CASE REPORT

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Preventing neurovascular invasion in desmoid tumors

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Desmoid tumors or aggressive fibromatoses are rare, non-encapsulated, infiltrative and locally aggressive tumors originating from deep musculo-aponeurotic structures. Traditionally, preferred treatment method for desmoid tumors is wide local excision. Depending on the side and type of resection, the reported local recurrence rates range from 15 to 77%. Similarly, in our institution there is a significant recurrence rate (24%) in patients who underwent surgery for desmoid tumor. After several recurrences, amputation may be inevitable following repeating vascular and nerve reconstructions. There is a need for a nonviable barrier in order to prevent the invasion of the viable tumor to the neurovascular structures which are also viable tissues. Depending on this need, we present two cases that we used synthetic vascular graft in their operations to cover neurovascular structures in order to prevent tumor invasion. For patients who are not suitable for radiotherapy and the neurovascular structures need to be secured because of the risk of local recurrence, this method can prevent possible future invasion of vessels and nerves.

Key words: Desmoid tumor; graft; neurovascular invasion.

Desmoid tumors or aggressive fibromatoses are rare, non-encapsulated, infiltrative and locally aggressive tumors originating from deep musculo-aponeurotic structures.[1,2] However, they lack the cytological features of malignancy and have essentially no metastatic potential.[3-6] Most extremity desmoids are located in the soft tissues of the lower and upper extremities and are frequently observed in the foot, calf, thigh, arm and shoulder.[7,8] The lesions are often painful because of their proximity to muscles and especially vessels and nerves. Although they may be seen on standard X-rays, MRI is very useful and most of the time they are found to be hypointense on T1 and either hypointense or hyperintense on T2.[7-9]

Traditionally, preferred treatment method for desmoid tumors is wide local excision.[6,10-12] Depending on the side and type of resection, the reported local recurrence rates range from 15 to 77%.[2,12-16] Over the past 10 years, adjuvant therapy employing radiation therapy, chemotherapy, anti-estrogenic agents such as tamoxifen and even NSAIDs have been used in an attempt to reduce the risk of local recurrence.[5,15,17]

Many reports have emphasized that the margin status is the single most significant determinant of recurrence in patients treated surgically.[6,8,12,18] However, some authors including the authors of this report believe that not only surgical margin but also localization of tumor, age, gender, or adjuvant radiotherapy affect disease-free survival.[19-22] In case of the local recurrence of tumor, dissection of neurovascular structures is difficult because of tumor invasion and sometimes treatment may result with amputation after several local recurrences.

The aim of this paper is to present two cases whose recurrence were treated with wide resection where syn-

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thetic vascular graft was used as a new and easy method to prevent neurovascular structures in local recurrence of desmoid tumors.

Case reports

Between 1990 and 2010, 54 patients were admitted to our clinic with extra abdominal desmoid tumors. Eight patients had recurrent tumor. Wide margins were achieved in 46 patients while the remaining three had microscopic and five had macroscopic residual disease following resection. Vascular reconstruction was performed in three patients. Thirteen patients (24%) relapsed after being treated in our institution. All patients were treated with excision as wide as possible for recurrent tumor. Four patients received postoperative radiotherapy after surgery for recurrent disease. Considering surgery of recurrent tumors, there is an increased risk of neurovascular reconstruction or sacrification due to the involvement of neurovascular structures compared to primary surgery. Therefore, we applied synthetic vascular graft to cover neurovascular structures in order to prevent tumor invasion in three patients. We could avoid the invasion of the vessels and nerves in two patients who had a relapse after the installation of the synthetic vascular graft.

