Fibromatosis: Where are we now?

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Abstract
Purpose of the study
Fibromatosis is a benign but locally aggressive tumour. A high rate of recurrence was noted in a number of patients treated by the senior author at a tumour and sepsis unit despite the fact that a wide surgical excision had been performed. The question was raised whether there are any alternate treatment modalities with a higher success rate available currently. A retrospective study and review of the literature was performed in order to ascertain whether new treatment modalities which can prevent recurrence more successfully have been developed recently.

Materials and methods
A retrospective study was performed. The files of all patients who presented at an orthopaedic practice with confirmed fibromatosis on histological examination in the past 19 years were reviewed. The following was looked at: age of the patient at first presentation; gender; tumour site; surgery performed; histological results; first line of treatment and recurrence rate. Patients were also contacted telephonically in order to ascertain whether any recurrence managed by another orthopaedic surgeon had been attended to.

Results
We evaluated 17 patients of which eight were males and nine females. The mean age was 25.87 years (range 2–52 years). All of the primary sites were extra-abdominal. Median follow up was 3.9 years (0–9) with a mean recurrence rate of 2.3 times. All the patients were treated with a wide marginal surgical excision without adjuvant therapy.

Conclusion
Fibromatosis has a high recurrence rate using current surgical treatment modalities. Complete surgical excision does not lead to a good outcome. The literature review revealed that a wide variety of treatment modalities, both surgical and non-surgical, are available. Non-surgical treatment modalities include: hormones; non-steroidal anti-inflammatory drugs; chemotherapy; radiotherapy. Wide surgical excision remains the mainstay of treatment but a multidisciplinary approach is necessary in order to optimise the efficacy of this treatment.

Level of evidence: Level III
Key words: Fibromatosis, retrospective study, histology, treatment modality, desmoid tumour
Introduction

Fibromatosis is a benign but locally aggressive tumour with a very high rate of recurrence. It has an infiltrative growth pattern which makes complete excision difficult. The National Comprehensive Cancer Network subdivides soft tissue sarcomas into four subdivisions: soft tissue sarcoma of extremity/trunk; retroperitoneal or intra-abdominal soft tissue sarcoma; gastrointestinal stromal tumours and desmoid tumours.¹⁻³ We will be focusing on the extra-abdominal division of desmoid tumours.

A high rate of recurrence was noted in a number of patients treated at the Pretoria tumour and sepsis unit despite the fact that a wide surgical excision had been performed. The question was raised whether there are more current alternate treatment modalities with a higher success rate. We looked at previously published articles regarding treatment modalities over the past few years and specifically did a review on the new treatment modalities published during the past 5 years.

Materials and methods

A retrospective study was performed. We evaluated all the patients who presented at the Pretoria tumour and sepsis unit with confirmed fibromatosis on histological examination during the past 19 years. The following was studied: age of the patient at first presentation; gender; tumour site; surgery performed; histological results; first line of treatment and recurrence rate. Patients were furthermore contacted telephonically in order to ascertain whether any recurrence managed by another orthopaedic surgeon had occurred.

Results (Table I)

Seventeen patients were evaluated. Desmoid tumour had to be confirmed histologically in order to be included in this study. One patient had Dupuytren's disease on histology and was excluded from this study. Seven patients were males and nine females (ratio: 0:0.8). The mean age was 25.87 years (range 2–52 years). All of the primary sites were extra abdominal, the highest percentage being on the extremities 62.5% (10/16). Median follow-up was 3.9 years (0–9) with a mean recurrence rate of 2.3 times. All the patients were treated with a wide surgical excision margin. No adjuvant therapy was given. Seven of the patients had complete surgical removal which was confirmed histologically; in all multiple recurrences occurred. The histopathologist was unable to determine the surgical margin in six of the specimens. Only three patients had no recurrence. No genetic screening was done on any of these patients.

