THE 14-YEAR FOLLOW-UP ON A MALIGNANT SOLITARY FIBROUS TUMOUR OF THE CAVATUS POPLITEUS

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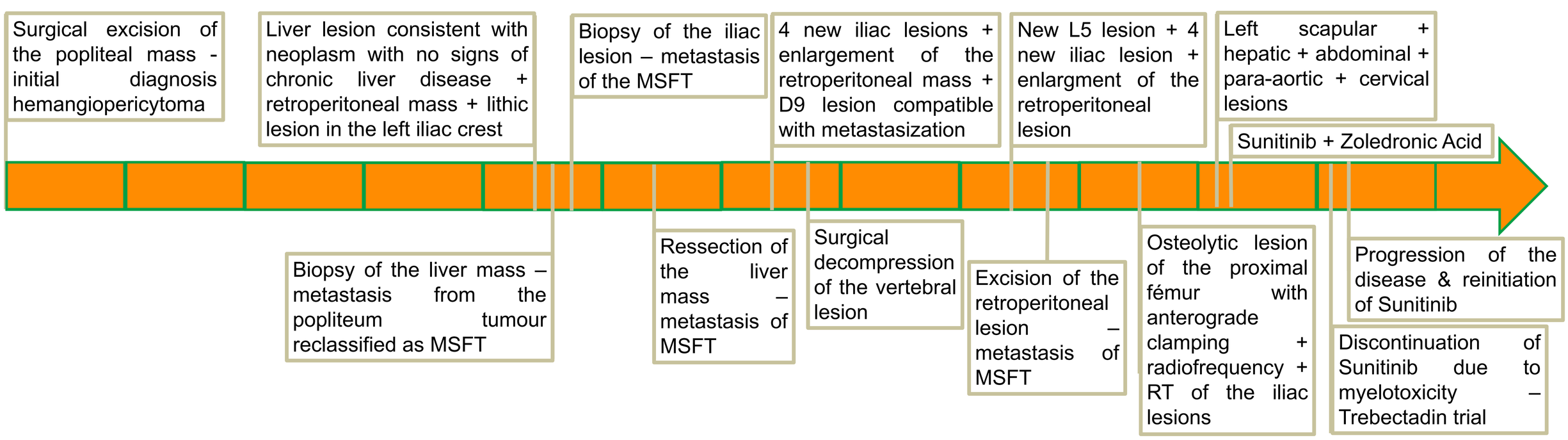
Abstract

A woman with pain and swelling of the right cavatus popliteus was submitted to an ultrasound-guided percutaneous biopsy. The diagnosis was hemangiopericytoma and wide resection was performed. Six years later, a liver lesion was diagnosed and biopsied, disclosing a metastasis of a MSFT. Pathologists revised the histology of the popliteum hemangiopericytoma and considered it the primitive tumour. She underwent right hepatectomy. One year later, a thoraco-abdominal CT-scan revealed metastases in the left iliac crest, treated with radiofrequency. Another year later, D8 metastases were found and treated with surgical decompression and radiotherapy. Another year past, the disease progressed with para-renal, later excised, and multiple bone metastases. Initiated sunitinib, thus stabilizing the disease. A trabectedin trial was tried due to sunitinib-related toxicity, but was soon abandoned due to impossibility of disease control, and restarted sunitinib. The disease has been stable ever since, and the patient is alive after 14 years of follow-up.

Case Presentation, Investigation and Treatment

A 20-years old woman with no relevant pathological background is referenced to a specialty appointment of orthopaedics by her general practitioner due to complaints of pain and swelling of her right cavatus popliteus with 2 months of evolution. There was no history of trauma to the knee. She had no family history of oncological disease. At physical examination, a small solid tumefaction was observable, with no signs of inflammation. A right knee magnetic resonance imaging (MRI) was performed in order to better assess the said tumefaction. The exam reported an image with an oval configuration, with a longitudinal axis of 85mm and posteroanterior of 51mm, that significantly captured contrast product (Gd STPA). At this point, the differential diagnoses included entities like synovial sarcoma, fibrous histiocytoma and giant-cell tumour. An urgent ultrasound-guided percutaneous needle biopsy was performed. The histological piece was analysed by the pathology lab and showed negativity for cytokeratins MNF, smooth muscle specific actin, desmin and protein-S100 and positivity for vimentin and CD34. The histological and immunocytochemical picture was compatible with glomangiopericytoma. The patient was proposed for urgent excision of the lesion, which happened about a month after the first appointment. The surgical piece was sent to the pathology lab for histological examination. The piece was a morphologically benign hemangiopericytoma. Due to the fact that the diagnosis was a benign entity, no adjuvant radiotherapy and chemotherapy was proposed. The surgery did not cause any compromise of function to the right lower limb, thus physiotherapy was not needed.

Follow-up Timeline



Discussion

Solitary Fibrous Tumours are rare group of neoplasms arising from mesenchymal cells, possibly from fibroblasts/primitive mesenchymal cells. The aggressive course of the disease is related to the size of the tumour, its histological degree (presence of mitosis, nuclear pleomorphism) and the presence of necrosis. In the tumours with a malignant behaviour the prognosis is worse and a tight follow-up programme is mandatory. The standard of care consists of chemotherapy, however there are some reports of it not being inferior to TKI-based therapy. it is important to advertise this sort of tumours in the literature: there are few publications on MSFTs and most of them do not report outcomes of patients under TKI-therapy, much less with 14 years of follow-up and three years of therapeutics with this novel drugs.

Learning Points/Take Home Messages

- Malignant Solitary Fibrous Tumours are rare neoplasms with an unpredictable clinical course, in which the diagnosis and therapeutics are not yet standardized.
- It is important to keep on track of what is being published when managing a patient with a rare neoplasm – it can be life-saving.
- Novel therapeutics with Tyrosine-Kinase Inhibitors are promising in what concerns the stabilization of a then multi-metastasized disease

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