Case 1

A 13-year-old female presented with a three week history of a swelling associated with a dull, aching pain over the back of right thigh after falling. At the clinical examination, patient had a hard, immobile and irregular, huge mass located over the thigh. Neurovascular examination was normal. The patient had no other complaints. Plain radiograms and MRI were obtained. MRI showed a well defined irregular tumor that was firmly attached to the periosteum of the proximal femur and measured 8×9×17 cm (Fig. 1a). Diagnosis was confirmed by tru-cut biopsy. It had also infiltrated to biceps and semitendinosus muscles and ischial nerve. The

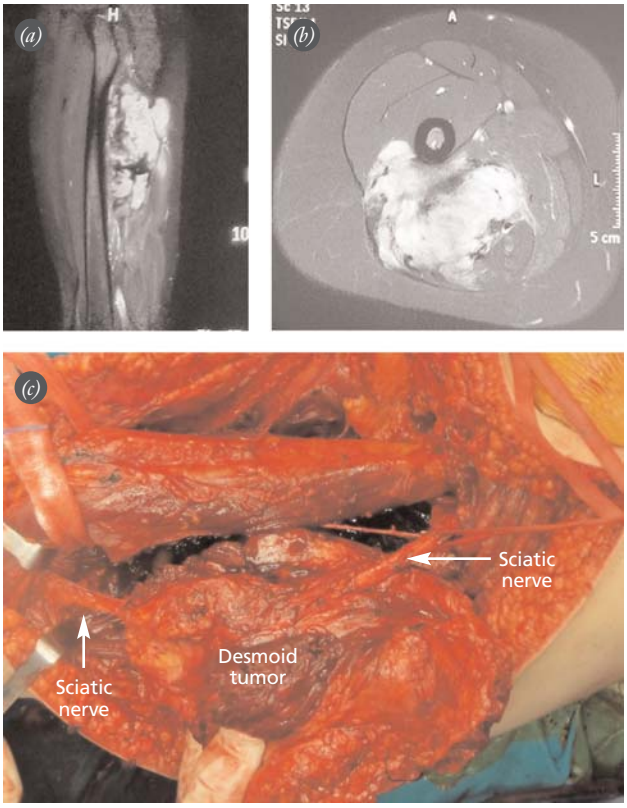

Fig. 1. Preoperative sagittal **(a)** and axial **(b)** MRI views showed irregular tumor that was firmly attached to the periosteum of the proximal femur and measured 8×9×17 cm. **(c)** Desmoid tumor and its relationship with sciatic nerve at the posterior thigh. Tumor was excised by sciatic nerve dissection. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

tumor was excised widely with sciatic nerve dissection (Fig. 1b) and sciatic nerve was covered with synthetic vascular graft to protect tumor invasion (Figs. 2a and b). At the second year follow up after the operation, recurrence was detected. Recurrent tumor was excised widely with removal synthetic vascular graft without neurovascular structure dissection. At recurrence surgery, it

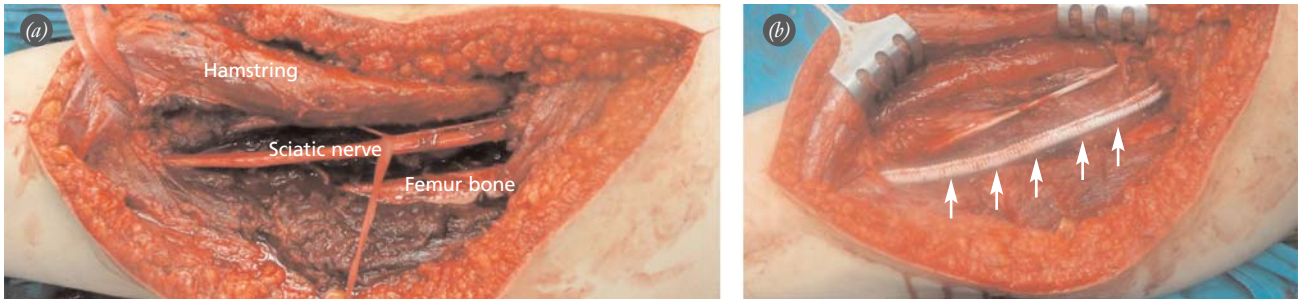

Fig. 2. Clinical view of the posterior thigh after tumor resection. **(a)** Sciatic nerve and femur bone are seen on the figure. **(b)** Clinical view of the sciatic nerve that was covered with synthetic vascular graft (arrows). [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

seemed that synthetic vascular graft was attacked by tumor (Figs. 3 and 4) and graft that cover sciatic nerve was blocked tumor invasion. Graft was removed with tumor mass without any nerve injury. There were no nerve and graft attachment. After obtain wide resection, synthetic vascular graft was covered neurovascular structure to prevent tumor invasion at recurrence.