A high rate of recurrence was noted despite the fact that a wide surgical excision had been performed

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age at first presentation</th>
<th>Sex</th>
<th>Follow-up in years</th>
<th>Amount of recurrence</th>
<th>Site</th>
<th>Follow-up till</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>19</td>
<td>F</td>
<td>8</td>
<td>7</td>
<td>Arm</td>
<td>2004-2012</td>
<td>C + R</td>
</tr>
<tr>
<td>2</td>
<td>39</td>
<td>F</td>
<td>6</td>
<td>2</td>
<td>Chest wall</td>
<td>2006-2012</td>
<td>C + R</td>
</tr>
<tr>
<td>3</td>
<td>36</td>
<td>M</td>
<td>9</td>
<td>4</td>
<td>Gluteal area</td>
<td>2003-2012</td>
<td>CM</td>
</tr>
<tr>
<td>4</td>
<td>46</td>
<td>F</td>
<td>7</td>
<td>7</td>
<td>Neck</td>
<td>2005-2012</td>
<td>C + R</td>
</tr>
<tr>
<td>5</td>
<td>14</td>
<td>F</td>
<td>2</td>
<td>3</td>
<td>Thigh</td>
<td>2002-2004</td>
<td>C + R</td>
</tr>
<tr>
<td>6</td>
<td>40</td>
<td>M</td>
<td>7</td>
<td>3</td>
<td>Leg</td>
<td>1993-2000</td>
<td>U</td>
</tr>
<tr>
<td>7</td>
<td>33</td>
<td>M</td>
<td>1</td>
<td>0</td>
<td>Posterior triangle neck</td>
<td>2000</td>
<td>U</td>
</tr>
<tr>
<td>8</td>
<td>5</td>
<td>M</td>
<td>3</td>
<td>1</td>
<td>Foot</td>
<td>2000-2003</td>
<td>U</td>
</tr>
<tr>
<td>9</td>
<td>25</td>
<td>M</td>
<td>1</td>
<td>1</td>
<td>Buttock, Thigh</td>
<td>2000-2001</td>
<td>C + R</td>
</tr>
<tr>
<td>10</td>
<td>6</td>
<td>F</td>
<td>3</td>
<td>1</td>
<td>Foot</td>
<td>2001-2004</td>
<td>IC</td>
</tr>
<tr>
<td>11</td>
<td>43</td>
<td>F</td>
<td>2</td>
<td>2</td>
<td>Foot</td>
<td>2003-2005</td>
<td>U</td>
</tr>
<tr>
<td>12</td>
<td>14</td>
<td>M</td>
<td>7</td>
<td>2</td>
<td>Foot</td>
<td>2002-2009</td>
<td>C + R</td>
</tr>
<tr>
<td>13</td>
<td>7</td>
<td>M</td>
<td>2</td>
<td>1</td>
<td>Foot</td>
<td>2002-2004</td>
<td>C + R</td>
</tr>
<tr>
<td>14</td>
<td>33</td>
<td>F</td>
<td>0</td>
<td>0</td>
<td>Paraspinal</td>
<td>2003</td>
<td>U</td>
</tr>
<tr>
<td>15</td>
<td>52</td>
<td>F</td>
<td>2</td>
<td>3</td>
<td>Elbow</td>
<td>2010-2012</td>
<td>U</td>
</tr>
<tr>
<td>16</td>
<td>2</td>
<td>F</td>
<td>2</td>
<td>0</td>
<td>Clavicle</td>
<td>2010-2012</td>
<td>C + R</td>
</tr>
</tbody>
</table>

C + R: Complete still recurr; U: Unable to tell histologically; IC: Incomplete
Aetiology and histopathology

Fibromatosis might be associated with genetic predisposing conditions (familial adenomatous polyposis; Gardner syndrome); hormonal influences (increased oestrogen levels, oral contraceptives, peripartum) and surgical trauma (especially at abdominal incision sites). Though it does not have the same histopathological features as a soft tissue sarcoma, clinical features do correlate with soft tissue sarcomas. Fibromatosis has a severe infiltrative pattern, with a high rate of recurrence and functional morbidity due to aggressive surgery.4

It is important to be familiar with the pathological features of this disease in order to treat it appropriately. We know that it is a benign proliferation of fibroblastic cells associated with the WNT (group of proteins)/APC (adenomatous polyposis coli)/β-catenin pathway which drives the process of formation of these tumours.5-8 The WNT/β-catenin pathway plays a role in transcription in the nucleus and cell adhesions. The APC (which is a tumour suppressor gene) controls the level of the β-catenin (which is an oncogene) by means of phosphorylation and this is indirectly controlled by the WNT pathway.9 β-catenin accumulation was originally demonstrated in desmoid tumours in FAP (familial adenomatous polyposis) patients who had a deficient APC complex. CTNNB1 (gene encoding β-catenin) was seen in a large population of sporadic desmoid tumours and leads to activation of the WNT pathway, thus increasing the β-catenin.9 The increased β-catenin level activates the T-cell factor. This causes transcription of the COX-2 (cyclooxygenase-2) leading to activation of the PDGFRA (platelet-derived growth factor receptor α) and PDGFRB (platelet-derived growth factor receptor β).10

