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Breast Diffuse Neurofibroma: Case Report

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Abstract: *Neurofibroma are considered to be the most common nerve sheath tumor. These tumors can occur either sporadically or be related to neurofibromatosis. Neurofibromas rarely occur in the breast, and have a predilection for the nipple-areola complex. The localized variant of neurofibroma is the most common form. We hereby report the case of a 29 years old female patient, presenting for a left areolar diffuse neurofibroma, with no association to neurofibromatosis.*

Introduction

Neurofibromas (NF) are considered to be the most common nerve sheath tumor. [1]

Neurofibromas most often occur in a sporadic manner. Meanwhile, they are encountered in 95% of neurofibromatosis1 patients.

Although the nipple-areola is the most frequent location for breast neurofibromas, few case reports can be found inthe English literature, and cases of breast neurofibromas that are not related to neurofibromatosis are even rarer. [2,3]

We hereby report the case of a 29 years old female patient, with a left areolar diffuse neurofibroma with noassociation to neurofibromatosis type 1.

Patient and observation

We report the case of a 29 years old female patient, gravida 3, para 3, with no family or personal history of breastcancer. She presented for an important diffuse thickening of the left areola and nipple. She reports a progressive enlargement of her breast since her young age, especially worsening in relation to periods of breast-feeding. She reports No associated pain, no blood or purulent discharge.

Clinical examination found a large 16cm soft and diffuse thickening of the areolar and nipple region, inducing a remarkable disfiguration of the breast. The lesion was slightly pigmented, with no ulceration or bleeding. (Figure 1)

An ultrasound was performed revealing a highly vascular and hypoechogenic nature of the lesion. The lesion appearedto be ill-circumscribed with no infiltration of the deep mammary tissue.

A contrast-enhanced magnetic resonance (MR) of the breast was also performed, revealing an ill-circumscribed hypo-intense lesion in T1 weighted sequence. The lesion showed heterogeneously high T2 signal intensity and an heterogenous hypersignal after gadolinium administration.

a core biopsy of the lesion was performed. The pathological examination revealed presence of loose, hypocellular spindle cell proliferation made of Schwann cells and fibroblasts. The stroma was loose and contained many mast cells.No mitoses or atypia were identified. A diagnosis of neurofibroma was established.

The patient has undergone a resection of the tumor, followed by reconstructive surgery. (Figure 2)

The resected specimen was received in the pathology laboratory. Microscopic examination showed a similar proliferation, as observed on the core biopsy. However, on the resection specimen, we could appreciate the ill- circumscribed and the focal infiltrative nature of the lesion. This proliferation was often entrapping lactiferous ductsand showed Meissner corpuscles. (Figures 3, 4 and 5)

After 9 months of follow up, no complications have been identified and no lesions suggestive of neurofibromatosishave appeared.

Discussion

Neurofibroma (NF) is considered to be the most common nerve sheath tumor. NFs can occur either sporadically, or berelated to neurofibromatosis type I (von Recklinghausen's disease). [1] They usually manifest after adolescence, in a younger age group when compared to schwannoma. [1]

NFs present as a polypoid or nodular, soft brownish or fleshcoloured lesions, and are frequently cutaneous and maydevelop anywhere in the tegument.

When diagnosing a NF, it is important to examine carefully the patient for other signs of neurofibromatosis. [1]

In our case, no other signs favoring neurofibromatosis has been found, and the patient is currently under follow-up.

NFs are known for their benign behavior although the plexiform variant have the highest tendency to undergo malignant transformation. NFs have a high rate of local recurrence. [3]

Histologically, NFs have a typical appearance, made of a well-circumscribed, unencapsulated dermal or subcutaneouslesion. A loose proliferation of spindle cells is observed. The cytoplasm is pale and scanty. The nuclei are elongated and wavy. The stroma is fibrillar, collagenous and sometimes myxoid. Sometimes, S-100 negative/CD34 positive multinucleated floret-like giant cells can be observed. [1]

The stroma classically contains mast cells, and shows no biphasic appearance as observed in schwannoma.

