Angiomatoid Fibrous Histiocytoma: Epidemiological, Anatomopathological, Clinical Profile, Treatment, and Outcomes in a Series of 15 Cases

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Histiocitoma Fibroso Angiomatoide: Perfil Epidemiológico, Anatomopatológico, Clínico, Tratamento e Resultados em uma Série de 15 Casos

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ABSTRACT

Introduction: Angiomatoid fibrous histiocytoma (AFH) is a rare soft-tissue neoplasm with intermediate behavior, originally described as a lesion occurring in the subcutaneous tissue of the extremities in young individuals. **Objective:** To strengthen the availability of pathological data and improve the clinical approach to this disease by healthcare providers. **Method:** A retrospective study analyzing 15 cases treated between 2000 and 2023, involving a review of medical records and an anatomopathological re-examination of stored slides. **Results:** The mean follow-up period was 7.16 years, and patient ages ranged from 5 to 44 years, with a predominance of females under 20 years old. The lesions were predominantly located in the limbs (60%). Surgical resection was the treatment of choice and the compromise of surgical margins was associated with risk of relapse, which was observed in five cases (33.3%), two of which resulted in death (mortality rate of 13.3%). Six patients (40%) received adjuvant therapy, two underwent margin widening with no subsequent recurrence. Metastases occurred in 27% of the patients, primarily affecting the lungs and lymph nodes. Oncological discharge was achieved in 66.6% of the cases. Among the patients who died, the mean survival time was 5.8 years. Histologically, the presence of characteristic cells in a syncytial arrangement, a rim of inflammatory cells, and pseudoangiomatoid spaces, along with immunohistochemical positivity for desmin, proved to be essential diagnostic features. **Conclusion:** Given the predominance of the disease in the limbs of young patients, the study highlights the importance of wide resection with clear margins to achieve best clinical outcome.

Key words: Histiocytoma, Malignant Fibrous; Retrospective Studies; Epidemiology.

RESUMO

Introdução: O histiocitoma fibroso angiomatoide (HFA) é uma neoplasia rara de partes moles, com comportamento intermediário, descrita a primeira vez como uma lesão que ocorria no subcutâneo das extremidades em jovens. **Objetivo:** Tornar mais robusta a disponibilidade de dados patológicos e aprimorar a abordagem dessa doença como prestadores de serviço de saúde. **Método:** Estudo retrospectivo que analisou 15 casos tratados entre 2000-2023, por meio de consulta dos prontuários e revisão anatomopatológica das lâminas armazenadas. **Resultados:** O tempo médio de acompanhamento foi de 7,16 anos e a idade variou de 5 a 44 anos, destacando-se menores de 20 anos e pacientes do sexo feminino. A localização foi predominantemente observada nos membros (60%). Quanto ao tratamento, houve preferência pela ressecção cirúrgica e o comprometimento de margens cirúrgicas foi associado ao risco de recidiva, observada em cinco casos (33,3%), dois destes evoluíram a óbito (13,3% de taxa de mortalidade). Seis pacientes (40%) receberam terapia adjuvante, sendo dois submetidos à ampliação de margem sem recidivas subsequentes. Metástases ocorreram em 27% dos pacientes, principalmente nos pulmões e linfonodos. Alta oncológica foi alcançada em 66,6% dos casos. Nos pacientes que evoluíram a óbito, a sobrevida média foi de 5,8 anos. Histologicamente, a presença de células típicas em conformação sincicial, o aro de células inflamatórias e espaços pseudoangiomatosos demonstram-se como características essenciais para o diagnóstico, bem como a positividade imuno-histoquímica da desmina. **Conclusão:** Dada a predominância nos membros de pacientes jovens, constatou-se a importância da ressecção ampla com margens livres para melhor evolução clínica.

Palavras-chave: Histiocitoma Fibroso Maligno; Estudos Retrospectivos; Epidemiologia.

