ABOUT EPITHELIOID SARCOMA

JUST THE FACTS

Sarcomas are a rare type of cancer that occur in hard tissues like bone or soft tissues like muscle.\(^1\)

Sarcomas that form in soft tissues—like muscle, fat, nerves, fibrous tissues (connecting muscle and bone), blood vessels and deep skin tissues—are called soft tissue sarcomas (STS).\(^1\)

There are more than 50 types of STS, of which epithelioid sarcoma (ES) is one.\(^1\)

ES forms as a hard lump in soft tissue such as under the skin or in the abdomen or groin.\(^2\)

WHO DOES ES AFFECT?

ES is rare and accounts for less than 1% of all STS.\(^3\)

Approximately 150-200 people in the U.S. are newly diagnosed annually with ES.\(^4\)

ES can affect people of all ages and genders. However, ES is more common in men and young adults.\(^5\)

Given the rarity of ES, patients should seek care from an experienced doctor and/or treatment center that specializes in managing STS.

ES OCCURS IN TWO FORMS

**Distal-type:** Classic form of ES and typically affects teenagers and young adults\(^6\)
- Associated with more favorable survival rates\(^8\)
- Typically occurs in the hands, forearms, feet or ankles\(^8\)

**Proximal-type:** Rarer, more aggressive form of ES and mainly affects older adults\(^6\)
- Associated with less favorable survival rates\(^6\)
- Typically occurs in the pelvic area or abdomen\(^7\)

ES SUBTYPING

More than 90% of ES tumors do not express the integrase interactor-1 (INI1) protein, which acts to suppress tumor growth.\(^8\) INI1 loss plays an important role in the diagnosis of ES.\(^8\)

DIAGNOSIS

Before receiving a formal diagnosis of ES, it’s common to receive an incorrect diagnosis (misdiagnosis) for several reasons, including:\(^3\)

- It’s rare and may not be top-of-mind for many doctors (a rare cancer is defined as fewer than 15 new diagnoses per 100,000 people per year, according to the National Cancer Institute)\(^9,10\)
- It looks harmless when it first forms and may be mistaken for non-cancerous conditions which do not require a biopsy\(^11\)
- It often occurs in presumably healthy young adults in their 20s and 30s\(^5\)

## References