Case 2

An 11-year-old male presented with a two-month history of a slowly-growing swelling associated with a dull, aching pain over the popliteal fossa of the right leg. At the clinical examination a huge hard, immobile, and irregular mass was found to be located over the popliteal region. The patient had no other complaints. Plain radiograms and MRI were obtained. In MRI imaging, a well defined irregular tumor that measured 8×9×8 cm was firmly attached to the periosteum of the dorsal popliteal plane of the femur. Longitudinally, the tumor extended from the division of the ischial nerve to the medial part of posterior capsule of the knee joint (Fig. 5a). At first operation tumor was excised with poplietal artery and vein dissection; however, common peroneal nerve was sacrificed because of tumor invasion that not dissected. Tibialis posterior nerve and poplietal artery and vein were covered with synthetic graft separately (Fig. 5b). At the follow up, recurrent tumor was detected after postoperative one year. MRI showed that tumor was so close to synthetic graft but it could not invade the neurovascular structure again (Fig. 6). At the recurrence surgery, popliteal artery and tibialis posterior nerve were seen to be preserved from tumor invasion by the help of the synthetic vascular graft which was installed at the previous operation. Tumor was dissected from graft surface without graft resection (Fig. 7). As a result, there was no need for vascular resection. During three years of follow-up, the patient had no complaints and there was no evidence of tumor recurrence clinically or radiologically.

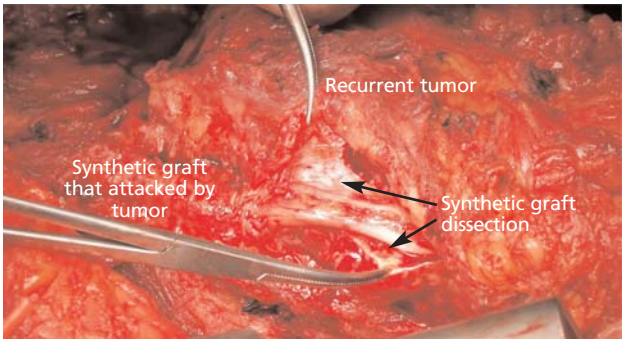

Fig. 3. Recurrent tumor resection. Figure showing synthetic graft attacked by tumor and its removal. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

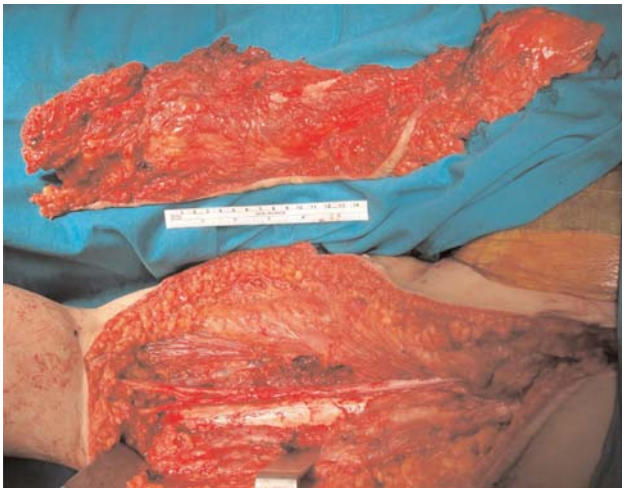

Fig. 4. Recurrent tumor resection material (top) and posterior view of thigh after resection (below). [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

Discussion

Extra-abdominal desmoid tumors cause significant and often serious problems for the patient, the orthopedist and the oncologist.[8] However, despite the adequate