RESUMEN

Introducción: El histiocitoma fibroso angiomatoide (HFA) es una neoplasia de tejidos blandos poco frecuente con comportamiento intermedio, descrita inicialmente como una lesión que se presentaba en el tejido subcutáneo de las extremidades en personas jóvenes. **Objetivo:** Aumentar la disponibilidad de datos patológicos y mejorar el enfoque de esta enfermedad por parte de los profesionales sanitarios. **Método:** Estudio retrospectivo que analizó 15 casos tratados entre 2000 y 2023, mediante la consulta de historias clínicas y la revisión anatomopatológica de láminas histológicas almacenadas. **Resultados:** El promedio del tiempo de seguimiento fue de 7,16 años y la edad osciló entre 5 y 44 años, con predominio de pacientes menores de 20 años y mujeres. La localización predominante fue en los miembros (60%). En cuanto al tratamiento, se empleó preferentemente la resección quirúrgica, y los márgenes quirúrgicos comprometidos se asociaron con el riesgo de recurrencia, observado en cinco casos (33,3%), de los cuales dos resultaron en muerte (mortalidad del 13,3%). Seis pacientes (40%) recibieron terapia adyuvante, y dos de ellos se sometieron a ampliación de margen sin recurrencia posterior. Se observaron metástasis en el 27% de los pacientes, principalmente en los pulmones y los ganglios linfáticos. El alta oncológica se logró en el 66,6% de los casos. En los pacientes que fallecieron, el promedio de supervivencia fue de 5,8 años. Histológicamente, la presencia de células típicas en conformación sincicial, el anillo de células inflamatorias y los espacios pseudoangiomatosos son características esenciales para el diagnóstico, al igual que la positividad inmunohistoquímica para desmina. **Conclusión:** Dado el predominio en los miembros de pacientes jóvenes, se confirmó la importancia de la resección amplia con márgenes libres para obtener mejores resultados clínicos.

Palabras clave: Histiocitoma Fibroso Maligno; Estudios Retrospectivos; Epidemiología.

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INTRODUCTION

Angiomatoid fibrous histiocytoma or fibro-histiocytoma angiomatoid (AFH) is a rare soft tissue tumor described initially by Enzinger in 1979 as an "angiomatoid malignant fibrous histiocytoma". Later, studies indicated that its nature was not totally malignant, currently classified by the World Health Organization (WHO) as a rare soft tissue tumor of uncertain differentiation that appear to be benign due to its macroscopy. Despite the ability of metastasizing, the term "malignant" was removed due to its low risk[1,2].

Epidemiologically, AFH affects mainly the extremities of children and young adults with a median age of 13 years within an age range of 7-65 years simulating a vascular neoplasia[2,3], predominantly as a slow-growth painless nodular subcutaneous tumor occurring in the lower extremities[1,3]. Nearly two-thirds of the cases occur in areas of lymph node chains (antecubital fossa, popliteal, axilla, inguinal and cervical), can manifest also in the ovary, vulva, peritoneum, lung, brain, bone, mediastinum and retroperitoneum[1,4]. The proportion men-women varies across the studies, but in some of them males predominate – 68%[5] – as in the study of Maqbool et al.[5].

Clinically and radiologically, it is similar to vascular tumors such as hemangioma, angiosarcoma or even hematoma. Magnetic resonance imaging (MRI) show T-1-weighted homogeneously hypointense lesions and T-2-weighted heterogeneously hyperintense lesions compatible with the aforementioned vascular lesions.

Costa and Weiss[3] demonstrate that surgical excision was the main therapeutic modality with high cure rate, similar to the findings of Maqbool et al.[5] in a study with 25 patients. Surgical approach should be performed with wide margins to ensure the removal of all tumor cells and minimize relapse. Recurrence was observed from 15% to 20% of the cases, with 80% surviving relapse-free while metastasis was rare - <1%, in addition of not being identified significant additional benefits with adjuvant therapies such as chemo or radiotherapy[4,5].

Macroscopically, AFH manifests as unspecified masses, its regular presentation is a circumscribed nodular lesion with solid white-to-yellow component and blood-filled cystic spaces, its most characteristic aspect[4,6]. The color can be brownish according to the quantity of hemosiderin, their limits can rarely be infiltrative[7,8].

Microscopically, AFH presents three essential morphologic components and one immunohistochemical component for the diagnosis according to WHO[1]:

- Syncytial-like nodules composed of ovoid or epithelioid cells with eosinophilic cytoplasm, vesicular nuclei and discreet atypia.
- Blood-filled vascular spaces lined by tumoral cells, the pseudoangiomatous.
- Rim of mononuclear inflammatory cells, such as lymphocytes or plasma cells with germinal centers, mimicking a lymph node metastasis.
- Immunohistochemistry markers utilizing desmin, EMA and/or CD99[6].