Immunohistochemically, expression of S-100 protein is observed in 30 to 50% of cells. Expression of CD34 and EMA isvariable. [1]

Ultrastructural evaluation of NFs shows an admixture of Schwann cells, fibroblasts and perineurial cells. [1]

Many NF variants have been described. Among these, the diffuse variant can be related to neurofibromatosis type I inup to 30% of cases. It generally occurs on the head, neck or trunk. [1] It presents clinically as an ill-defined area of subcutaneous thickening, as was the case for our patient, whereas the conventional form of NF tends to present as appendices of the nipple, mimicking an accessory nipple or molluscum pendulum. [4] Histologically, it is characterizedby a diffuse infiltrative growth, with a collagenous stroma. This variant classically presents many Meissnerian corpuscles, as found in our case. [1]

Neurofibromatosis type 1 is the most frequent genodermatosis. It is a dominant genetic disorder, occurring secondaryto an alteration of the NF1 gene expression. The NF1 gene is located on chromosome 17 and has a tumor suppressorfunction, which explains the occurrence of numerous benign and malignant tumors when its function is lost. [5]

Neurofibromas are encountered in 95% of neurofibromatosis 1 patient and are therefore considered a hallmark of thisentity.

The diagnosis of NF1 can be easy when cutaneous stigmata are present. In some instances, no multiple NFs can be identified and café-au-lait patches can be discreet, emphasizing a careful examination of a patient diagnosed with aNF. [6]

NFs rarely occur in the breast and nipple-areola complex is the most usual location of this entity. [6]

The majority of reported breast NFs in the literature are related to neurofibromatosis type 1. to our knowledge, this isthe first case of breast diffuse neurofibroma that is not related neurofibromatosis 1. [7]

Solitary neurofibromas are ideally treated with surgical excision. [8]

Conclusion

Nipple-areola complex neurofibromas are rare and occur also in patients with no history of neurofibromatosis type 1.The diagnosis of NF should lead to a careful skin examination to identify any cutaneous stigmata or other neurofibromas.

Competing interests

The authors declare no competing interest.

Authors' contributions

All mentioned authors have equally contributed to: research in the English literature, redaction of the manuscript,collection of the patient's data, analyzing of the radiological and pathological data.

Acknowledgements (if any)

