Tumors of Muscles
and Synovial Tissues

Tapas K. Das Gupta, M.D., Ph.D.
and Richard D. Brasfield, M.D.

Tumors arising in muscle and synovium are relatively rare and clinical diagnosis is difficult unless the examining physician is alert to these lesions. The Table on page 380 shows the common benign and malignant forms of tumors arising in muscle and synovium.

**Tumors of Smooth Muscle**

**LEIOMYOMA**

Leiomyomas are extremely rare outside the gastrointestinal tract and uterus. The occasional occurrence of leiomyoma in the somatic tissues is usually in the skin and the subcutaneous tissues. Stout and Lattes pointed out that these leiomyomas can be either superficial or vascular.1 Superficial leiomyomas arise from the arrectores pilorum muscles. The vascular leiomyomas which arise from the smooth muscle of the small vessels can be either single or multiple and are often very vascular. Biologically all leiomyomas behave benignly though in extremely rare instances recurrence has been recorded.

Quan and Berg reported on the natural history of leiomyomas of the gastrointestinal tract in a series of 57 patients.2 Forty-four leiomyomas were found in the stomach, four in the small bowel, three in the colon and six in the rectum.

---

Dr. Das Gupta is Professor of Surgery and Head, Division of Surgical Oncology, University of Illinois, Abraham Lincoln School of Medicine, Chicago, Illinois.

Dr. Brasfield was Associate Attending Surgeon, Gastric and Mixed Tumor Service, Department of Surgery, Memorial Hospital for Cancer and Allied Diseases, New York, New York. He died May 4, 1970.

It was found that if a diagnosis is accurately established these can be considered truly benign tumors which do not require aggressive therapy.

**LEIOMYOSARCOMA**

Like their benign counterparts, leiomyosarcomas are relatively rare tumors; however, they occur more often in somatic tissue than do benign leiomyomas. These tumors develop in the smooth muscle coats of major arteries and veins in all areas of the human body.3 They are also found in the gastrointestinal tract, uterus, urinary bladder and prostate in young children. In one series of 119 leiomyomatous tumors, 62 leiomyosarcomas occurred in the gastrointestinal tract.2
TUMORS OF MUSCLE AND SYNOVIAL TISSUE

| Tissue of Origin | Benign Tumors            | Malignant Tumors                  |
|------------------|--------------------------|-----------------------------------|
| Smooth muscle    | Leiomyoma                | Leiomyosarcoma                     |
| Striated muscle  | Rhabdomyoma              | Rhabdomyosarcoma                   |
| Synovial tissue  | Ganglion                 | Malignant Synovioma                |
|                  | Giant cell tumor of      |                                   |
|                  |  tendon sheath           |                                   |
|                  | Synovial xanthoma        |                                   |

Tumors of Striated Skeletal Muscle

Tumors of the striated muscles were first described in 1854. A similar type of muscle tumor has also been recorded in fishes, birds and rodents. These tumors can be either benign (rhabdomyoma) or malignant (rhabdomyosarcoma). (See Table.)

Rhabdomyoma

This is an extremely rare form of tumor. Stout and Lattes found ap-
Fig. 4. Embryonal rhabdomyosarcoma in the upper end of the anterior thigh in a two-year-old girl.

Fig. 5. Embryonal rhabdomyosarcoma in the lower end of thigh in a 25-year-old man.

Fig. 6. Embryonal rhabdomyosarcoma in the scapular region of a 10-year-old boy.

approximately 12 such recorded cases, all located in the tongue, neck muscles, larynx, eurila, nasal cavity, axilla or vulva. A special variant of these tumors can be found in the heart muscle. These tumors are entirely benign and are treated accordingly.

**Rhabdomyosarcoma**

Sarcomas composed exclusively of rhabdomyoblasts are of two types. One is usually seen in children and is commonly known as embryonal rhabdomyosarcoma; the other usually occurs in adults and is designated as pleomorphic rhabdomyosarcoma.

**Embryonal Rhabdomyosarcoma**

Although this variety of rhabdomyosarcoma may be found in all age groups, it occurs predominantly in patients 15 years of age or younger. Embryonal rhabdomyosarcoma is also known as juvenile rhabdomyosarcoma, sarcoma botryoides or alveolar rhabdomyosarcoma. Rhabdomyosarcomas that develop near the mucosal surface in various sites such as the urogenital tract, pharynx, orbit, nasal cavity, the auditory canal, etc., tend to be lobulated and myxoid, with the surface resembling a bunch of grapes (which accounts for the term sarcoma botryoides). The microscopic appearance of these types of rhabdomyosarcoma is not germane to this clinical presentation.

Embryonal rhabdomyosarcomas can develop in any area of the body (Figs. 4, 5 and 6), but are found predomi-
nantly in the head and neck region. Although these tumors usually metastasize through the blood stream, about 25 percent also show evidence of regional lymphatic metastases.

The treatment of these tumors has undergone some changes with the expanded use of chemotherapy. At the present time tumors located in the trunk and extremities are usually treated by wide excision in conjunction with sequential chemotherapy. In selected cases supplemental radiation therapy is tried. Excision must be radical and in some cases may mean amputation, although tumors located in the head and neck region may be adequately controlled by judicious use of radiation therapy and radical excision is not always indicated.

The prognosis for this tumor is not favorable and only a handful of patients can be expected to survive. However, it must be pointed out that tumors located in the head and neck region behave less aggressively than those in the trunk or extremities. With the use of sequential chemotherapy it is possible that the present overall survival in 35 percent of these patients might be considerably improved.

Pleomorphic Rhabdomyosarcoma

This type is also known as adult rhabdomyosarcoma. Like the embryonal type, pleomorphic tumors can occasionally arise in children under 15 years of age. The majority of these tumors are found in the trunk and extremities and usually are deep-seated. (Fig. 8.) The microscopic appearance of these tumors varies greatly and recognition is not easy. Stout and Lattes emphasized the necessity of searching for rhabdomyoblasts, which can assume several shapes and sizes. Good fixation and careful staining are essential for an accurate diagnosis.

The tumors spread locally, and inadequate excision of the primary tumors is usually associated with local recurrence. (Figs. 9 and 10.) Metastases commonly occur through the bloodstream and again, as in cases of embryonal rhabdomyosarcoma, regional nodal metastases are frequent. In fact, pleomorphic rhabdomyosarcoma has a higher incidence of regional nodal metastases than any other adult soft tissue sarcoma.

The ideal treatment of pleomorphic rhabdomyosarcoma is adequate wide excision and, when indicated, amputa-
tion of the extremity. Unlike its embryonal counterpart, radiation therapy and/or chemotherapy do not significantly affect the prognosis.

The survival rate is poor; Keyhani and Booher reported an over-all five-year survival rate of 29.4 percent. Most authors have reported a similar low salvage in the adult type of rhabdomyosarcoma.

**Tumors Arising in the Synovial Tissue**

Synovial tissue is composed of an inner lining of polyhedral cells and an outer layer of spindle cells of fibrous tissue. Synovial tissue modifies itself according to function and location. Furthermore, in some areas the tissue is constantly exposed to trauma and as a result some degenerative changes are more common in specific locations. Despite the variations in synovial tissue all synovial tumors, whether arising from joint, tendon sheath or bursa, have the same parent tissue and possess a similar natural history.

**Ganglion**

A ganglion is a cystic tumor-like lesion which is found attached to a tendon sheath, and is composed partly of myxoid tissue. The true incidence of ganglions is not known, because many are treated without being reported.

Ganglions most frequently occur on the dorsum of the wrist, arising from the tendon sheaths of the extensor tendons. No instance of malignant degeneration has been noted and simple excision is the preferred treatment.

