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STRANGE LOCALIZATION OF A MESENCHYMAL TUMOR - PARATESTICULAR LIPOSARCOMA

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Abstract: *Liposarcoma is a very rare mesenchymal tumor that appears from an unknow cause. All around the world there are just 200 cases of paratesticular liposarcomas reported. We presented the case of a male patient, 66 years old, that was first clinically diagnosed with orchi-epididymitis. Orchidectomy was performed the histopathological exam led to a tumor from the spectra of liposarcomas, which is a rare diagnose. The aspect of the lesion macroscopically was of a well circumscribed nodular structure, yellow, with white areas. Immunohistochemistry was conducted for confirmation and the profile was characteristic for a primary pleomorphic liposarcoma.*

Key words: *liposarcoma, pleomorphic, paratesticular*

1. Introduction

Liposarcoma is a rare mesenchymal malignant tumor that arises from the lipoblasts and involves the soft tissue. This neoplasm is the most common soft tissue sarcoma and holds 20% of the total of all malignant mesenchymal neoplasms. The most frequently affected site is the retroperitoneum and the other common area of interest includes the lower extremities, especially the popliteal fosa [10].

Histologically, there were several classifications made during years. Currently, according to the latest morphological features from the 8th edition of the American Joint Committee on Cancer, these lipomatous tumors are represented by the following types: well differentiated liposarcoma, dedifferentiated liposarcoma, myxoid-round cell liposarcoma and pleomorphic liposarcoma. The well differentiated liposarcoma (WDL) and dedifferentiated liposarcoma (DDL) are the most common types and together they represent 40-50% from the total of liposarcomas [3].

WDL appearance resembles the normal lipomatous tissue. The difference between the malignant proliferating cells and the mature adipocytes is made by nuclear pleomorphism, and fibrous septation, along with fibrous stroma. This tumor has

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a better prognosis than other liposarcomas due to the fact that it can recur, but the metastatic capacity is very low. However, this type can transform in a 10% of the cases in a DDL.

The DDL type can appear under the form of a pleomorphic sarcoma or a spindle cell sarcoma and can resemble other type of tumors. The diagnosis of the particular type is extremely important because the tumor has overall a better prognosis than other tumors it can be mistaken with, such as leiomyosarcoma, because of the lower risks of recurrence and metastasis.

2. Case Report

We present the case of a 66-year-old male patient that presented with an enlarged right testicule and was clinically diagnosed with orchi-epididymitis. Orchidectomy was performed and the sample was sent to the Pathology Department of the Mureș County Hospital (Târgu Mureș, Romania) for examination and histopathological diagnosis. The tissue was fixed in formaline and processed according to the protocols.

At the macroscopical examination, on the gross section, we observed a 80x60x25 mm well circumscribed nodular mass that was compressing the testicular parenchyma. The lesion had well defined borders, multinodular structure and yellow color with white areas, no other significant features.

On the microscopical examination, the tumor was composed by a solid proliferation of lipoblasts. The cells varied in dimensions and shapes, from round to spindle and the cytoplasm was pale, multi-/univacuolated. The nuclei showed high pleomorphism, bizarre shapes and hyperchromatosis. Between the neoplastic cells, multinucleated cells were also observed, especially at the peripheral region of the neoplasm. In some areas, myxoid differentiation was shown, along with thin and numerous branched vessels. Furthermore, necrosis was present in more than 50% of the tumor. The number of mitosis, most of them atypical, was 7/10 HPF. These last two components, necrosis and mitosis, along with the score for the resemblance of the tumor with the normal tissue are important for determine the grade of the lesion. In our case, the tumor was scored with 3 points for the grade of differentiation (abnormal cells that to not resemble mature adipocytes), 2 points for necrosis and 1 point for the number of mitosis. The final result classified the liposarcoma in G3 according to AJCC (American Joint Committee of Cancer) with a total score of 6 points.