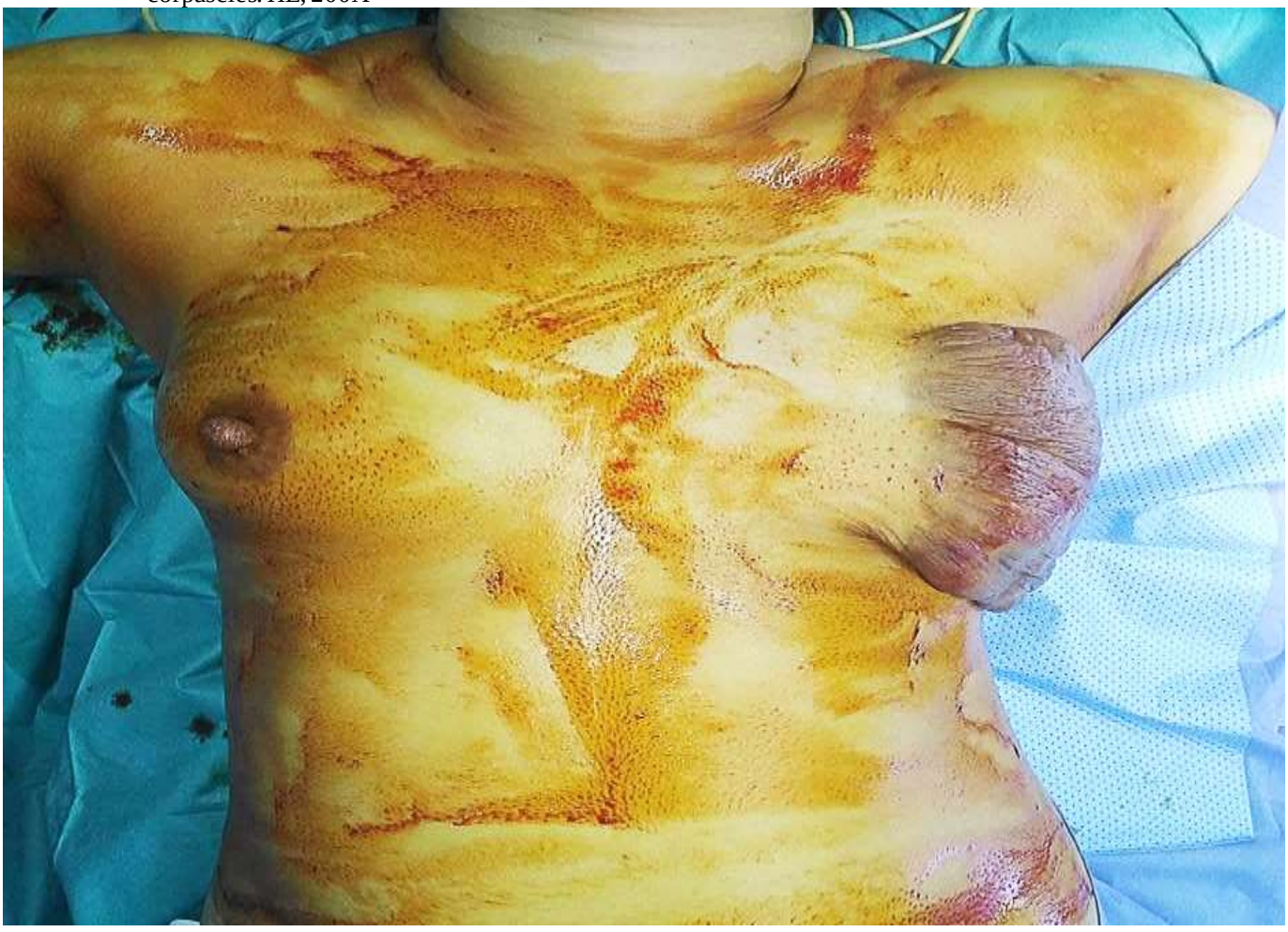
None

