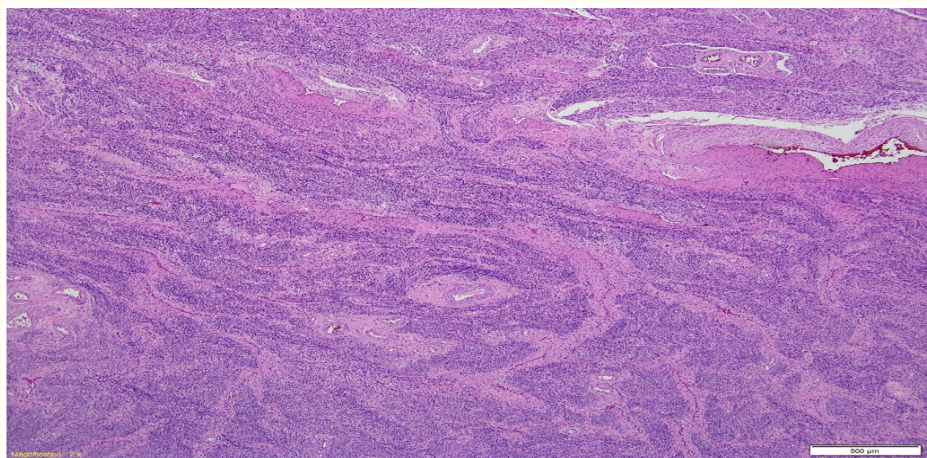


Figure 6: Pediculated cutaneous superficial angiomyxoma of the arm: pathological examination. Microscopic aspect (x40).

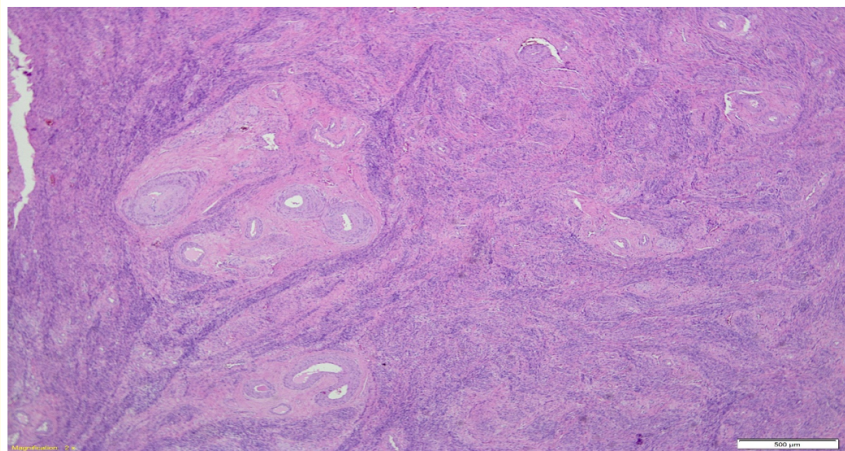


Figure 7: Pediculated cutaneous superficial angiomyxoma of the thorax: pathological examination. Microscopic aspect (x40).

Healing by primam.

Discussion

The term "superficial" cutaneous angiomyxoma was first used in 1989 by Calonge E., *et al.* [3], to define skin tumors with benign, non-invasive, peripheral evolution, as opposed to the aggressive ones, occurred with predilection in pelvic and in the female genital area [2].

In the English literature, the incidence is extremely low: 0.008% - 3% [4], more common in men (with a prevalence of 57%), than in women [5], at an average age of 36 - 45 years [6].

It is known that the most frequent locations are at the head and neck level, the first cases described in the literature being in the parotid region [1,6,7], less frequently in the limbs [6].

In "superficial" locations, angiomyxomas are known as noninvasive, with a benign structure, unique and very rarely multiple, as opposed to pelvic, perineal, genital locations, which are described as "aggressive", due to their local invasive potential and relapsing into incomplete resections [7-11]. Microscopic it is characterized by the proliferation of small vessels arranged in nests and poor cell architecture, but with abundant myxoid stroma and diffuse inflammatory infiltrate, considered by election for this neoplasm, less epithelial cells [1,2].

Also, it is known that "superficial" peripheral angiomyxoma, especially multiple forms, is frequently associated with the Carney complex, which is an autosomal dominant syndrome, which includes the existence of cardiac, cutaneous, mammary myxoid fibroadenomas, skin hyperpigmentation and endocrine hyperactivity (Cushing's syndrome, early sexuality, acromegaly) [2,6,7]; cases of angiomyxomas without Carney complex, especially synchronous forms, are extremely rare [3,12].

Thus, regarding our patient, there were multiple skin pedicles, different sized angiomyxomas, located on the thorax in the upper extremity, the lateral and anterior cervical region and in the axillary area (Figure 2 and 3), the largest being at the arm (Figure 1), which suggests that at this levels, synchronous neoplasia began - this being a particular clinical aspect in the described case. Although it is well

known that synchronous forms are associated with the Carney complex, however, our patient did not present the clinical features characteristic of this syndrome, which is another very rare feature.