Wu et al11 did a study on mice and found a correlation between fibromatosis and mesenchymal progenitor cells supporting tumour genesis via the β-catenin pathway. Matono et al2 evaluated 74 samples and came to the conclusion that there is overproduction of the VEGF (vascular endothelial growth factor) in desmoid tumours and especially in the recurrent group. They could not however find any correlation between the ‘β-catenin mutation and VEGF mRNA expression.

The histologist determined the positive or negative surgical margin which we are referring to in this article. It describes the margin of the tumour in the specimen which was resected and evaluated. ‘Positive’ means tumour at the border of the specimen excised (possible incomplete excision) and ‘negative’ means a clear, tumour-free surgical margin.

Fibromatosis has a severe infiltrative pattern, with a high rate of recurrence and functional morbidity due to aggressive surgery.

Treatment modalities

Conservative treatment

Barbier et al12 evaluated 26 cases of extra-abdominal fibromatosis of which 11 had no previous surgery and 15 recurred after surgery. They found that 24 of the cases stabilised at a mean of 14 months; only two regressed and showed evolution at 23 months. They came to the conclusion that a ‘wait-and-see’ principle can be optimal especially in those with a high risk of severe functional or cosmetic deformities after surgery. A few other smaller studies were also discussed in this article and showed similar results. They suggested that surgery makes the tumour more aggressive. The patient populations in these studies were however small.

Stoeckle et al13 evaluated 106 patients over a period of 123 months and came to the conclusion that the tumours stabilised after 3 years and some even regressed thereafter. They suggested a ‘wait-and-see’ approach and medical management in all tumours except those located primarily in the lower trunk wall/girdle. In the case of these tumours response was more favourable when surgery was performed.

Pignatti et al14 evaluated 83 cases and suggested that conservative management can be considered for non-progressive recurrent lesions. A retrospective study done in France by Bonvalot et al15 on 112 patients found a similar event-free survival: 65% versus 68%. The gender, age, tumour size and treatment period were not statistically significant but there was a correlation between the tumour site and quality of surgery with extremity tumours and positive surgical margins having a poorer outcome. A study done by Salas et al16 also found that age, tumour size, tumour site and surgical margins had a significant impact on progression-free survival, and suggested dividing desmoid tumours into prognostic subgroups to decide on management.

The above studies suggest that the least invasive method be used as first-line treatment; this however excludes patients with severe pain; a massive tumour or severe loss of function. Each patient still needs to be evaluated individually.

Radiotherapy options and the influence of the surgical margin

Gluck et al17 did a retrospective study and reviewed 95 patients treated at their institute over 24 years with surgery, radiotherapy or both. They found equivalent local control rates in all the groups; no difference between the outcomes of positive or negative surgical margins. They noted a higher recurrence rate (36.8% versus 16.8%) in head and neck tumours versus trunk, extremities, pelvis and retroperitoneal tumours. It is however important to note that the head-and-neck group was very small in relation to the other group (19 patients versus 54 patients).
They suggested adjuvant radiotherapy for patients with recurrence after surgery (possibly in positive surgical margins); site of origin – head and neck; and a poor functional outcome after surgery. Melis et al. published a review on how important a negative surgical margin is and came to a conclusion that you should not resect desmoid tumours due to the consequence of a poor functional outcome in order to achieve a negative surgical margin. In this review they concluded that treating recurrent lesions with surgical excision and radiotherapy had better results than surgery alone.

Gronchi et al. came to the conclusion after reviewing 203 cases retrospectively (35-year follow-up) that microscopic positive disease does not necessarily affect disease-free outcome in patients with primary lesions but definitely at recurrence. They found that 75% of those patients who presented with a primary lesion were cured even though they had a positive margin on histology. Forty of their patients had adjuvant radiation with a disease-free survival rate of 78% versus 72% at 5 years for those who were not treated with adjuvant radiation therapy.

