EUROPEAN OPHTHALMIC PATHOLOGY SOCIETY (EOPS)

60th ANNUAL MEETING

Date of the meeting: 25th May – 28th May 2022.

Location: Valencia, Spain.

Invited member's name: Nuno Jorge Lamas

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Case number: H20-10778.

Material distributed: 1 histology H&E glass slide.

SOLITARY FIBROUS TUMOUR OF THE ORBIT

CLINICAL HISTORY

A 53-year-old female with a right orbital swelling, ptosis and visual change evolving for 8 months. The CT scan showed an expansive lesion in the superomedial quadrant, with grossly ovoid and lobulated contours, ≈22x22x11 mm, with regular and well-defined limits and homogeneous content, hypodense in relation to the muscle tissue. The lesion apparently contacted the eyeball, the medial rectus and superior rectus muscles, deflecting them, with no signs of invasion. Surgery was performed to remove the lesion.

OCULAR PATHOLOGY

MACROSCOPY

An irregular fragment of elastic and brownish tissue measuring 25x17x5 mm with a spongy and brownish cut surface was received in the Pathology laboratory.

MICROSCOPY

The histological analysis showed fibroadipose tissue almost completely occupied by a mesenchymal neoplasia consisting of ovoid to fusiform cells (Figure 1A, H&E, 100x magnification), with poorly defined cytoplasmic limits, with scarce cytoplasm and small to intermediate-sized hyperchromatic nuclei, sometimes with evident nucleoli and occasional images of nuclear pseudoinclusions (Figure 1B, H&E, 200x magnification). The cellular pleomorphism was mild to moderate. Two mitotic figures in 10 high-power fields (Figure 1C, yellow circle H&E, 250x magnification) were identified. The neoplastic cells were distributed in a disorganized manner around a rich vascular network, sometimes with staghorn vessels (Figure 1D, H&E, 200x magnification). There were hemorrhagic zones with macrophages rich in hemosiderin pigment. Necrosis was not identified.

COMPLEMENTARY STUDIES

The imunohistochemistry study conducted showed that the neoplastic cells had strong and diffuse staining for CD34 (Figure 1E, 200x magnification) and STAT6 (Figure 1F, 200x magnification). In addition, the neoplastic cells were focally positive for MDM2 (Figure 1G, 200x magnification) and were negative for the expression of CAM5.2 (Figure 1H, 200x magnification) and SOX10 (Figure 1I, 200x magnification). To further understand MDM2 focal positivity we conducted a cytogenetics study through fluorescence *in situ* hybridization (FISH), which did not reveal MDM2 amplification.

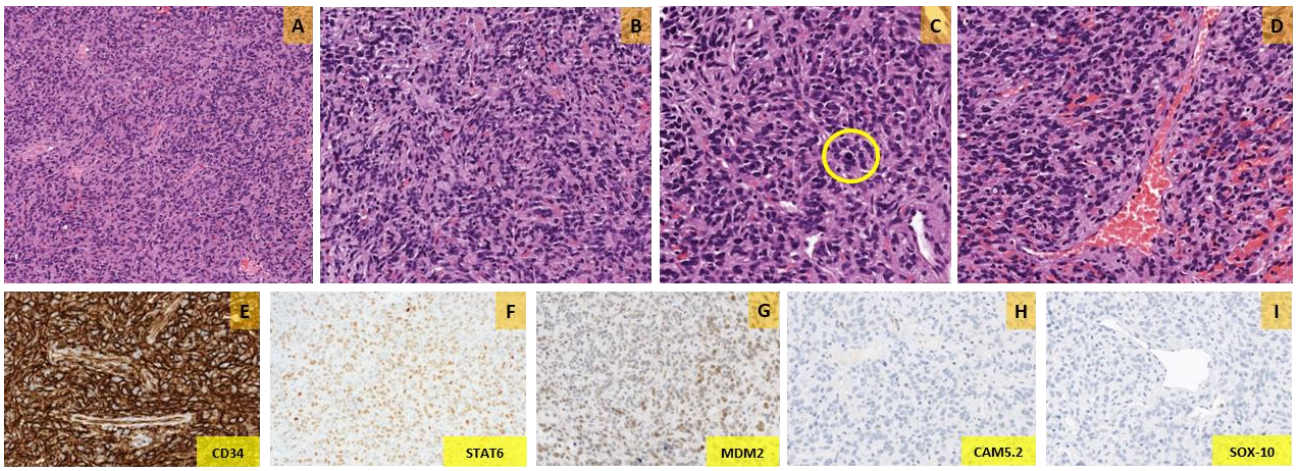

Figure 1. Solitary fibrous tumor of the orbit in a middle-aged woman.

The lesion was apparently incompletely excised. The combined WHO score for metastasis risk was demonstrated to be low [4-variable model = 1 point; Patient age in years < 55 (0 points); Mitoses per 10 HPFs = 2 (1 point); Tumor size in cm = 0 – 4,9 (0 points); Tumor necrosis < 10% (0 points)].

DISCUSSION

Solitary fibrous tumours (SFTs) occur rarely in the orbit and seem to afflict both sexes equally. Orbital SFTs present in younger patients (median 42 years) than the 5th to 7th decades for other sites. SFTs are characterized by a NAB2-STAT6 fusion resulting from a genomic inversion involving the 12q13 gene locus. Dedifferentiated liposarcomas (which have MDM2 amplification) with a SFT-like morphology normally have STAT6 nuclear expression, which could constitute an important diagnostic pitfall.

SFTs generally have a benign clinical behaviour, however, nearly 1/3 of orbital SFTs have local recurrences. Distant metastasis are uncommon and might only occur after extended periods. There are several proposals of risk stratification models for the prediction of metatastic risk in SFT cases, which are based on the clinical and pathological findings. The currently most widely accepted model (WHO score) includes patient age, mitotic index, largest tumoral dimension and the presence of necrosis. Recently, Lester Thompson and colleagues proposed a risk prediction model unique to orbital SFTs, which is based on patient age > 45 years, tumor size > 3 cm, tumor necrosis, mitoses of > 4/2 mm2, moderate to high cellularity, and moderate to severe pleomorphism.

The long term follow-up of these patients is extremely important, since metastasis might develop on the long term. For example, in a recent case report, a japanese patient was shown to have developed lung metastasis along with orbital recurrence 41 years after the initial orbital SFT diagnosis and surgical treatment. Based on the WHO score, the risk of metastasis for the patient presented in this meeting is low. Nine months after the surgery, the patient underwent a cycle of adjuvant stereotactic fractionated radiotherapy. Currently, 22 months have elapsed since the initial orbital ressection surgery and the patient remains well, without signs of local or distant disease.

REFERENCES

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