The pathogenesis of these tumors is still incompletely elucidated. Some chromosomal mutations of the translocation type at 12q13-15 have been described, in the aggressive angiomyxoma invading a pelvis, vagina and bladder [10].

Hypothetically, in our case, synchronous tumors that are clinically in different stages of evolution, suggest the possibility of other microscopic cutaneous oncogenesis centers, so it may be an evolving multicenter neoplasia, with the risk of developing tumors as a result of neo-oncogenesis, more frequently than relapses, taking into account their location in the upper extremity of the body and the fact that all multiple tumor excisions were curative.

The paraffin histopathological diagnosis of the operative parts was defining: angiomatous proliferation of small and medium vessels, disposed subepidermally and intradermally, with few acicular or oval cells, grouped in nests, with abundant myeloid stroma, without atypia. The presence of the mixed inflammatory infiltrate, considered characteristic, which differentiates them from other types of myxoid tumors [3,8], is an aspect identified also in our case (Figure 6 and 7); In 20% of cases, epithelial structures of epidermoid cyst or squamous epithelium have been described [3].

Immunohistochemically, there are no specific markers, although, according to some opinions [2], only vimentin is constantly positive, and positive desmin represents an important differentiating factor between benign and aggressive evolutionary angiomyxomas, and the presence of stromal inflammatory infiltrate and the lack of atypia differentiate them from the other superficial myxoid tumors [4,13].

Sporadically, CD34, S-100 protein, muscle fibers, actin or pankeratin [3] may be identified, but future studies are needed to identify a specific immunohistochemical profile of these tumors.

Although they are considered rare and benign neoplasms as structure and evolution, differential diagnosis is very important because they can clinically and at imagistic examinations mimic external genital malignancies [9,10], a spermatocoele [13] or malignant bone tumors [14]. In this respect, the ultrasound examination can identify the vascular component, with positive Doppler signal [15,16], an aspect that differentiates them from the epidermal cysts [8]. For invasive and relapsing tumors in the pelvis and the soft side, nuclear magnetic resonance is beneficial.

The superficial cutaneous ones must be differentiated by the benign nevus, the myeloid cutaneous cysts, the vulgaris wart, the lipomas, the plexiform fibrocystic tumors [15,16], the solitary fibroma, the dendritic or malignant hamartoma, dermatofibrosarcoma protuberans, atypical fibroxanthoma and myxofibrosarcoma [17] or sarcoma with focal cutaneous mucinosis [18].

In our case, the differential diagnosis was difficult because the synchronous clinical forms are extremely rare. The dermatofibrosarcoma, which can be multicentric, was considered due to the large dimensions for the ones in the peripheral skin locations, which have made significant progress in recent months, the polylobate aspect with surface tumor buds, elastic consistency and multicentricity, liposarcoma, skin papillomas, lipomas, cell atypia and histopathological study were the defining ones for the diagnosis.

The treatment is surgical, the tumor excisions with cutaneous margins within the limits of oncological safety are sufficient.

Recurrences are possible in incomplete resections, on average after 18 months, in 30 - 40% of cases [1,3], especially in pelvic, perineal and genital locations, which have a significant local invading potential [9,11,13], although cases have been reported after 20 years [7].

For head and neck regions, recurrences can occur in 16% of cases [6]. According to some opinions [2], angiomyxomas with epithelial components in the structure have a higher relapse potential compared to those without epithelial components.

In selected cases of aggressive invasive angiomyxomas in the pelvis in women and in relapses with estrogen-receptor and progesterone-receptor-positive tumor cells, neoadjuvant, preoperative hormone therapy has been proposed to reduce tumor size or adjuvant in incomplete resections. These receptor-positive tumors may be candidates for gonadotropin-releasing hormone agonist (Gn-RH) therapy, considered a good therapeutic alternative in aggressive or relapsed tumors, in pelvic, perineal or genital locations, but with important side effects such as: depression, early menopause, long-term osteoporosis, but also with complete remission in some cases [19,20].

Chemotherapy and radiotherapy are considered ineffective in these tumors, due to their reduced mitotic activity, and intra- and preoperative radiotherapy have had questionable results [21].

For the "aggressive" pelvic angiomyxomas, due to the recurrence potential, clinical monitoring and in some cases serial interventions are required. It is not considered to be metastasizing tumors, although in the literature there have been reported 2 cases with pulmonary and mediastinal localization, both in women [22].

In our patient, although multiple excisions and focal electrocauterisation were curative at an early stage, and their locations were not "aggressive", but peripheral, however, they require long-term clinical monitoring, as clinical tumor synchronism suggests a possible multicenter neoplasm. It is still evolving, which may require future surgery.

Conclusion

Superficial skin angioma is a mesenchymal neoplasm of soft tissue, with very rare, single or multiple vascular and myxoid component, which is commonly associated with the Carney complex, especially in synchronous forms; clinical forms without Carney complex are very rare.