In addition, a compatible clinical presentation with small and painless subcutaneous nodulation is also considered an essential criteria[1]. The presence of all the findings simultaneously is not necessary or expected to confirm the diagnosis. The presence of a pseudocapsule with deposits of hemosiderin is a typical finding[1,4]. Regardless of being listed as essential, the peritumoral infiltrate and pseudoangiomatous spaces can be absent, therefore, it is possible that an AFH is totally solid. Older lesions exhibit extensive fibrous areas with deposits of hemosiderin and rare islets of neoplastic cells[4]. The identification of pleomorphic cells, the presentation as small, round, blue cells neoplasia and the presence of myxoid area are rare characteristics[1,4,5].

Further to the variable positiveness for desmin, EMA and CD99 – one of which should be identified – there is variable staining for CD68, calponin and caldesmon[1,4,8]. The negativity for vascular markers as CD34 and CD31, in addition to MYOD1 and myogenin are useful to rule out differential diagnoses[1,4,8].

AFH molecular studies are necessary for diagnostic confirmation. The most frequent genetic alterations involve the formation of fusion genes as the translocation t(2;22)(q33;q12), that creates fusion *EWSR1-CREB1* and occurs in more than 90% of the cases, the translocation t(12;22)(q12;q12), responsible for the fusion *EWSR1-ATF1*, is the second most common and some cases are related to the fusion *FUS-ATF1*[1,4,6].

Given its low incidence, few reports and reviews are found in the literature, which reinforces the importance of discussing the theme, most of all in view of the national scenario.

The present study presents a series of 15 patients treated in the National Cancer Institute (INCA) in order to share experiences, make the availability of pathology data more robust and improve the approach to this disease by healthcare providers.

METHOD

Observational, retrospective cohort study of patients diagnosed with localized AFH enrolled in an oncological reference center who received some type of treatment between October 2000 and January 2023 at INCA's department of connective-bone tissue (TOC). The

clinical data were extracted from medical charts. INCA's pathologists and study participants reviewed the diagnosis. Cases where anatomopathological evaluations between the biopsy material and the surgical piece presented discrepancies were excluded in addition to those with insufficient clinical documentations. The data of the cohort were analyzed in a descriptive manner. For discreet variables, proportions were utilized (%) and for continuous variables, mean, median and standard-deviation have been calculated.

INCA's Ethics Committee approved the study, report number 4501050 (CAAE (submission for ethical review): 40424120.6.0000.5274) in compliance with Directive 466/2012[9] of the National Health Council.

RESULTS

15 predominantly female patients diagnosed with AFH, accounting for nine of the cases (60%) were enrolled, eight of which claimed they were Brown (53.3%). Follow-up ranged from 2.6 and 18.4 years, with mean of 7.16 years. When surgical procedure was performed, the age-range varied between 5 and 44 years, standing out younger than 20 years, representing 80% of the cases (12).

Limbs were the most affected in nine cases (60%), most of all the lower limbs with six patients (40%). 13.3% of the cases submitted to surgery presented compromised surgical margins after the first procedure and surgical margin-widening was performed in 40% of the patients. Among adjuvant therapies, radiotherapy was applied in five (33.3%) of the patients while chemotherapy was utilized in two (13.3%) both as neoadjuvant and adjuvant (Table 1).

Local relapse was found in 33.3% of the patients and metastases, in 26.6% , being lungs and lymph nodes the most affected. 13.3% died due to the disease progression as portrayed in Table 2.

Macroscopic anatomopathological evaluation was feasible in 12 cases (Table 3). Circumscribed margins were observed in 58% of the macroscopic reports while staining was described as white and yellow in 75%, brownish in 17% and greyish in 8%. Cystic formations with blood content were cited in 33% of the cases and solid lesions in 66%. One case presented myxoid/gelatinous aspect.

15 reports have been evaluated microscopically and in all of them, nodules of typical cells in a syncytial arrangement were present (Figure 1.A) and microscopic capsule (Figure 1.B). A rim of inflammatory cells was noticed in 87%, 73% with pseudoangiomatous spaces (Figure 1.C). Cicatricial fibrosis was present in 67% and 27% manifested as solid lesions without pseudoangiomatous spaces. Two cases presented predominantly solid spindle cell pattern (Figure 1.D) and one solid with myxoid areas (Figure 1.E). Morphologic signs suggestive of malignancy were not identified in any case. All the cases are presented in Table 3.