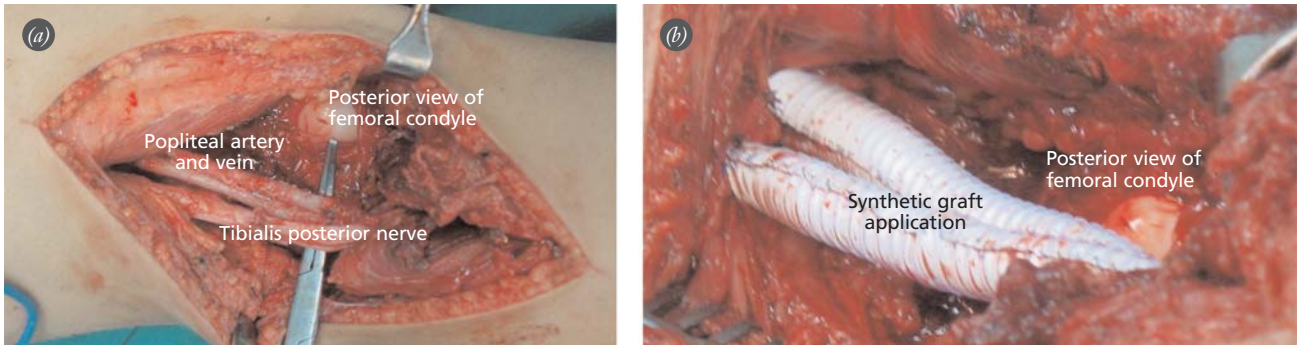

Fig. 5. **(a)** Clinical view after tumor resection located at popliteal region. **(b)** Tibialis posterior nerve and popliteal artery and vein were covered with synthetic graft separately. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

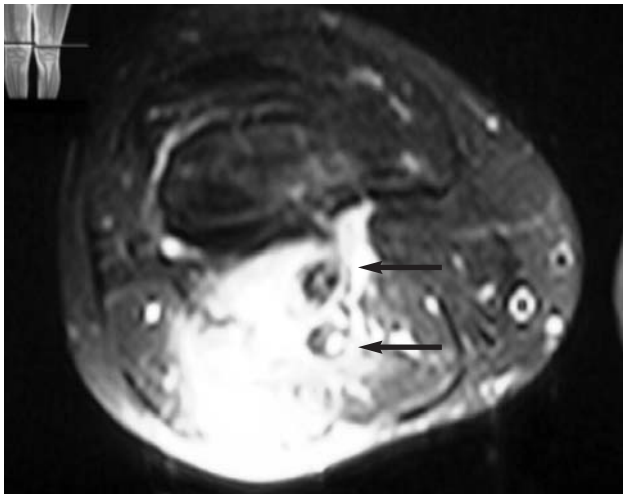

Fig. 6. Local recurrence is shown in the same area in axial T2 weighted MRI image. Arrows show synthetic graft.

Fig. 7. The recurrent tumor is dissected at the second operation, and there is no invasion of vascular structures which are protected by the synthetic graft. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

margins, local recurrence is a significant problem that has been reported to range from 25 to 77% in ten years.[12,14,15] Similarly, we met a significant recurrence rate (24%) in patients who underwent surgery for desmoid tumor. Surgical excision is the basis of treatment in both the primary and relapsed settings. Wide resections with negative margins have generally been correlated with lower recurrence. Many studies have found external radiotherapy to be helpful in the management of this disease. However, patients with open physis are not suitable for radiotherapy.[20-24] Patients have to learn to live together with tumor recurrences. After several recurrences, amputation is inevitable following recurrent vascular and nerve invasions and reconstructions. Both at the initial and recurrence surgery, neurovascular invasion by the tumor increases the morbidity of surgical treatment. Also, as nerve reconstruction is not done, the loss of functionality is at a higher rate. For patients in whom radiotherapy can't be used and local recurrence is expected, neurovascular structures can be secured from tumor invasion with this method.

There is a need of a nonviable barrier in order to prevent the invasion of the viable tumor to the vessels and nerves, which are also viable tissues. Depending on this need, we found that covering the vessel with a nonviable vein graft is applicable. We applied this technique in three patients, and could prevent the invasion of the neurovascular structures in two patients who had a relapse after the installation of the synthetic vascular graft, which serves as a nonviable barrier between the tumor and neurovascular structure.

Invasion of neurovascular structures by desmoid tumor affects the morbidity rate of the disease. In fact we can protect neurovascular structures against tumor invasion by synthetic graft coverage. This method decreases morbidity rate but it is not effective on mortality rate. Today, we cannot change desmoid tumor biology with further treatment but we can protect vital neurovascular structures against by desmoid tumor invasion with our technique which acts as synthetic barrier.

For patients in whom radiotherapy cannot be used and local recurrence is expected, neurovascular structures can be secured from tumor invasion by this method.

Conflicts of Interest: No conflicts declared.

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