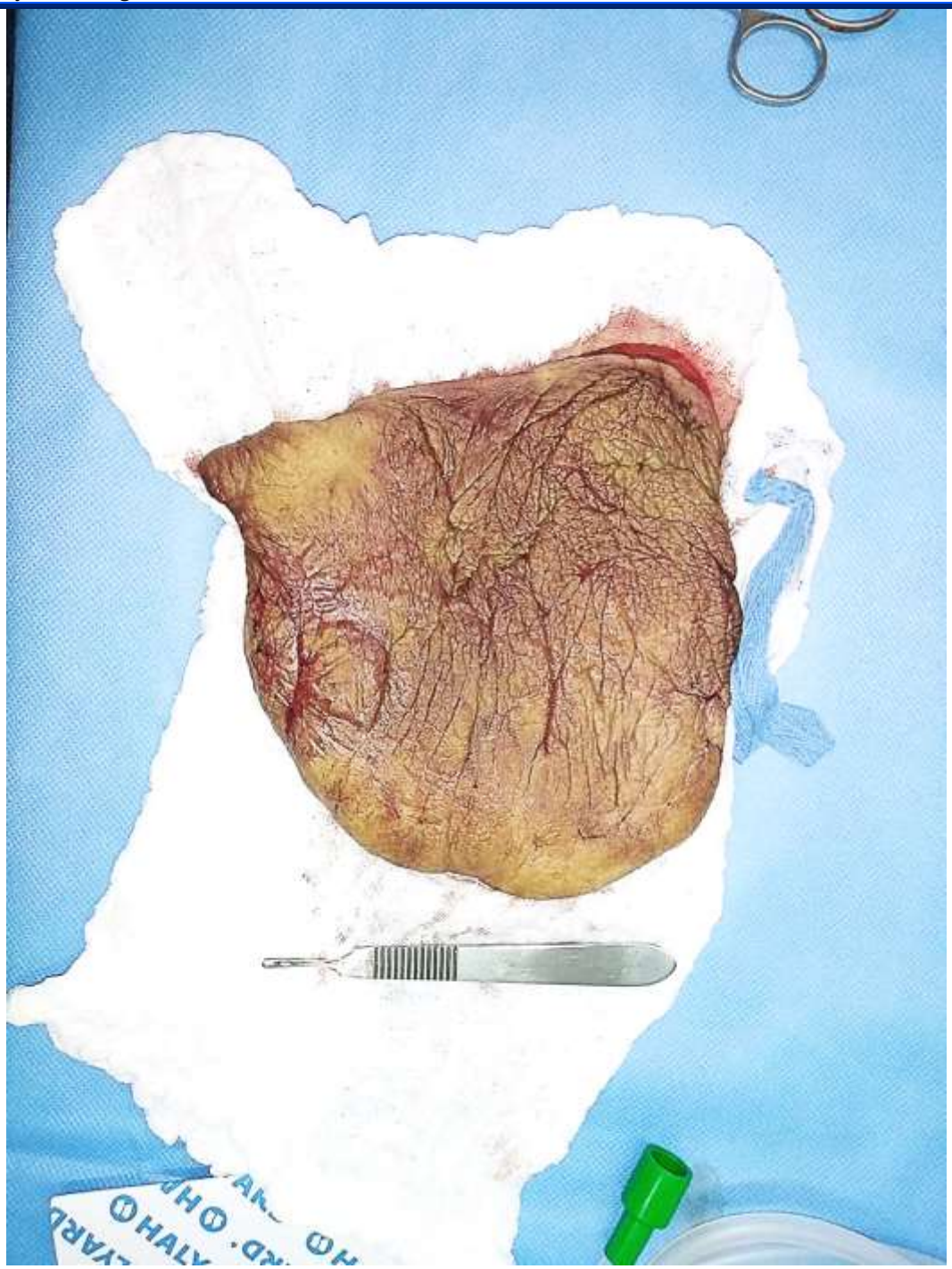
Tables and figures (if any)

