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Angiosarcomas

What are angiosarcomas?

Angiosarcomas are uncommon malignant tumours (cancers) that arise from the cells that are normally used to make up the walls of the blood or lymphatic vessels. Haemangiosarcomas start in blood vessel walls and lymphangiosarcomas start in lymph vessel walls.

Angiosarcomas may occur in any organ of the body but are more frequently found in skin and soft tissue. They can also originate in the liver, breast, spleen, bone or heart.

What causes angiosarcomas?

The cause of angiosarcomas is usually unknown. The tumours may develop as a complication of a pre-existing condition. Certain patient groups may be at greater risk of developing angiosarcomas, these include:

- Patients with chronic [lymphoedema](#) (accumulation of lymph fluid in the arms) whom have undergone a radical mastectomy for breast cancer (removal of breast and all lymph nodes under the arm)
- Radiotherapy patients (especially those who have been exposed to radiation emitting contrast agent Thorotrast – this agent is no longer used but angiosarcomas have occurred in people decades after exposure)
- Patients with foreign material (such as Dacron, shrapnel, steel, and plastic) in the body
- Patients exposed to environmental agents such as sprays containing [arsenic](#), and vinyl chloride in the plastic industry.

What are the signs and symptoms of angiosarcomas?

The signs and symptoms of angiosarcomas differ according to the location of the tumour. Often symptoms of the disease are not apparent until the tumour is well advanced.

Affected organ	Features
Soft tissue	<ul style="list-style-type: none"> • Rapidly growing tumour in the extremities or abdomen • Abdominal tumours may grow to large sizes before being detected as the abdomen can accommodate tumours • Haemorrhage, anaemia, haematoma, gastrointestinal bleeding • Adjacent lymph nodes enlarged
Skin	<ul style="list-style-type: none"> • Enlarging bruise, a blue-black nodule, or unhealed ulceration • Lesions may bleed and be painful • Angiosarcomas occurring on the head and neck in elderly people are one of the most common forms of cutaneous angiosarcoma
Bone	<ul style="list-style-type: none"> • Tumours may grow on multiple bones of the same extremity • Pain and tenderness of the affected area is common

	<ul style="list-style-type: none"> • Swelling and increased size of the affected limb may be present
Breast	<ul style="list-style-type: none"> • Rapidly enlarging palpable mass without tenderness • Often there is no pain • Tumours often grow deep within breast tissue and cause diffuse breast enlargement with associated bluish skin discolouration
Other organs	<ul style="list-style-type: none"> • Hepatic – non-specific symptoms such as fatigue, weight loss, right upper quadrant pain • Lung – chest pain, bloody sputum, weight loss, cough, difficulty breathing

Cutaneous angiosarcomas

This is the most common form of angiosarcoma not associated with chronic lymphoedema. The disease is primarily located on the head and neck of elderly persons and is also known as Wilson-Jones angiosarcoma, senile angiosarcoma or malignant angioendothelioma.

Clinical features of this form of angiosarcoma are:

- Lesions most commonly appear on the scalp or upper forehead.
- In the early stages of disease lesions may be singular or multifocal, slow spreading patches of livid or dusky red colour, somewhat like an ill-defined bruise.
- Lesions may develop into elevated nodules or plaques that may bleed and ulcerate.
- Lesions grow rapidly and spread, making margins difficult to define.

Angiosarcoma



What is the treatment of angiosarcomas?

Treatment of angiosarcomas is dependent on the location of the angiosarcoma and the extent of the tumour. Treatment includes chemotherapy, surgery, radiotherapy, or a combination of these treatment modalities.

The optimum treatment of cutaneous angiosarcomas has not been defined. These angiosarcomas are difficult to treat because of their multifocal nature and extensive spread pattern. Radical surgery and postoperative radiotherapy is generally used but tumour recurrence is common after treatment. Despite aggressive treatment, prognosis is poor in patients with cutaneous angiosarcomas.

Related information

References:

- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Skin lesions, tumours and cancers](#)

Other websites:

- [Angiosarcoma](#) - emedicine dermatology, the online textbook
- [Skin, Angiosarcoma of the Scalp: Surgical and Nonsurgical Treatment](#) - emedicine dermatology, the online textbook

Books about skin diseases:

See the [DermNet NZ bookstore](#)

Author: Vanessa Ngan, staff writer

DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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