A soft tissue sarcoma arising in the blood and/or lymph vessels is relatively rare; yet, when it occurs, this type of tumor may present a challenge to the physician. Since the vast majority of such sarcomas are benign blood vessel hemangiomas, which seldom require any treatment, when a true malignant hemangioendothelioma is encountered, the physician may be at a loss in deciding on either management or prognosis. To counter this, some general findings in blood and lymph vessel tumors are discussed below. Remember, however, that none of these sarcomas can be positively identified without proper histological examination—the essence of good management.

**Tumors of the Blood Vessels**

Blood vessel tumors constitute one of the most frequently seen soft tissue neoplasms. According to Pack and LeFevre, they comprise 46 percent of all soft tissue tumors in their series. Of note, is that this figure represents only those neoplasms arising in the blood vessels and not those tumors in which a preponderance of blood vessels are found. It’s encouraging, however, that the vast majority of such tumors are benign.

| Clinically Important Blood Vessel Tumors |
|-----------------------------------------|
| **Benign tumors**                       |
| 1. Hemangioma                           |
| 2. Angiomatosis                         |
| 3. Rendu-Osler-Weber’s disease and other types of anomalies |
| 4. Glomus tumor                         |
| **Malignant tumors**                    |
| 1. Angiosarcoma                         |
| 2. Kaposi’s sarcoma                     |
| 3. Hemangiopericytoma                   |

**Benign Tumors**

_**Hemangioma:**_ Sharp distinction must be made between a hemangioma and a varicosity or aneurysmal dilation of a pre-existing blood vessel. This clinical distinction is based largely on the index of suspicion: in a varicosity, one generally visualizes multiple dilations of a vessel, usually of long-standing duration; in an aneurysmal dilation, one can

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always locate a fusiform or sacular dilatation, usually pulsatile, with no evidence of surrounding soft tissue sarcoma.

The exact origin of a hemangioma has aroused certain controversy. According to some embryologists, a hemangioma is an embryonic sequestration of unipotent angioblastic cells. These clusters of cells remain discrete while other cells develop to form mature blood vessels. In contrast, other authors argue that a hemangioma is due to elongation of the vessel comprising the tumor, within a more or less closed territory or capsule, without involvement of the surrounding vessels. Since it is now generally accepted that all circumscribed hemangiomas— with the possible exceptions of cirrhotic or racemos types—possess only one efferent and one afferent vessel, this concept of elongation is certainly acceptable for the majority of circumscribed lesions. However, in rare instances of angiomatosis of an extremity or other part of the body (systemic angiomatosis), the first hypothesis must be taken into consideration.

If a tumor is encountered in infancy or childhood, a hemangioma will be suspected since it is the most common tumor found in this age group. (Seventy-five percent of hemangiomas are evident at birth, and the remainder appear in early infancy.) In one of five patients, hemangiomas are multiple and occur preponderantly in females with about 50 percent occurring in the skin of the head and neck.

Although hemangiomas usually bleed when incised, the majority of circumscribed tumors can be removed with relative ease by ligation of one, two or, occasionally, three main vessels.

**Capillary Hemangioma:** This tumor—the most common of all hemangiomas—is apparent at birth and grows with the child. It occurs generally in the skin, the face being the most common site, and occasionally, in the mucous membrane, especially the oral mucosa. The capillary hemangioma may grow rapidly and varies in size from minute spots (DeMorgan spots) to a large, flat, port-wine stain. However, the most common type is a circumscribed, sessile, lobulated, bright red tumor. (Fig. 1.) Management requires infinite patience and caution. Any form of aggressive operation or irradiation therapy to these children is condemned.

**Spider Angioma:** The spider angioma, usually found in older patients, is commonly located on the face, arms, fingers or upper trunk; it is extremely rare in the hairy regions of the body. The tiny, red angioma, resembling a spider, is associated with hepatic disease and its high incidence in alcoholics has long been known. A congenital analogue is found in Rendu-Osler-Weber’s disease.

**Sclerosing Hemangioma:** There is no age preponderance for sclerosing hemangioma and although it is frequently seen in the thigh, abdominal wall or the neck, it is quite uniformly distributed. In this type of tumor, the connective tissue proliferation is so excessive that the perivascular and endovascular thickenings form a solid mass. Thus the tumor is firm, noncompressible and
Fig. 2. Sclerosing angioma in a 40-year-old woman. Such lesions are commonly mistaken for various forms of dermatosis.

frequently the lumen of the major feeding vessel cannot be found. (Fig. 2.) Quite often a sclerosing hemangioma is mistaken for a small fibroma, neurofibroma or nonpigmented melanoma.

*Cavernous Hemangioma:* This tumor, most likely a congenital malformation, is seen at any age. The cavernous hemangioma becomes much more dilated than the capillary angioma and presents as a soft, readily compressible mass, frequently extending into the subcutaneous tissues. (Figs. 3-5.) All the visceral hemangiomata are of this type and occasionally attain enormous size. (Figs. 6a and 6b.)

The management of localized hemangiomata is similar to that for a capillary hemangioma—patience and caution.