**Giant Cell Tumor of Tendon Sheath**

These tumors are a type of fibrous histiocytoma. They are quite variable in growth rate and appearance and are usually found on the hands and feet. (Fig. 11.) When they are attached to a joint capsule or a tendon sheath they form a tumor mass. Frequently
Fig. 12. Synovial sarcoma in the medial aspect of the knee in a 40-year-old man. The tumor was originally diagnosed as thrombosed varicose vein.

Fig. 13. Locally recurrent synovial sarcoma in the dorsum of the foot in a 34-year-old woman.

Fig. 14. Synovial sarcoma in the index finger of a 50-year-old man. This was painful and thought to be an infection. Incision and drainage had been attempted.

Fig. 15. Synovial sarcoma in the left shoulder joint area, which is an uncommon site.

these lesions are also designated as villonodular synovitis. Although local excision is usually sufficient treatment, these tumors sometimes recur despite apparent adequate excision.

SYNOVIAL SARCOMA

This rare form of sarcoma commonly occurs in the extremities, with the lower extremity more frequently involved than the upper. Synovial sarcomas appear in the knee joint more often than in all other joints, with the foot and hand, respectively, second and third in frequency. (Figs. 12-15.)

Grossly, the tumor may be either encapsulated, circumscribed with a pseudocapsule, or diffuse, infiltrating the surrounding structures and producing distortion of the adjacent tissues. The size varies with the stage and duration of the disease. Although synovial sarcomas may assume huge proportions, they are usually not very large, varying from 3 to 6 cm. in maximum diameter. Occasionally they may show spotty areas of calcification. The microscopic pattern of synovial sarcomas is very striking; they are usually composed of what may be termed synovioblastic elements. However, some of these tumors may have a preponderance of fibroblastic elements, which makes accurate histologic diagnosis difficult.
Synovial sarcomas are first seen as a mass which may or may not be painful, although the majority are painless. The painful masses, particularly in the finger, are often misdiagnosed as infections and only microscopic examination will give the diagnosis. (Fig. 14.)

Synovial sarcomas, like rhabdomyosarcomas, have a high tendency to metastasize to the regional lymph nodes, a fact which must be considered when planning therapy. The bloodstream is the major route of distant metastasis. The lung is most commonly involved although pancreatic metastases are frequent and there is also a high incidence of bone metastases.

The generally accepted form of treatment for synovial sarcoma is wide excision or major amputation. Irradiation alone or in conjunction with chemotherapy is not curative.

The survival rate following complete excision of the primary tumor before any metastases have taken place is difficult to assess, because most reports have so few patients. Pack and Ariel found that about 30 percent of their patients survived for five years. In our opinion, based on our own experience in a small number of patients with localized synovial sarcoma, 25 to 35 percent of patients were free of disease at five years.

References
1. Stout, A. P., and Latessa, R.: Tumors of the Soft Tissue. Second Series, Fascicle 1. Washington, D.C., Armed Forces Institute of Pathology, 1967. 196 p.
2. Quan, S. H. Q., and Berg, J. W.: Leiomyoma and leiomyosarcoma of the rectum. Dis. Colon Rectum 5: 111-120, 1962.
3. Stout, A. P., and Hill, W. T.: Leiomyosarcoma of the superficial soft tissues. Cancer 11: 844-854, 1958.
4. Pack, G. T., and Ariel, I. M.: Tumors of the soft somatic tissues. New York, Hoeber and Harper, 1958. 820 p.
5. Sutow, W. W.: Chemotherapeutic management of childhood rhabdomyosarcoma. In: Neoplasia in Childhood. M. D. Anderson Hospital and Tumor Institute, Twelfth Annual Clinical Conference on Cancer, Houston, 1967. Chicago, Year Book Medical Publishers, 1969. Pp. 201-208.
6. Sutow, W. W.; Sullivan, M. P.; Ried, H. L.; Taylor, H. G., and Griffith, K. M.: Prognosis in childhood rhabdomyosarcoma. Cancer 25: 1841-1890, 1970.
7. Keyhani, A., and Booher, R. J.: Pleomorphic rhabdomyosarcoma. Cancer 21: 956-967, 1968.