Our patient was diagnosed with multiple superficial cutaneous angiomyxomas, with different dimensions, located on the limbs, in the upper chest, axils and cervical region, clinically without associated Carney complex - extremely rare.

Particularly for this patient, although the apparent surgical treatment was curative, long-term resections may be required for new tumors, which are currently within microscopic dimensions and less likely for relapses.

Although "peripheral" skin angiomyxomas generally have a good prognosis, in this case the prognosis remains difficult to assess.

Conflict of Interest

The Authors have no conflicts of interest to declare.

Declaration of Patient Consent

The authors certify that they have obtained patient consent form for his clinical aspects and images to published and their name not published in the journal.

Bibliography

1. Allen PW, *et al.* "Superficial angiomyxomas with and without epithelial components. Report of 30 tumors in 28 patients". *The American Journal of Surgical Pathology* 12 (1988): 519-530.
2. Fetsch J, *et al.* "Superficial Angiomyxoma (*Cutaneous Myxoma*): A Clinicopathologic Study of 17 Cases Arising in the Genital Region". *International Journal of Gynecological Pathology* 16.4 (1997): 325-334.
3. Calonje E, *et al.* "Superficial Angiomyxoma: clinicopathologic analysis of a series of distinctive but poorly recognized cutaneous tumors with tendency for recurrence". *The American Journal of Surgical Pathology* 23 (1999): 910-917.

4. Ali N., *et al.* "Recurrence of a rare skin tumour Superficial angiomyxoma in the eyelid". *Canadian Journal of Ophthalmology* 46 (2011): 205-206.
5. Lee CU., *et al.* "Sonographic findings of prescrotal superficial angiomyxoma". *Japanese Journal of Radiology* 33 (2015): 216-219.
6. Rodriguez PR., *et al.* "Superficial Angiomyxoma of the Parotid Region and Review of the Literature". *Acta Otorrinolaringologica Española* 63.2 (2012): 147-149.
7. Allen PW. "Myxoma Is Not a Single Entity: A Review of Concept of Myxoma". *Annals of Diagnostic Pathology* 4.2 (2000): 99-123.
8. Wang YC., *et al.* "Superficial angiomyxoma of the penis A case report of a 6-year follow-up". *Asian Journal of Andrology* 19 (2017): 262-263.
9. Aydin AM., *et al.* "Long-standing aggressive angiomyxoma as paratesticular mass: A case report and review of literature". *SAGE Open Medical Case Reports* 5 (2017): 1-4.
10. Binta Patel and Hussain Ali. "Aggressive Angiomyxoma. A Rare Presentation". *Journal of Case Reports* 6.2 (2016): 157-160.
11. Gaunay GS., *et al.* "Aggressive angiomyxoma of the scrotum". *Clinical Imaging* 37.6 (2013): 1122-1124.
12. Yamamoto K., *et al.* "A Case of Superficial Angiomyxoma". *The Tokai Journal of Experimental And Clinical Medicine* 51.2 (2006): 43-45.
13. Mona Kafka., *et al.* "Incidental resection of a scrotal aggressive angiomyxoma mimicking a spermatocele: a case report". *European Surgery* 51 (2019): 31-33.
14. Bhat AK., *et al.* "Superficial angiomyxoma of the thumb mimicking a malignant bone tumor: Case report". *The Journal of Hand Surgery: American Volume* 39 (2014): 937-939.
15. Lee CU., *et al.* "Sonographic findings of prescrotal superficial angiomyxoma". *Japanese Journal of Radiology* 33 (2015): 216-219.
16. Lu Chen., *et al.* "Sonographic Findings in Superficial Angiomyxoma of the Vulva in a Perimenopausal Female". *Journal Medical Ultrasound* (2019).
17. Hongyu Y and Limin Y. "Cutaneous and Superficial Soft Tissue CD34+Spindle Cell Proliferation". *Archives of Pathology & Laboratory Medicine* 141 (2017): 1092-1100.
18. Wilk M., *et al.* "Cutaneous angiomyxoma: A benign neoplasm distinct from cutaneous focal mucinosis". *Journal of the American Academy of dermatology* 33.2 (1995): 352-355.
19. Bigby SM., *et al.* "Aggressive angiomyxoma. (Corrected) of the female genital tract and pelvis- clinicopathologic features with immunohistochemical analysis". *International Journal of Gynecological Pathology* 30.5 (2011): 505-513.
20. Shinohara N., *et al.* "Medical management of recurrent aggressive angiomyxoma with gonadotropin-releasing hormone agonist". *International Journal of Urology: Official Journal of The Japanese Urological Association* 11.6 (2004): 432-435.
21. Nyam D and Pemberton J. "Large aggressive angiomyxoma of the perineum and pelvis an alternative approach. Report of a case". *Diseases of the Colon and Rectum* 41.4 (1998): 514-516.
22. Blandamura S., *et al.* "Aggressive angiomyxoma: a second case of metastasis with patient's death". *Human Pathology* 34.10 (2003): 1072-1074.

Volume 3 Issue 3 March 2020

©All rights reserved by Vidu Viorica., et al.