Usually these tumors are treated either by freezing or by injection of sclerosing fluid; occasionally, surgery or radiation therapy is indicated. Aggressive therapy is never indicated. *

*Rare Forms of Hemangioma:* The systemic hemangioma is a congenital, arteriovenous, aneurysmal anomaly. It presents as a diffused, vascular tumor which may occupy an entire extremity—generally the upper extremity—or portion of the trunk and is evidenced immediately by local gigantism. (Figs. 7a, 7b.)

* The exact principles of therapy will be described in a forthcoming, final section on soft tissue sarcoma.
and 7b.) In some patients, the accompanying developmental problems are of such magnitude that little can be done.

Telangiectasias, and other congenital conditions of the blood vessels analogous to tumors of blood vessel origin, are so rare as to be beyond the scope of this presentation.

Glomus Tumor: The small, benign and rare glomus tumor is usually single (but may be multicentric) in origin. It generally occurs in the skin and subcutaneous tissues of the hands, feet and often the nail bed. The pain associated with the glomus tumor is pathognomonic. Successful treatment usually requires excision.

Malignant Tumors

Angiosarcoma: This rare, malignant tumor of the blood vessels, which can mimic any conceivable tumor, has a diverse distribution in the body, although the breast is one of the more...
common sites. Angiosarcomas are usually solitary, firm, bulky tumors situated deep in the soft tissue. Usual therapy is irradiation; however, in selected cases, excision of the lesion is advisable. The prognosis is poor since these tumors are extremely aggressive.

Kaposi’s Sarcoma: Since its original description by Kaposi in 1872, this tumor has been well documented. Seen frequently in Africa, it is virulent in African children to a degree not seen in this country. In the great majority of patients, this sarcoma originates in the skin of the lower extremities, although it has been described in all parts of the body and is frequently associated with a variety of malignant lymphomas. The appearance is so classic that a diagnosis can be made by inspection alone: the initial lesion appears as a reddened, well demarcated macule or nodule; in advanced cases, multiple, elevated, bluish-purple, angioma-like lesions are found. (Figs. 8a, 8b and 9.)

Irradiation is the treatment of choice for localized Kaposi’s sarcoma. It is gratifying to see localized lesions regress with institution of this mode of therapy. In advanced cases, however, systemic chemotherapy has been of some value.

Hemangiopericytoma: This rare, vascular tumor composed of capillaries surrounded by peculiar, elongated, contractible cells known as Zimmermann’s pericytes, was first described by Stout and Murray in 1942. Observed in all age groups, a hemangiopericytoma occurs wherever capillaries are found. Usually, it is painless, in contrast to its...
possible counterpart, the painful, benign glomus tumor. The hemangiopericytoma tends to recur locally and metastases occur via both the bloodstream and lymphatics. O'Brien and Brasfield reported their experience in 24 patients with hemangiopericytoma, and concluded that these tumors are highly malignant, not in a five-year concept, but over a lifetime. Wide excision is the best method of treatment.

Tumors of the Lymph Channels

The clinically important tumors arising in the lymph vessels are the benign lymphangioma and its malignant counterpart, the lymphangiosarcoma.

*Lymphangioma:* This tumor occurs much less frequently than does the hemangioma and can be found anywhere on the body surface. The lymphangioma, a large sac-like dilatation, may be either papillary or cavernous; however, the most frequently encountered clinical problem is the cystic hygroma of the neck found in infants. (Fig. 10.) Although excision is the best method of treatment in all types of lymphangioma, treat the cystic hygroma with caution. In uncomplicated and asymptomatic patients the infant should be allowed to
Fig. 11. Lymphangiosarcoma of the forearm of a patient with postradical mastectomy edema. The lesions developed 14 years after surgery.

Fig. 12a. Lymphangiosarcoma developing in congenital edema of the upper extremity. The patient was 36 years old when the initial diagnosis was made.

Fig. 12b. Same patient as in Fig. 12a showing the original excision with recurrence.

Fig. 12c. The cut section of the upper extremity from the same patient as in Figs. 12a and 12b. Note the marked hemorrhagic foci of the tumors extending to the subcutaneous tissue. He had forequarter amputation three years ago and is still doing well.
grow and excision performed at a later date—not in infancy unless the patient is symptomatic. A number of unfortunate technical mishaps have been reported when a neck dissection has been attempted in such cases.

Edema and lymphangiectasia may develop in certain instances, due either to congenital hypoplasia of the lymph vessels in the extremity or to deliberate disruption of the lymph node basin.

**Lymphangiosarcoma:** Although the more dramatic form of this disease is seen in a postmastectomy edematous arm (Fig. 11.), first described by Stewart and Treves, it must be emphasized that this tumor can occur in any chronically edematous extremity. (Figs. 12a, 12b and 12c.) Both wide excision and irradiation have been used to treat a lymphangiosarcoma, but the end result is poor. Hermann in 1965 studied 91 cases of lymphangiosarcoma and found that by either method the salvage rate was low.

Rarely, tumors such as extraosseous osteogenic sarcoma, extraskeletal chondrosarcoma as well as a number of extranodal soft tissue lymphomas will also be encountered.

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### Announcement

A National Conference on Human Values & Cancer, sponsored by the American Cancer Society, will be held at the Regency Hyatt House, in Atlanta, Georgia, on June 22, 23 and 24, 1972. For additional information, please contact William M. Markel, m.d., National Conference on Human Values & Cancer, American Cancer Society, Inc., 219 East 42nd Street, New York, N. Y. 10017.