Merchant et al. evaluated 189 patients of whom 105 complied with their inclusion criteria (with primary lesions) and found that there was no statistically significant difference microscopically between a positive or negative surgical margin with a local recurrence rate of 22% in those with a positive resection margin and 24% in those with a negative resection margin (p=0.51). There was also no difference in the group treated with adjuvant radiation therapy. Seven of the 31 patients (23%) who had radiation developed recurrence versus the 23% (17 of 74 patients) recurrence rate of those who didn't have radiation (p=0.82).

Spear et al. evaluated 107 patients who had surgery alone, radiation therapy alone, or both, and found that they had better local control in those patients with a negative surgical margin as well as those with a positive margin and radiation therapy.

Ballo et al. came to the conclusion that radiation therapy is effective in unresectable masses alone or in combination with a positive surgical margin. Tumours with negative surgical margins had a 10-year relapse rate of 27%; positive surgical margins had a relapse rate of 54%; and radiation therapy a relapse rate of 24% with a 75% expected control in those patients who were treated with radiotherapy over the long term.

Nuyttens et al. did a Medline search in 2000 and compared 22 articles. They came to the conclusion that radiotherapy alone or surgery plus radiotherapy had a better outcome than surgery alone.

There are numerous studies stating that adjuvant radiotherapy has a better outcome in selected cases but also many others stated no difference in the outcome.

Definitive radiotherapy must be considered in a patient where surgery will be severely debilitating. The benefits of radiation therapy versus the complications of radiation therapy such as wound breakdown, infection, secondary malignancies, lymphedema and pathological fractures should however be weighed up carefully when planning treatment.

Neoadjuvant radiation therapy has not been proven to have a better outcome. Studies are however limited.

Intra-operative electron radiotherapy (IOERT) has been studied by Roeder et al. They evaluated 30 individualised patients in whom surgical intervention alone would have had severe complications. They had a 3-year local control rate of 82% and concluded that IOERT might be feasible in a multimodal approach.

Radiofrequency ablation has been suggested as an alternative treatment option for desmoid tumours but studies are limited.

Percutaneous cryoablation in small desmoid tumours have been suggested by Kujak et al. They found that this treatment modality appeared to have adequate local control of the tumour as well as pain relief. This was however a retrospective study of only five patients.

When reviewing the above and many other studies one can come to the conclusion that surgical margin is a controversial measurement of recurrence. There is furthermore no clear indication as to whether radiation therapy should be administered or not. It does however make sense to give radiation therapy first in those patients with a high possibility of morbidity post-surgery. A multidisciplinary team approach is necessary in order to correctly individualise treatment. The surgeon, oncologist, pathologist and patient must be part of the decision-making team.

Systemic therapy
Systemic therapy has been experimented with for years in order to see if there is a less invasive way of managing desmoid tumours than with surgery and radiation therapy. Some of the systemic treatment options are: cytotoxic agents (anthracyclines); molecular targeted agents: imatinib; and antiestrogen hormonal agents (e.g. tamoxifen, toremifene),interferon and NSAIDs.

De Camargo et al. did a retrospective review of 68 patients seen at their institution who received some form of systemic therapy from 1994–2007. The median follow-up was 63 months. They found the following: partial response (PR): 19%; stable disease (SD): 58%; progressive disease (PD) 23%. It was concluded that systemic therapy is an option for patients with debilitating disease. They also noted that anthracyclines and hormonal therapy had a better response rate than dacarbazine/temozolomide or tyrosine kinase inhibitors. This is however a retrospective study over 13 years, the type of treatment and dosages were surgeon-dependent and could have also been influenced by the severity of the disease.

A multidisciplinary team approach is necessary in order to correctly individualise treatment.
Tyrosine kinase inhibitor (imatinib)
The exact mechanism of action for imatinib on a desmoid tumour is still being investigated. There have been a few studies evaluating the target of imatinib, and some of the possible target sites were: PDGFRα, PDGFRβ, c-KIT, PDGFα, PDGFβ and macrophage colony stimulating factor.38-43 We know it is a tyrosine kinase inhibitor. Table II summarises the studies we evaluated in order to see what the outcome of imatinib in desmoid tumours is.

