Cancer Association of South Africa (CANSA)

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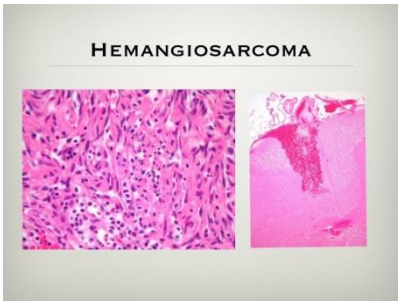
Fact Sheet on Haemangiosarcoma

Introduction

Cancerous (malignant) tumours of the connective tissues are called "sarcomas". Sarcoma arises in the connective tissue of the body. Normal connective tissue include, fat, blood vessels, nerves, bones, muscles, deep skin tissues, and cartilage.



[Picture Credit: Hemangiosarcoma]

Sarcomas are divided into two main groups, bone sarcomas and soft tissue sarcomas. They are further sub-classified based on the type of presumed cell of origin found in the tumour. They all share certain microscopic characteristics and have similar symptoms.

Sarcomas can develop in children and adults. For children under 20 approximately 15 percent of cancer diagnosis are sarcomas. The majority of childhood sarcomas are one of three types. Although they are often called *Paediatric Sarcoma* because of their prevalence in children, they can occur in adults as well. As these cancers are relatively common among childhood cancers, there are relatively standard treatments.

Adolescents and young adults (AYA) with paediatric sarcoma face more than the usual challenge from cancer. In cancer treatment, *adolescents and young adults* are people between age 15 and 39. One of the challenges is that it is studied less than sarcoma in children and older adults. But the major challenge for youth with sarcoma is how to get the best treatment.

Haemangiosarcoma

Haemangiosarcoma is a very aggressive, rapidly growing form of cancer that affects the blood vessels. It can form anywhere in the body, but more than half of the cases originate in the spleen; this condition is also known as "splenic angiosarcoma." This form of cancer can develop in patients of any age, but mainly affects those over 50 years of age.

Haemangiosarcoma of the spleen is a rare tumour. However, it is the most common malignant, primary, non-lymphoid tumour of the spleen. It usually presents itself as a well-defined haemorrhagic nodule, or it involves the spleen diffusely. Clinical symptoms usually are diffuse

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abdominal pain and left upper quadrant mass. Associated clinical findings are anaemia, thrombocytopenia, and coagulopathy. Haemolytic anaemia is caused by damage to erythrocytes from irregularly lined vascular channels in the tumour. The prognosis is poor. Metastases are usually to the liver, lung, and lymph nodes. Survival is usually less than 1 year.

Liu,L., Kakiuchi-Kiyota, S., Arnold, L.L., Johansson, S.L., Wert, D. & Cohen, S.M. 2013. Hemangiosarcomas are uncommon aggressive vascular tumors that have recently become the focus of attention because several chemicals and pharmaceuticals increase their incidence in mice. The relevance of these mouse vascular tumors to humans is unclear. In the present study, we semiquantitatively evaluated the expression profiles of hematopoietic stem cell markers (CD117 [c-kit], CD133, CD34, and CD45), endothelial cell markers (vascular endothelial growth factor receptor 2, CD31, and factor VIII-related antigen), and a myeloid lineage cell marker (CD14) in human hemangiosarcoma (n = 12) and hemangioma (n = 10) specimens using immunohistochemistry. CD133 was completely negative in almost all cases of hemangiosarcomas and hemangiomas. Most hemangiosarcomas, but not hemangiomas, stained for CD117 and CD45. Both groups diffusely expressed CD34, vascular endothelial growth factor receptor 2, and factor VIII-related antigen; however, hemangiomas had more intense and diffuse CD34 and factor VIII-related antigen expression compared with hemangiosarcomas, whereas CD31 was positive in all hemangiosarcomas but only half of the hemangiomas. CD14 staining was negative in most hemangiosarcoma and hemangioma cases. Our results indicate that multipotential bone marrow-derived hematopoietic stem cells or early endothelial progenitor cells (EPCs) expressing CD117, CD34, and CD45 are involved in hemangiosarcoma formation, whereas hemangiomas originate from late EPCs or differentiated endothelial cells, which have lost the expression of most hematopoietic stem cell markers. This contrasts with our previous results that demonstrated that both hemangiosarcomas and hemangiomas in mice may be derived from early EPCs that are not completely differentiated.

Masuzawa, M., Fujimura, T., Hamad, Y., Fujita, Y., Hara, H., Nishiyama, S., Katsuoka, K. Tamauchi, H. & Sakurai, Y. 1999.
"A cell line (ISO-HAS) has been established from tumor tissue of a human hemangiosarcoma arising on the scalp by the use of conditioned medium from a murine-phenotypic angiosarcoma cell line (ISOS-1). Cells have been cultured for more than 2 years with up to 100 passages. The cells retained endothelial-cell properties, such as a characteristic cobblestone appearance at confluency, contact-inhibited growth, active uptake of acetylated low-density lipoprotein labeled with 1,1-dioctadecyl 1,3,3,3,3-tetramethyl-indocarbocyanine perchlorate (DiI-Ac-LDL) and CD31 expression. However, they were weakly positive for von-Willebrand-factor (vWf) antigen and for binding of Ulex europaeus agglutinin-I (UEA-I) lectin, and lacked tube-formation activity. These findings indicate that ISO-HAS is a poorly differentiated endothelial cell line. ISO-HAS cells showed accumulation of p53 protein in the nuclei, and a new-typed p53-gene point mutation was found in exon 7 at codon 240. When inoculated s.c. into severe-combined-immunodeficiency (SCID) mice, the cells showed solid-tumor growth that caused death. These properties suggest that ISO-HAS is a malignant endothelial cell line with high tumorigenicity."

Incidence of Haemangiosarcoma in South Africa

The National Cancer Registry (2016) does not provide information regarding the incidence of Haemangiosarcoma in South Africa.

Causes, Symptoms and Treatment of Haemangiosarcoma

Long-term exposure to certain environmental hazards and chemicals, including arsenic, vinyl chloride, and thorium dioxide, has been linked to a number of cases of haemangiosarcoma. Patients who have been exposed to these types of chemicals - often used in certain types of industry - are at particular risk for developing this form of cancer.

Symptoms are often mistaken for those of other conditions, which can make rapid diagnosis and treatment very difficult. Distinctive symptoms typically include abdominal pain or splenomegaly (an enlargement of the spleen).

Patients with this condition also run a high risk of splenic rupture, which can result in severe blood loss and often death. This condition develops in the spleen and will often spread to the lungs and/or liver.The highly aggressive nature of this form of cancer, and lack of symptoms until it has progressed fairly significantly, makes it very difficult to treat. A physician will usually conduct a thorough physical exam, followed by scans that might include a CT or CAT, MRI, or X-rays.

These scans can help to determine the size and precise location of the mass; an MRI can provide a more detailed image of the tumour that can help to determine the progression of the condition. The size, location, and stage of the tumour will then determine the best course of treatment.

A combination of chemotherapy and a splenectomy, surgery to remove the tumour on the spleen, have been the most successful. A splenectomy itself can give the patient a survival time of 1-3 months; the addition of chemotherapy can extend the survival rate to 5-7 months.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

Medical Disclaimer

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