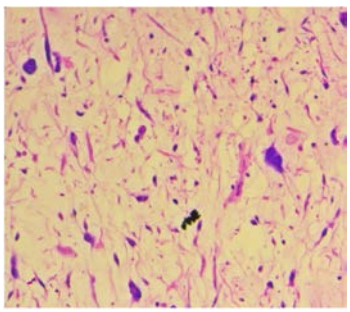

Fig.1. *Liposarcoma, Hematoxilin & Eozine stain, 500x magnification*

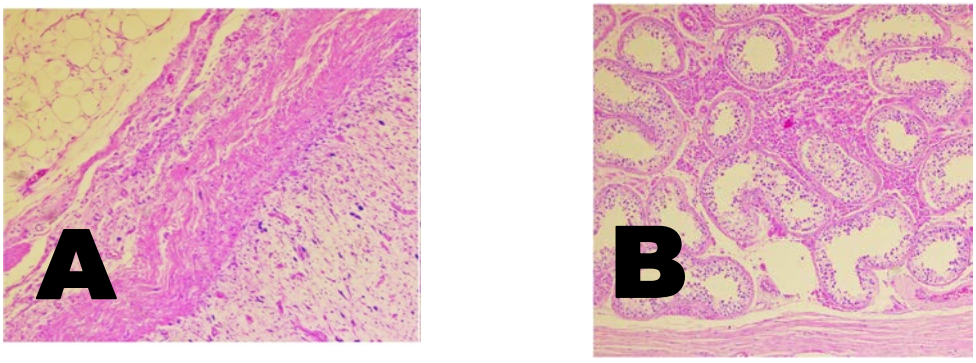

Fig. 2. *The border of the tumor and the testicular parenchyma, Hematoxilin & Eozine (A and B)*

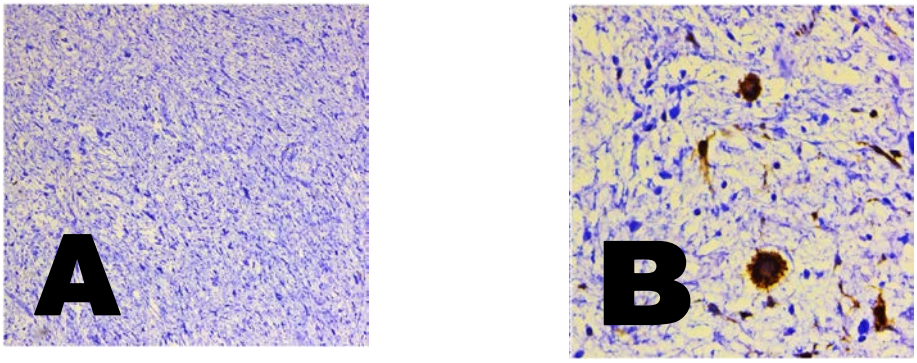

Fig. 3. *Marker S100 nuclear stain A -200x magnification, B- 500x magnification*

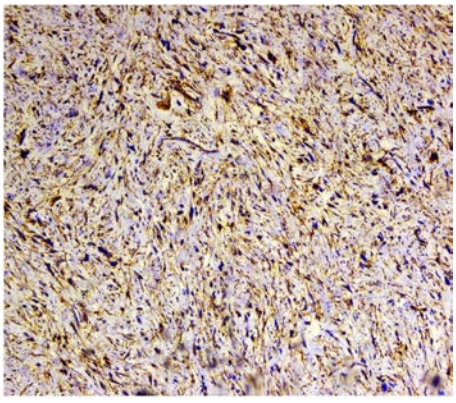

Fig.4. *Marker CD34 staining marks the tumor cell, 500x magnification*

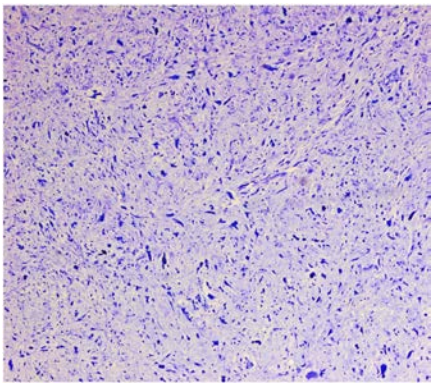

Fig.5. *Marker Ki67, 200x magnification*

3. Discussions

The rarest and less understood type is the pleomorphic liposarcoma, a lesion that appears in 5% to 10% of the cases. This type of sarcoma is a very rare entity that can rise challenges in diagnosis and it is characterized by a major grade of atypia. This tumor usually arises as a painless mass that grows in dimension slowly and has a compressive character. The most common areas in which this type of sarcoma occurs are upper and lower extremities with a higher involvement of the second, the abdominal wall, the chest wall, the spine and the cavities (pleural, pericardium, pelvic) and surgery is the main therapeutic option. It has the highest capacity of producing invasion, metastasis and also recurrences. The therapeutic options are currently controversial and studies all around the world are working to find a progress and develop further treatments [1].

The pleomorphic liposarcoma was described in various regions of the human body, such as esophagus, lower abdomen, lower extremities, mediastina and the regional organs, also head and neck areas [8, 7], [10].