In immunohistochemistry, desmin was positive for 90% of the AFH (9/10), while the other essential markers, EMA and CD99, presented positivity of 67% (4/6) and 50% (2/4) respectively. CD68, non-essential, was positive in 89% (8/9), and the highest diffuse staining index (75%, 6/8). Other findings were focal positivity for angiomyolipoma in 29% (2/7) and CD31 in 33% (1/3).

DISCUSSION

The 15-case sample presented mean follow-up of 7.16 years, ranging from 2.67 to 18.42 and revealed that 80% of the patients were younger than 20 years old, predominantly in the age range of 10-20 years, representing 46.7% of the cases, which corroborates findings of the literature[5,7,10].

Females predominated (60%) in comparison with classical articles, but similar to recent publications[11,12].

For the 11 patients submitted to incisional biopsy, 36.4% relapsed and 18.2 metastasized (two patients died). Of the four patients submitted to excisional biopsy, only one (25.0%) relapsed while 50% metastasized and none of them died.

These data suggest that excisional biopsy offers a more complete diagnostic approach and possibly a clearer conformation of the tumor, optimizing adjuvancy and approach of remote lesions. The differences found were most likely influenced by the tumor's intrinsic factors such as size, location and biologic aggressiveness that determine the type of biopsy for each case, since excisional biopsy can be preferred for small easy-to-resect lesions while the incisional can be utilized in larger tumors or complex locations.

Similar to previous publications, the analysis of the tumor location indicated that 60% of the tumors occurred mainly in the limbs[1,3,11,12]. It is believed that this fact associated with the anatomic functional structure and the intent to preserve the function and of the limb itself mirrored the necessity of widening the margin in six of the 15 cases.

There were no significant differences regarding the reported race in comparison with the general Brazilian demographic profile or proportion thereof or in terms of incidence or association of severe events such as metastases, relapses and death.

Wide local excision was performed in 40% of the patients, usually in situations where the margins were compromised in frozen section in the first procedure or, in some cases, where the initial resection was performed out of INCA. Other studies did not address this procedure[11-15].

Table 1. Distribution of the patients according to clinical data

Clinical data	Indicator//Subgroup	n (%)
Length of follow-up	Minimum time	2.67 years
	Maximum time	18.42 years
	Mean time	7.16 years
Sex	Female	9 (60%)
	Male	6 (40%)
Reported race	White	4 (26.7%)
	Black	3 (26.7%)
	Brown	8 (53.3)
Age	< 10 years	5 (33.3%)
	10-20 years	7 (46.7%)
	> 30 years	3 (20%)
Location of the tumor	Shoulder	2 (13.3%)
	Thigh	3 (20%)
	Leg	3 (20%)
	Thorax	1 (6.7%)
	Axillary	1 (6.7%)
	Dorsum	1 (6.7%)
	Head	1 (6.7%)
	Abdomen	1 (6.7%)
	Inguinal	1 (6.7%)
	Elbow	1 (6.7%)
Adjuvancy	Only chemotherapy	1 (6.7%)
	Only radiotherapy	4 (26.6%)
	Chemo + radiotherapy	1 (6.7%)
	Total with adjuvancy	6 (42%)
Surgical margin	Free	13 (86.7%)
	Compromised	2 (13.3%)
Relapse	Yes	5 (33.3%)
	No	10 (66.7%)
Metastasis	Yes	4 (26.7%)
	No	11 (73.3%)
Death	Progression of the disease	2 (13.3%)
	Other causes	2 (13.3%)
	Oncologic discharge	11 (73.3%)
Total cases	-	15 (100%)

Even with 80% of the cases with free-margins, the patients with compromised margins (cases 2 and 8) presented higher risk and relapsed, which reinforces the importance of proper surgical monitoring. The local relapse rate was 33.3% of the patients who submitted to margin-widening with compromised margins.

Interestingly, within the scenario of disease spread, case 7 presented lung implants of a primary lesion originated from the axillary lesion, the patient was submitted to surgical resection without adjuvancy, surviving for eight years and died of meningitis unrelated to the progression of AFH.