<table>
<thead>
<tr>
<th>Study</th>
<th>No. of patients</th>
<th>Dose</th>
<th>Prior intervention</th>
<th>Response</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wcislo et al⁴</td>
<td>1</td>
<td>400 mg daily For 38 months</td>
<td>Yes</td>
<td>Responded</td>
</tr>
<tr>
<td>Goncalves et al⁵</td>
<td>1</td>
<td>400 mg daily For 10/52 600 mg daily For 10/52</td>
<td>Yes</td>
<td>First responder after 20/52 66% smaller at 34 weeks</td>
</tr>
<tr>
<td>Penel et al⁶</td>
<td>40</td>
<td>400 mg daily For a year (1 case 600 mg, 8 cases 800 mg)</td>
<td>Yes</td>
<td>Responded 1 year: CR: 1/35 (2.9%) PR: 3/35 (8.6%) SD: 28 (80%) PD: 3 (8.6%) NP: 67% PF: 55% 50% defaulted</td>
</tr>
<tr>
<td>Heinrich et al⁷</td>
<td>186 (different malignancies of which 20 (fibromatosis))</td>
<td>400 mg daily – 800 mg daily Mean duration 2.5 months &gt;20 months in 3/20</td>
<td>Unknown</td>
<td>92.5% defaulted due to unsatisfactory results PR: 2/20 (10%) SD: 8/20 (40%) PD: 7/20 (35%) Median time to progress: 9.1 months</td>
</tr>
<tr>
<td>Heinrich et al⁸</td>
<td>19 (7 – extra-abdominal)</td>
<td>800 mg/day</td>
<td>Yes: 18/19</td>
<td>1 year control rate: 36.8% No PR in extra-abdominal type PD: 1/7 (14.3%) SD: 6/7 (85.7%)</td>
</tr>
<tr>
<td>Mace et al⁹</td>
<td>2</td>
<td>400 mg BD</td>
<td>Yes</td>
<td>Responded</td>
</tr>
<tr>
<td>Chugh et al⁹⁵</td>
<td>51 (10 – Intra-abdominal; 41 – extra-abdominal)</td>
<td>100–300 mg daily (dependent on BSA)</td>
<td>Yes</td>
<td>4 months: CR: 43/51 (84%) PD: 5/51 (10%) 3/51 not evaluated CBR: 84% 42 patients defaulted treatment after 4/12</td>
</tr>
<tr>
<td>Kasper et al⁹⁶</td>
<td>9</td>
<td>400 mg – 800 mg daily</td>
<td>-</td>
<td>Used &quot;PET CT for evaluation SD: 7/9 (77.8%) PD: 2/9 (22.2%) All showed a 27% decrease in the standardised uptake value of the tumour</td>
</tr>
</tbody>
</table>


There is an indication for imatinib in the treatment of desmoid tumours with stable disease ranging between 28% and 84% and of progressive disease of 10%–35%
After reviewing the studies in Table II, the following conclusion was reached: there is definitely an indication for imatinib in the treatment of desmoid tumours with stable disease (SD) ranging between 28% and 84% and of progressive disease (PD) of 10%–35%. What was however concerning was the number of patients who defaulted in two of the studies. In the study by Penel et al44, 50% of patients defaulted due to: disease progression (9), haematological toxicity (4), refusal of treatment (6), and investigator’s decision. Heinrich et al45 had a 92% default rate due unsatisfactory results; this study however included 186 different malignancies with imatinib-sensitive tyrosine kinases or PDGFRα. To date, these are not necessarily the targeting receptor on those tumours.

Anti-oestrogen therapy
Bocale et al50 published a systematic review in 2011 evaluating the efficacy of anti-oestrogen therapy. They evaluated 168 patients. Forty-one articles were reviewed. The anti-oestrogen therapy treatment was started as first-line treatment in 92 patients for a period of 9 months and in 34 patients with recurrent disease following surgery; they were unable to determine if it was started as first-line treatment or recurrence in 42 patients. They found the following:

• Outcome of anti-oestrogen therapy in FAP/Gardner’s syndrome and desmoid tumour as shown in Table III.
• Monotherapy with anti-oestrogen had a significantly higher CR/PR than those treated with anti-oestrogens and NSAIDs.
• Tamoxifen and NSAID had a 35% response rate versus 58% in those treated with tamoxifen alone; they however had a higher stabilisation rate. The authors came to the conclusion that this could be biased as more aggressive desmoid tumours might have been treated with combination treatment rather than monotherapy.
• Tamoxifen and toremifene are equally effective.

