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Short Communication

Retroperitoneal Liposarcoma: The Endless Challenge

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Liposarcoma is the most common soft tissue sarcoma accounting for approximately 20% of all sarcomas in adults. Based on WHO criteria Liposarcomas are classified into four histological subtypes:

- Well-differentiated Liposarcoma (WDLS)
- De-differentiated Liposarcoma (DDLS)
- Myxoid/round cell Liposarcoma (M/RLS)
- Pleomorphic Liposarcoma (PMLS)

Well-differentiated LS is a low grade lesion and is the most common among the four types. It has a high local recurrence rate but a minimal metastatic potential. Five year survival is recorded up to 90% in several series. De-differentiated LS is a high grade sarcoma with high local recurrence probability. The distant metastatic rate goes up to 10-15% and 5year survival is 75%. Myxoid/round cell and pleomorphic LS are also high grade but rare types of liposarcoma [2].

Well- differentiated LS and De-differentiated LS often coexist when firstly diagnosed. In addition, in every subsequent recurrence a fraction of WDLS can evolve to DDLS.

Retroperitoneal Liposarcoma (RPLS) can grow into quite big masses since the large retroperitoneal space gives the opportunity to this "silent" neoplasm to reach gigantic dimensions. Firstly diagnosed retroperitoneal Liposarcomas size between 10-26 cm in large series. Although, imaging modalities per se are not considered to be very helpful for preoperative differential diagnosis, Neuhaus., *et al*. in his publication from Royal Marsden at 2005 reports that CT scan can provide important information about tumor grade [3]. Nevertheless, despite all diagnostic tools available, precise histological identification of large tumors seems a very difficult task to achieve, since it's only after removal and dissection of the whole specimen that the histological subtype and grade is identified securely.

Although high grate tumors have more aggressive behavior compared to low grate, the initial operation in all cases should be designed in the same way. Several authors agree that R0 resection is mandatory during the first operation, even if adjacent structures have to be removed [1-4,6]. Clear margin is defined as the absence of microscopically detectable tumor within 1mm of inked specimen. Samuel Singer., *et al*. [2] in a retrospective study of 177 patients with primary liposarcoma at 2003 they conclude that, as long as complete resection can be achieved tumor burden and nephrectomy have no influence in survival, though they clarify that sacrificing the kidney is not necessary if the tumor is confined on the capsule. In that case capsulectomy without nephrectomy is an acceptable option.

Lucas Matthyssens., *et al*. [4] consider surgery the cornerstone of treatment for non-metastatic RPLS advocating en-block resection of involved structures aiming to reduce the risk of local relapse. According to their data, complete (R0) resection should be the optimal target despite the fact that it often requires removal of psoas, abdominal wall or even sections from arteries and veins.

Even if complete (R0) resection and the extent of differentiation (grate) have proven to be the two most significant predictors for disease specific survival (DSS) for patients with primary tumors, when it comes to recurrence disease the data suggest otherwise. No matter how aggressive the initial surgical approach it may be, over 80% of all cases eventually will recur locally. Chemotherapeutic agents and localized radiotherapy have been tested with disappointing results, although MDM2 and CDK4 antagonists and tyrosine kinase antagonist Sunitinib could be promising especially for advanced or unresectable LPS [4]. Since there is no study proven effective adjuvant therapy, surgical treatment remains the best option for recurrence cases too. The question is how extensive the resection should it be.

During the first operation contiguous organ resection is justifiable. This is less frequently indicated in reoperations since it doesn't change the final course of the disease whereas it increases significantly postoperative morbidity and mortality [8]. According to the

Royal Masrden Hospital team [3], surgical palliation for recurrence is effective in terms of treating disturbing or severe symptoms, it is well tolerated and can be applied as many times as possible. On the other, hand according to the same publication, visceral resection may still be necessary to facilitate subsequent resection or to "correct" previous suboptimal operation. So one should balance between palliative "conservative" resection but up to the point that one doesn't live behind obvious gross mass, aiming to prolong the interval until next relapse and also achieve the best possible QOL.

In conclusion, since it's usually not feasible to know the histological subtype of a firstly diagnosed large retroperitoneal liposarcoma, the treatment of choice should be radical resection with intention to cure. A histological 1mm negative margin is the gold standard in order to achieve good oncological results and contiguous organ removal should be performed if necessary.

No matter how aggressive the first operation local recurrence occurs up to 30% during the first two years, usually indicating a biological behavior that affects residual "normal" fatty tissue. Although distant metastases are rare, death often occurs as a result of local progression, making local control mandatory.

Taking into consideration the indolent course of the disease one must create a long term therapeutic strategy. Even if radical intervention is initially justifiable, extended surgical resection for recurrent disease doesn't seem to improve DSS. Debulking for palliation seems to postpone the development of critical tumor mass and subsequent fatal results.

Although no chemotherapeutic agent has proven to be effective, several targeted agents are being tested that may contribute to the available treatment options. This could change the natural history of the disease but until then, surgery remains the gold standard of treatment for all stages.

Finally, one cannot stress enough the need for a multidisciplinary team management for these cases during the full course of the disease. Since it usually affects people between 40 - 60 years of age team cooperation to achieve prolongation but also quality of life should be the primary target.

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