The data related to adjuvant therapies showed that 40% of the patients submitted to complementary treatments, predominantly radiotherapy. Likely, this therapeutic choice is associated with the presence of risk factors for relapse such as compromised margins or previous relapse. Among the six cases submitted to margin-widening, two did not relapse and in one of them, adjuvant chemotherapy was applied. Of the four cases who relapsed, only one underwent neo and adjuvant therapy (case 2) and evolved to death.

The available literature on the implementation of systemic treatments is scarce, but some publications present positive experiences of chemotherapy in patients who initially would not be eligible to surgery, the golden standard[16,17]. In that sense, it is interesting to mention case 3, a patient registered at the institution with lung and lymph node metastases. He survived for nine years after surgical resection of tumors on the thigh, pulmonary and inguinal lesions associated with neoadjuvant chemotherapy and adjuvant radiotherapy.

Metastatic spread was observed in four cases (27%), in two cases (50%) on the lungs and lymph nodes, the main site of secondary implants.

Remarkably, the analysis of the outcome revealed that only two of the 15 cases evolved to death directly associated with tumor progression (cases 2 and 3), a death rate of 13.3%. The other two cases interrupted the follow-up post death unrelated to histiocitoma (breast cancer and meningitis, respectively). Oncologic discharge (last visit) was achieved by 66.6% of the patients (n = 10), indicating a favorable global evolution in most of the cases. One case is still in oncologic follow-up (case 4, with 2.6 years of follow-up).

CONCLUSION

The present study is consistent with the epidemiologic profile of age and location of AFH found in early publications, with prevalence of cases of lesions on the limbs and in young individuals in contrast with the data of sex with high number of women in this series.

It is reinforced the importance of surgical limits. Although 80% of the cases have been submitted to free-margins resection, those with compromised margins relapsed more frequently and demanded therapeutic procedures. This aspect was definitive for the evolution because the two cases of death had surgical limits compromised and/or necessity of margin-widening and also relapse.

Table 2. Clinical data collected and individual cases

Age	Location	Surgery date	Limit	Margin widening	Relapse	Chemotherapy	Radiotherapy	Metastasis	Death	Last visit	Type of biopsy
17	Shoulder	10/18/2000	Free	N	N	N	Adjuvant	N	N	3/21//2019	Incisional
19	Thigh	10/30/2002	Compromised	N	Y	N	N	Retroperitoneum	9/6/2008	X	Incisional
14	Thigh	09/19/2003	Free	Y	Y	Neo and adjuvant	Adjuvant	Lymph mode, pulmonary and musculoskeletal	9/23/2012	X	Incisional
35	Leg	01/10/2023	Free	N	N	N	Adjuvant	N	N	11/28/2014	Incisional
39	Thigh	08/10/2007	Free	N	N	N	Adjuvant	N	13/12/2010 breast cancer	X	Incisional
8	Thorax	11/01/2006	Free	N	N	N	N	Linfonodal	N	6/15//2016	Excisional
8	Axillary	05/01/2006	Free	N	N	N	N	Pulmonary	2015 - 2010 meningitis	X	Excisional
12	Leg	05/29/2008	Compromised	Y	Y	N	N	N	N	7/3/2016	Excisional
13	Leg	06/17/2010	Free	Y	Y	N	N	N	N	3/11/2016	Incisional
11	Shoulder	12/15/2010	Free	Y	N	N	N	N	N	6/26/2015	Excisional
17	Dorsum	01/11/2013	Free	N	N	N	N	N	N	3/12/2020	Incisional
5	Head	07/13/2018	Free	Y	N	Adjuvant	N	N	N	12/17/2023	Incisional
44	Abdomen	07/30/2020	Free	N	N	N	Adjuvant	N	N	4/3/2023	Incisional
6	Inguinal	12/08/2021	Free	N	N	N	N	N	N	11/15/2024	Incisional
8	Elbow	12/26/2020	Free	Y	Y	N	N	N	N	1/13/2025	Incisional

Table 3. Macroscopy and microscopy characteristics

Characteristic	Description	n (%)
Macroscopy	Circumscribed margins	7 (58.3%)
	White-to-yellow color	9 (75%)
	Brownish color	2 (17%)
	Greyish color	1 (8.4%)
	Solid lesions	7 (58.3%)
	Blood-filled cystic formations	4 (33.3%)
	Myxoid/gelatinous aspect	1 (8.4%)
Microscopy	Nodes of typical cells in syncytial conformation	15 (100%)
	Microcapsule (non-essential)	15 (100%)
	Rim of inflammatory cells	13 (87%)
	Pseudoangiomatous spaces	11 (73%)
	Areas of perinodular fibrosis	10 (67%)
	Solid lesion (without pseudoangiomatous space)	4 (27%)
	Cytologic pleomorphism	4 (27%)
	Fusiform growth pattern with xanthomatous macrophages	1 (7%)
	Solid tumor with myxoid areas	1 (7%)

The mean follow-up time of 7.16 years and the death rate of 13.3% indicate the efficacy of the multidisciplinary management of local control of the disease. It is remarkable the role of adjuvant therapy where even in patients with metastatic disease, survival time was higher than described for soft tissues neoplasms in general.