Anti-oestrogen treatment is definitely an option for systemic therapy in desmoid tumours. It has fewer side effects than the other systemic drugs and is tolerated better by the patients.

Hong et al49 did a study on mice and came to the conclusion that testosterone regulates the β-catenin level and cell proliferation and that anti-testosterone might be effective in the treatment of aggressive fibromatosis. This however needs to be further investigated with a prospective human trial.

Chemotherapy
The French Sarcoma Group Garbay et al52 did a retrospective study of 62 patients who had chemotherapy. Forty-four patients had combination chemotherapy and 18 monotherapy of which 13 patients had an anthracycline-containing regimen. They found a higher response rate in the anthracycline group versus the nonanthracycline-containing group: 54% versus 12% (p=0.0011). Forty-eight per cent did not need any other treatment intervention after chemotherapy and the most commonly used regimen was methotrexate-vinblastine.

A retrospective study on 39 patients by Constantinidou et al54 found similar results (Table IV).

Chemotherapy can have severe side effects such as: mucositis, vomiting, neutropaenia, neurotoxicity, cardio toxicity (irreversible) and treatment-induced malignancy.55,56 Though chemotherapeutic agents are a possibility for the treatment of desmoid tumours, the significant morbidity due to the side effects of this treatment modality have to be considered. The benefit must outweigh the risk.

Table IV

<table>
<thead>
<tr>
<th>Disease</th>
<th>Number of patients</th>
<th>CR/PR</th>
<th>SD</th>
<th>PD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Garbay et al52</td>
<td>62</td>
<td>1.6%/19.4%</td>
<td>59.6%</td>
<td>19.4%</td>
</tr>
<tr>
<td>Constantinidou et al54</td>
<td>39</td>
<td>--- / 11%</td>
<td>60%</td>
<td>22%</td>
</tr>
</tbody>
</table>

CR: Complete Response, PR: Partial Response, SD: Stable Disease, PD: Progressive Disease

NSAIDs
COX-2 is involved in the cell proliferation due to β-catenin stabilisation in desmoid tumours.57 Nishida et al58 did a prospective and consecutive case-control trial where they treated 22 patients with meloxicam (10 mg daily) for a median period of 20 months. All the specimens showed a strong positive staining with COX-2 on immunohistochemistry. Two patients defaulted due to gastritis, pneumonia or diarrhoea. Of the 20 patients who continued with the study, 95% had a good outcome (stable disease or better) (complete response: 1; partial response: 7, progressive disease: 1; not determined: 2; stable disease: 11). This was however a small prospective study and a bigger randomised trial will be ideal in order to evaluate the above results adequately.

Anti-oestrogen treatment is an option for systemic therapy in desmoid tumours. It has fewer side effects than the other systemic drugs and is better tolerated.
Conclusion

After reviewing the literature and evaluating the outcome of our patients the following treatment regimen was decided upon:

- The ‘wait-and-see’ principle is valid in a patient who is asymptomatic, stable and with non-progressive disease. They have to be followed up closely every 3–6 months to monitor the disease progress. Rather intervene if there is any doubt regarding the disease progression.
- Surgery has a high recurrence rate, whether a positive or negative surgical margin is achieved. Radiotherapy in a patient with a positive surgical margin would not necessarily have changed the outcome of our patients. You have to take the side effects of radiation into consideration before exposing your patient to this modality as these side effects will also limit the success of your surgical intervention, if needed, later (e.g. wound breakdown).
- Radiation therapy can be considered in patients with large tumours causing functional incapacity, neurovascular involvement or where surgery will cause severe morbidity (unresectable tumour).
- Systemic therapy can be given in any recurrent tumour, unresectable tumour, progressive disease or where the least-invasive treatment is to be used as first-line treatment.
- Radical surgery for an unresectable tumour is rarely needed and radiotherapy or systemic therapy should be considered first.

The most important concept is to individualise each patient and to have a multidisciplinary team approach involving the surgeon, radiation and medical oncologist, pathologist, radiologist, psychologist, occupational therapist, physiotherapist and most importantly the patient’s opinion.

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