Angiosarcoma is a rare, aggressive tumor of the cells that line blood vessels, that comprises 1% of all soft-tissue sarcomas. It carries a poor prognosis if detected in later stages, with a reported 5-year survival rates ranging from 12% to 25% and has a high rate of local recurrence and metastasis. These tumors arise from lymphatic or vascular endothelial cells showing atypia and can grow along vascular channels, sinusoidal or cavernous spaces. They may also appear as poorly organized vessels, solid masses, or nodules. The cause for angiosarcomas is still being studied; however, definitive risk factors that have been identified include history of chronic lymphoedema and prior radiation therapy.

Angiosarcomas can be further classified into cutaneous or visceral angiosarcomas, depending on the primary site of the tumor. They most commonly present as a cutaneous manifestation in elderly white men, with a median age between 60 and 71 years, or on the head and neck. It has also been commonly reported on the chest wall after receiving radiation therapy for breast cancer.

Surgical resection with negative margins followed by radiotherapy is the mainstay of treatment for localized angiosarcoma and is indicative for the best outcome in terms of overall survival. Other treatment options include chemotherapy, such as paclitaxel, for advanced angiosarcoma; however, disease outcome greatly vary and depend on tumor site, size, resectability, and tumor type (i.e. de novo vs radiation induced). Emerging therapeutic modalities are still being studied, most notably investigating angiogenetic agents and vascular permeability factors as they play a prominent role in angiosarcoma etiology.

References: