

Epithelioid sarcoma is a kind of cancer that usually begins as a lump in the hands, fingers, forearms, lower legs or feet. This fact sheet will discuss the following about epithelioid sarcoma:

- Appearance and characteristics
- Types and stages
- How it is treated
- Emotional concerns when dealing with epithelioid sarcoma



Appearance and Characteristics

A soft tissue sarcoma is a cancer tumor that appears in muscles, ligaments, tendons and other soft tissues in the body.

Epithelioid sarcoma is rare form of soft tissue sarcoma. It most commonly starts under the skin on the finger, hand, wrist or forearm, knee, lower leg or foot. It usually appears as a small, firm growth or lump under the skin.

It does not usually cause pain, but can become painful or tender as it grows and pushes into nearby nerves.

Epithelioid sarcoma is considered slow-growing but aggressive. This means that it is initially slow to develop, but can be hard to remove as it grows and can spread into other parts of the body. It also has a high rate of recurrence, which means it can again.



Types and Stages

The two types of epithelioid sarcoma are:

- Distal type – tumors start in the hands, fingers, forearms, lower legs or feet. These most often affect teens and young adults
- Proximal type – tumors form in areas closer to the core of the body, so in the head, neck, chest, abdomen and genitals. These most often affect adults.

Epithelioid sarcoma is given a stage to indicate how far it has progressed. This is determined through a biopsy.

- Stage I – The tumor is small, low-grade and has not spread to nearby lymph nodes
- Stage II – The tumor is small, but is higher-grade, meaning it is likelier to grow, but still has not spread
- Stage III – The tumor is larger, has grown deeper and often has spread to nearby lymph nodes
- Stage IV – The cancer has spread to distant organs in the body, most often the lungs, or distant lymph nodes



Treatment Options

Because epithelioid sarcoma is very rare, a specialist who knows more about this form of cancer can help your health care team, if one is available.

The common ways of treating epithelioid sarcoma include:

Surgery: Surgery is the most common form of treatment. Your surgeon will remove the entire tumor while working to preserve the way your hand, arm, foot or leg looks and functions. The surgery may also remove surrounding lymph nodes.

Radiation therapy: Radiation beams may be used to shrink tumors before surgery, or after in order to kill any cancer cells that remain.

Chemotherapy: If the cancer has spread, you may need chemotherapy to kill any cancer in other parts of the body. It may also be used if surgery is not an option.

Targeted therapy: Targeted therapy attacks specific chemicals within the cancer cells that help them grow and therefore causes the cancer to die. Tazemetostat (Tazverik) is a common drug used to treat advanced epithelioid sarcoma.



Emotional Concerns

A cancer diagnosis can be very difficult to handle, not just affecting yourself, but for everyone around you. It is important to take care of your feelings. You may find emotional support from friends, family and loved ones. There are also organizations that can help.

CancerCare provides one-on-one counseling and support groups to connect you with others in a safe and supportive environment. We can also help find other resources to help you in many ways.

CancerCare® Can Help

For over 80 years, CancerCare has empowered millions of people affected by cancer through free counseling, resource navigation, support groups, educational resources, advocacy and direct financial assistance. Our oncology social workers improve the lives of people diagnosed with cancer, caregivers, survivors and the bereaved, by addressing their emotional, practical and financial challenges.

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This fact sheet is made possible by Ipsen Biopharmaceuticals, Inc.