Figure 1. A large disfiguring, slightly pigmented mass is observed on the left breast.
Figure 2. The resected specimen containing the tumoral mass.
Figure 3. Microphotography showing a hypocellular ill-circumscribed proliferation made of bland spindle cells(star). A lactiferous duct is also seen (arrow). HE, 100X

Figure 4. Microphotography showing an admixture of Schwann cells (Arrow), fibroblasts (arrow head) and presence of many mast cells (star). HE, 400X

Figure 5. Microphotography showing infiltration of the normal adipose tissue and presence of numerous Meissner corpuscles. HE, 200X





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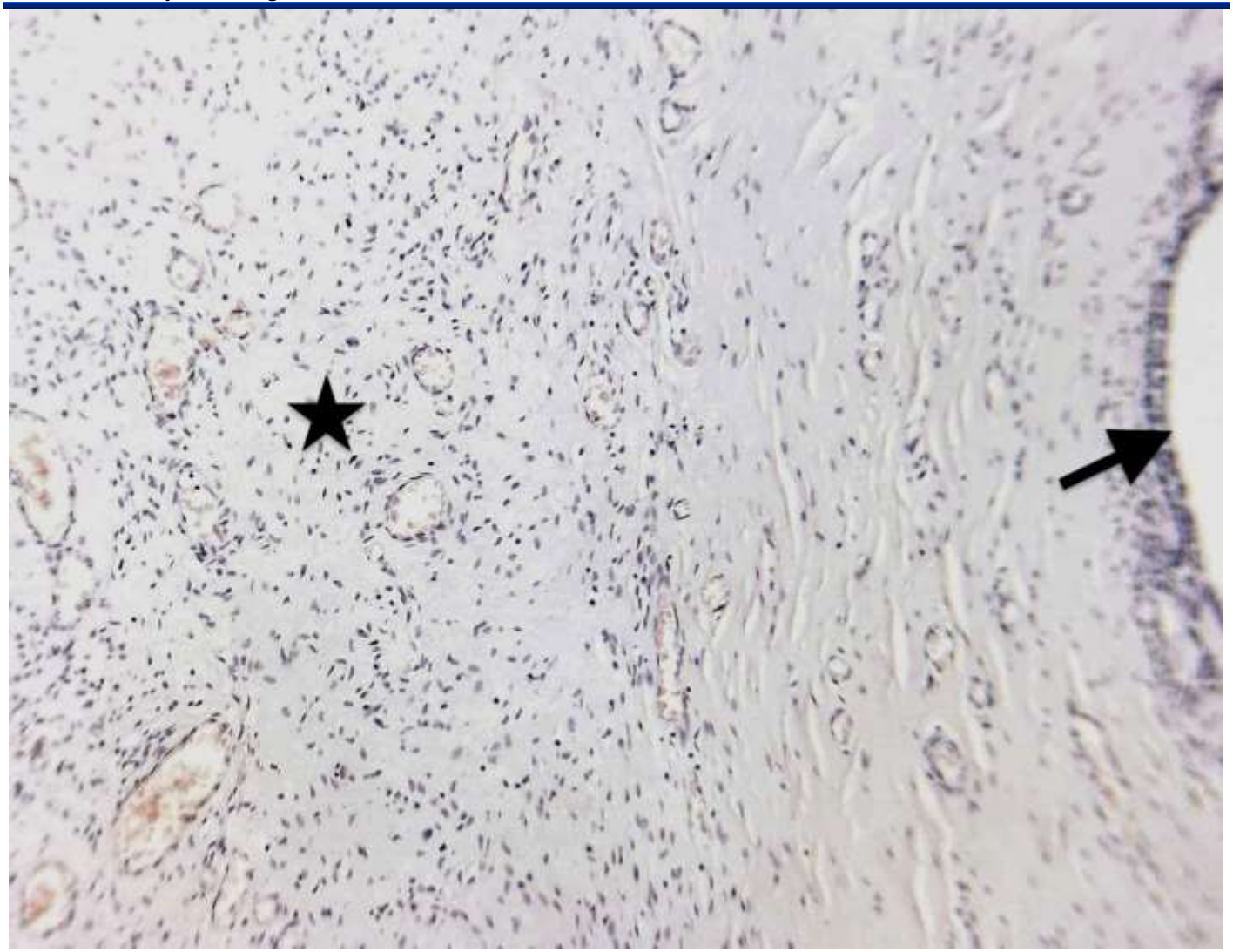