The anatomopathological findings concur with previous articles regardless of the variability of immunohistochemistry markers. It is expected a harmonization of histopathological reports of AFH after the most recent edition of WHO's tumor classification.

Future studies with larger samples and multivariate analyzes are essential to identify with great accuracy the risk factors influencing the potential aggressiveness of this rare tumor and the associations with anatomopathological characteristics and usefulness of adjuvancy.

CONTRIBUTIONS

All the authors contributed substantially to the study conception and design, acquisition, analysis and interpretation of the data, writing and critical review. They approved the final version for publication.

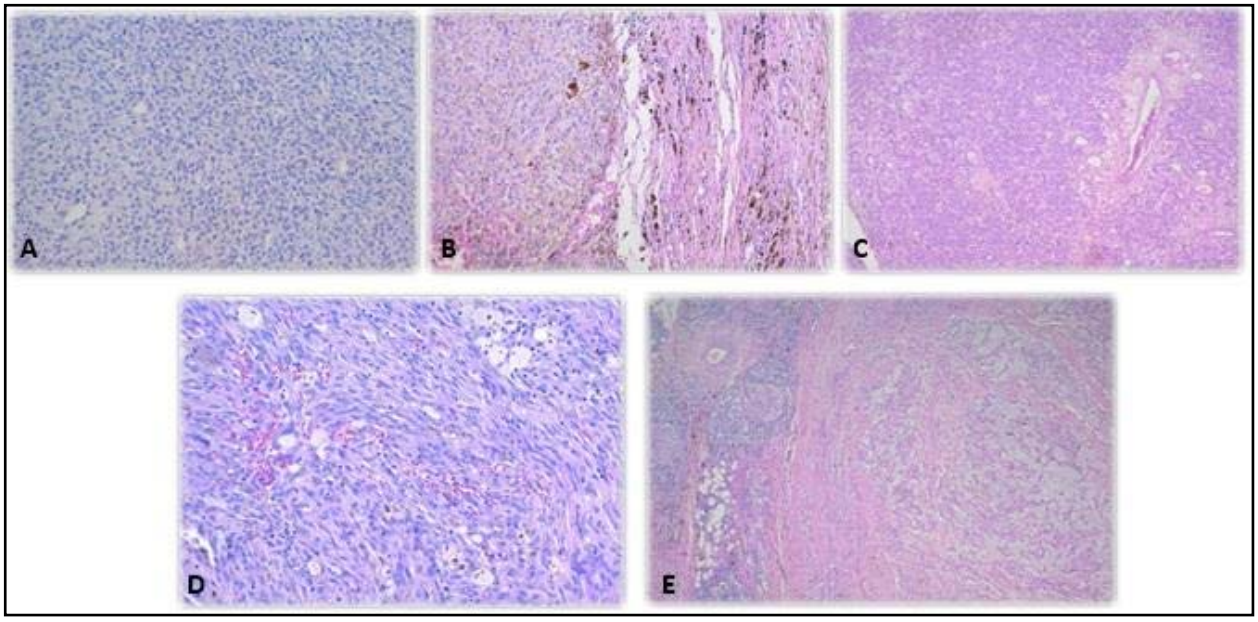

Figure 1. A – Nodules of typical cells in a syncytial arrangement; **B –** Microcapsule; **C –** Pseudoangiomatous spaces; **D –** Predominantly solid fusiform grow pattern; **E –** Myxoid areas

DECLARATION OF USE OF AI

AI was utilized to support the drafting of the manuscript.

DECLARATION OF CONFLICT OF INTEREST

There is no conflict of interest to declare.

DATA AVAILABILITY STATEMENT

Data should be requested to the corresponding author by e-mail to protect the anonymity of the patients.

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