Desmoid Tumor

One of the more common benign soft tissue tumors is the extra-abdominal desmoid tumor, sometimes referred to as aggressive fibromatosis or simply desmoid tumor. As compared to the multitude of different clinical types of infantile fibromatosis, the desmoid tumor occurs in older children or young adults up to the age of 40 years. The abdominal desmoid tumor is usually seen in young adult females following pregnancy, whereas the extra-abdominal desmoid tumor is found more commonly in men, usually in proximal parts of the body such as the shoulder, buttock or posterior thigh area. Other areas include the popliteal space, the arm and forearm. Although they are usually solitary lesions, multicentric involvement can occur. In "Gardner's syndrome", the desmoid tumor can be associated with polyposis of the large bowel and cranial-facial osteomas. Desmoid tumors originate in muscle fascial planes and tend to infiltrate into surrounding tissue with poorly defined margins that make surgical resection difficult. They can also occur in tendon sheathes, joint capsules and even inside bone. Clinically, the desmoid tumor has the appearance of a low-grade fibrosarcoma but tends to be more invasive in character and can engulf adjacent neurovascular structures. The histological appearance of the desmoid tumor is similar to a low-grade fibrosarcoma but will never metastasize to distance parts, however, it can be multifocal in its presentation in the extremity. Clinically, the desmoid tumor is very firm to palpation. It frequently causes pain and is tender on deep palpation. Microscopically, the tumor is heavily collagenized but with a very low mitotic index. Its appearance is similar to plantar fibromatosis.

The desmoid tumor does not calcify but is radiodense on ordinary radiographs and can be easily identified on a soft tissue window CT scan. The MRI is the best imaging study for this tumor. It will be low signal on the T1-weighted images but only intermediately high signal on the T2 because of the low water content in this tumor which helps separate it from malignant MFH and fibrosarcoma. The desmoid tumor is related to physical activity whereas the higher grade spindle cell tumors are more spontaneous in origin.

Treatment consists of a wide surgical resection, if possible. Because of the high recurrence rate (about 50 per cent), most clinicians advise postoperative radiation therapy to the extent of 5000 centigray to the surgical site starting two weeks after surgery. This reduces to 10-15 percent the chance of local recurrence. In rare instances, where the patient has multiple local recurrences following radiation therapy, amputation may be indicated. Even with amputation, the

tumor can recur at the amputation site. In patients over 40 years of age, it is not unusual to see a spontaneous involution of the tumor requiring no treatment whatsoever. Estrogen may play a role in the development of desmoid tumors and this accounts for the use of tamoxifen with some reported clinical benefit. Other adjuvant therapies include nonsteroidal anti-inflammtory agents and even the use of mild chemotherapeutic agents has proven beneficial in certain cases.