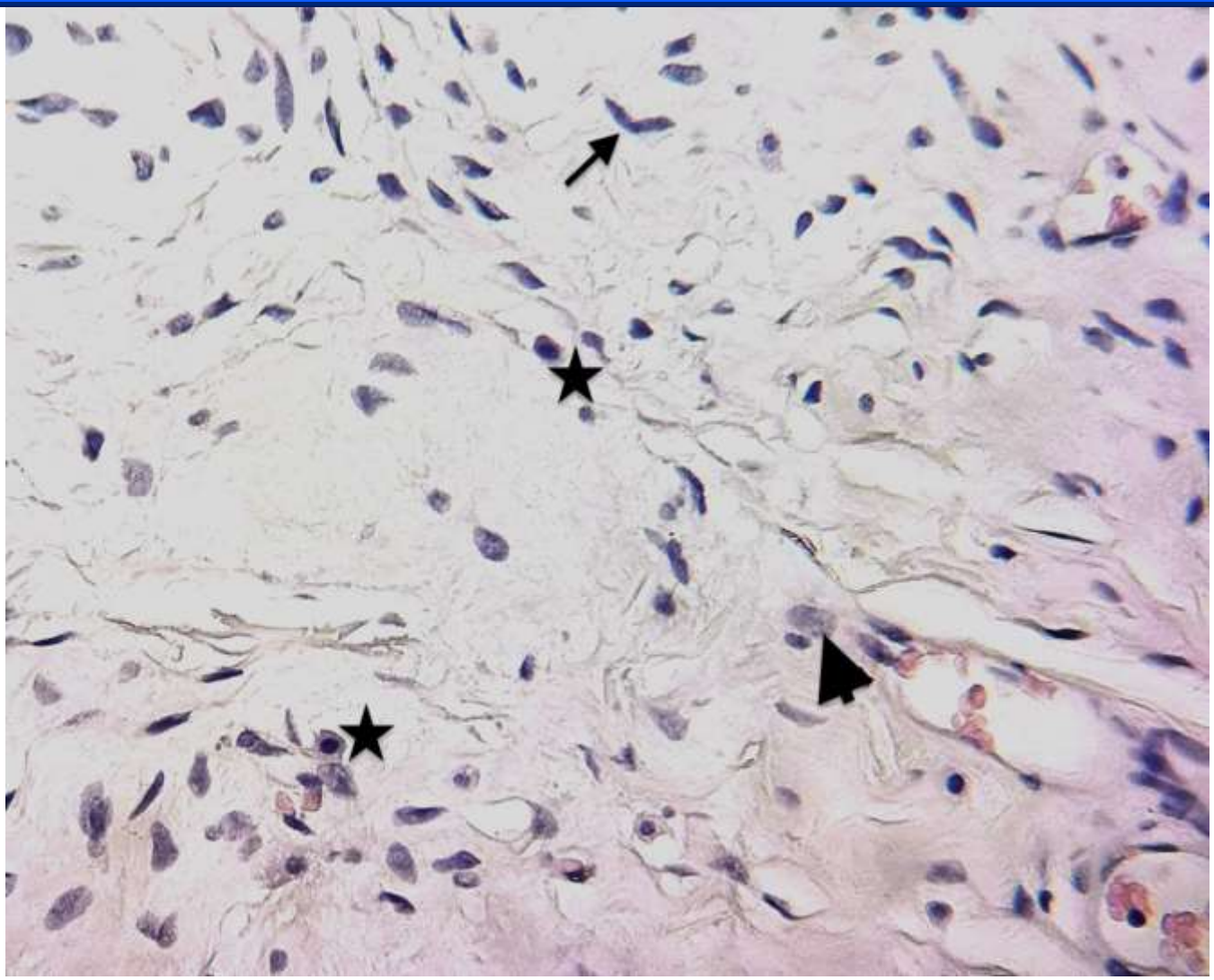

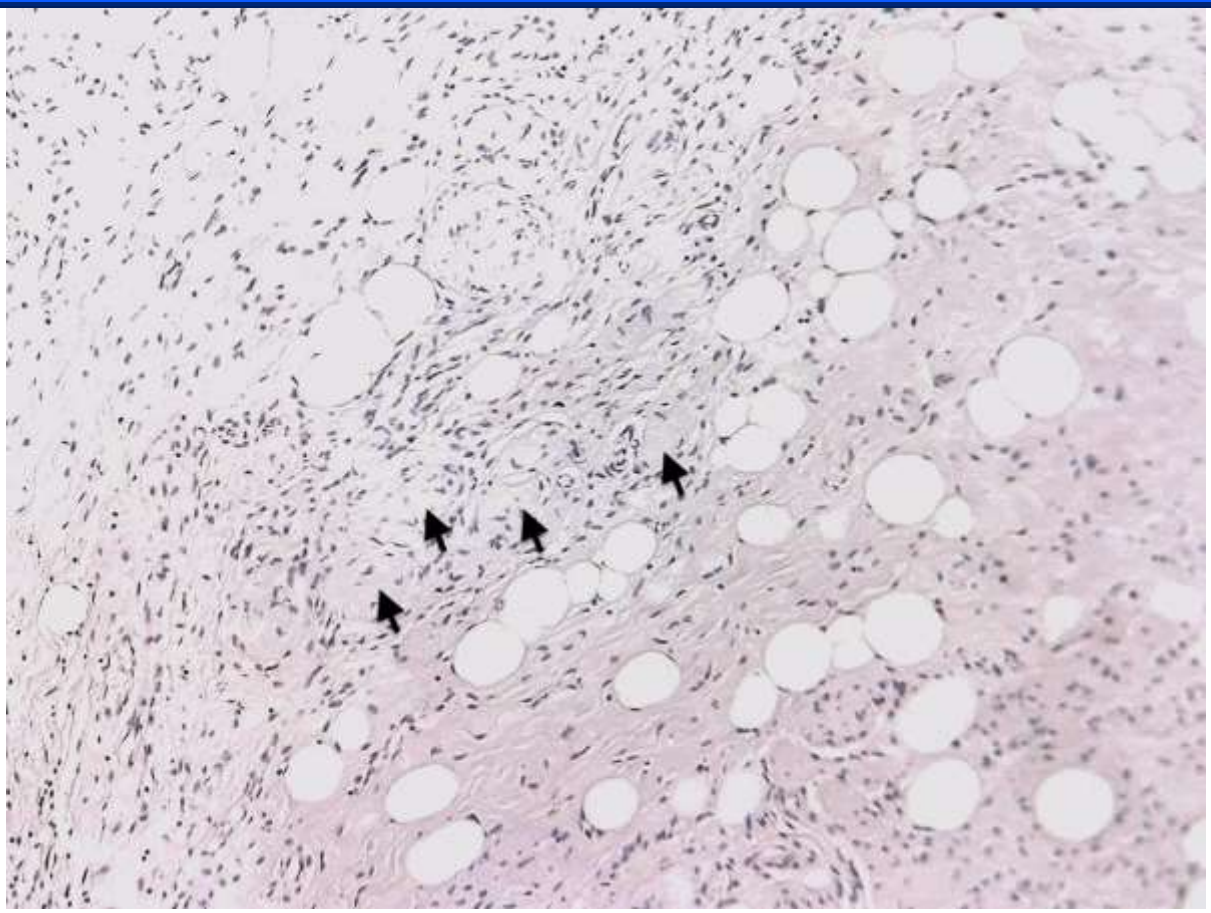

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