Studies showed the most common age for this lesion had a peak at the interval between 50-70 years [9].

While taking in consideration immunohistochemistry, according to World Health Organization, the tumor is positive for the following markers: S100, SMA and CD34. Other stains that could be expressed in pleomorphic liposarcoma are represented by EMA, CTK AE1/AE3, while markers such as CDK4 and MDM2 are usually negative [4].

Regarding paratesticular tumors, only a proportion of maximum 10% can be reserved to lipopsarcomas and they usually arise from the spermatic cords. A proportion of only 20% of these tumors is represented by lipomatous neoplasms from all categories, benign and malignant [9, 10].

After analyzing the histological aspect of the lesion, we took into consideration several differential diagnosis. The most important aspect arises in cases in which the liposarcoma embraces a myxofibrosarcoma appearance, being called “myoxofibrosarcoma like”.

The other important neoplasms we considered in this particular tumor are sarcomas with epithelioid aspect, poor differentiated carcinomas with origin in renal cell carcinoma or melanoma. A liposarcoma that has a well-developed vascular pattern has to be differentiated from as well hemangiopericytoma or angiosarcoma [6], [9].

Other lesions we took into consideration were represented by other types of sarcomas, especially dedifferentiated liposarcoma with leyomiomatous differentiation (this type also has well differentiated component, missing in our case), metastatic carcinoma (no liposarcomatous differentiation) pleomorphic lipoma (collagen, very rare mitotic figures and low pleomorphism) pleomorphic rhabdomyosarcoma (has skeletal

muscle differentiation) and pleomorphic undifferentiated sarcoma.

To confirm and support the diagnosis of pleomorphic liposarcoma, we carried additional tests involving immunohistochemical reactions, by choosing the most specific markers for this lesion. The results had the further outcome: the malignant cells showed positivity for markers S100, CD34 and Ki67 was represented in a 50%. The tumoral cells were negative for CD68. According to most studies, this profile is characteristic for pleomorphic liposarcoma [5].

4. Conclusions

The paratesticular area for liposarcoma is extremely uncommon, with just 200 cases described in the literature up to this point. Pleomorphic liposarcoma is the rarest subtype and its appearance was described more frequently in the retroperitoneum. The presence of this tumor in the paratesticular region is extraordinary.

References

1. Anderson, W.J., Jo V.Y.: *Pleomorphic liposarcoma: Updates and current differential diagnosis*. In: Semin Diagn Pathol. 2019 Mar; 36(2), p. 122-128.
2. Benna, M., Zarraâ, S., et al.: *Rare Case of a Well-Differentiated Paratesticular Sarcoma of the Spermatic Cord in a 60-Year-Old Patient.* In: Case Rep Urol., 2017; 2017: 7903242.
3. Cates J.: *Staging System for Soft Tissue Sarcoma of the Extremities or Trunk: A Cohort Study of the SEER Database.* In: J Natl The AJCC 8th Edition *Compr* Canc Netw., 2018, 16(2), p. 144-152.
4. Fletcher, C., Bridge, J., et al.: *Pleomorphic liposarcoma*. In: World Health Organization Classification of Tumors o soft tissue and bonem International Agency for Research on Cancer. Lyon, 2013.
5. Galosi, A.B., Scarpelli, M. et al.: *Adult primary paratesticular mesenchymal tumors with emphasis on a case presentation and discussion of spermatic cord leiomyosarcoma.* In Diagn Pathol., 2014 May 6; 9, p. 90.
6. Jevremovic, V., Yousuf, A., et al.: *A rare presentation of myxo-fibrosarcoma as a Pancoast tumor: a case report.* In: J Med Case Rep. 2017; 11(1), p.61.
7. Morales-Codina, A.M., Martín-Benlloch, J.A., et al.: *Primary pleomorphic liposarcoma of the spine. Case report and review of the literature.* In: Int J Surg Case Rep. 2016, 25, p. 114-119.
8. Mumert, M.L., Walsh, M.T., et al.: *Pleomorphic liposarcoma originating from intracranial dura mater*. In: J Neurooncol., 2010, 97(1), p. 149-153.
9. Sheela, K.M., Lailaraji, N., et al,: *Pleomorphic Sarcoma in Paratesticular Region*. In: World Journal of Pathology Volume, No. 9, 2017.

10. Wang, L., Luo, R., et al.: *Pleomorphic liposarcoma-An analysis of 6 case reports and literature review*. In: Medicine 2018 Feb; 97(8), p. 9986.