

(Leiomyosarcoma, liposarcoma, fibrosarcoma, rhabdomyosarcoma, Kaposi's sarcoma)

DEFINITION

1. Cancer is a term which embraces a large number of different diseases, the common feature of which is a malignant tumour. This is a growth (neoplasm) which is not circumscribed but which infiltrates the surrounding tissues and metastasies (spreads to other sites in the body, thereby producing secondary deposits). Any tissue in the body may be affected.
2. Cancers are classified according to the tissue of origin. **Carcinoma** arises from epithelial tissue and **sarcoma** from connective tissue. The suffix - **blastoma** implies a tumour of embryonic origin.
3. **Soft tissue sarcomas** are of varied histological type dependent on tissue of origin. 30% are **leiomyosarcomas** (arising in smooth muscle) which are found mainly in the female uterus and the gastrointestinal tract in the male. **Liposarcomas** (arising in fat), **fibrosarcomas** (fibrous tissue) and **rhabdomyosarcomas** (skeletal muscle) comprise about 40% of the remainder and are predominant in males.
4. A particular type of soft tissue sarcoma is **Kaposi's sarcoma** of which there are 3 forms. The first type is sporadic in distribution and affects elderly people particularly Jews. Another type occurs in relation to AIDS infection and a third variety affecting the legs is seen in young male Africans.

CLINICAL MANIFESTATIONS

5. Soft tissue sarcomas most commonly present as lumps which are usually painless. They sometimes have the features of infected lesions. Sarcomas arising in deep tissues produce symptoms in relation to the organ affected. Diagnosis is made by biopsy.

AETIOLOGY

6. Cancer is not one disease but a group of widely different diseases. While some aetiological factors may be common to a number of different types of cancer, each type should be recognised to be an individual disease with its own specific aetiology.
7. The common feature of all cancers is the loss of control over normal cell division and differentiation. Cell division proceeds by a complex sequence of events. For this to be maintained in a normal way it must be strictly controlled. It has been found that certain regions of the chromosomes are vital to this control. These regions are called oncogenes. While the oncogenes perform normally, cell division and differentiation remain under control.

8. The process whereby oncogenes lose control of cell division and differentiation is known as activation. When this occurs cell division and differentiation become chaotic and neoplasia (carcinogenesis) ensues. The factors which activate oncogenes are numerous and varied, some being endogenous, others environmental. There is evidence that in most types of cancer a number of different factors play a part at different stages of the neoplastic process.
9. Some types of cancer are strongly genetically determined with a family history, for example retinoblastoma. In other types of cancer an external agent is the dominant factor, for example aniline dyes, which will cause carcinoma of the bladder in 100% of cases following sufficient exposure.
10. Some individuals are **genetically determined** to be more likely to develop cancer and there is a strong history of a certain type of cancer in their family of origin. Some cancers are more common in one sex than the other.
11. During life many **constitutional factors** are present which may activate oncogenes. These include humoral factors, immunological factors and the normal ageing process during which spontaneous changes affect the genes (somatic mutations).
12. For the most part, cancer is commoner at the extremes of life. This may be because the immune system is relatively less efficient in the young and the elderly. In addition, with increasing age, the summation of naturally occurring somatic mutations and any exposure to carcinogens may become sufficient to activate oncogenes.
13. **Environmental factors** play a part in the aetiology of some types of cancer. The following groups of factors have been identified:
 - 13.1. **Chemical**, for example dyes and carcinoma of the bladder.
 - 13.2. **Physical** agents, for example asbestos and mesothelioma.
 - 13.3. **Ionising radiation** which when a certain dose is exceeded will cause cancer in some, but not all, tissues.
 - 13.4. **Ultraviolet radiation** which may cause cancer of the skin. Its tissue penetration is limited and so it does not cause cancer in the deeper tissues.
 - 13.5. Some specific **viruses** have been identified which play a part in the causation of particular types of cancer, for example hepatitis B and primary carcinoma of the liver.
 - 13.6. It has been suggested that a wide variety of other environmental factors may cause certain types of cancer. Many of these suggestions have been based on animal studies, in vitro experiments or on epidemiological studies with small samples or inadequate controls. These contentions are still at the stage of speculation.
14. Soft tissue sarcomas are rare tumours. This has made the quest for aetiological factors difficult.

15. Some soft tissues sarcomas are familial and families have been described where a benign condition such as neurofibromatosis with multiple tumours may show progression of some of these lesions to malignancy.
16. Soft tissue sarcomas have been found in relation to vinyl chloride monomer and thorotrast. Other than following the use of thorotrast, no evidence has been produced of any causal link between exposure to excessive ionising radiation and the subsequent development of a soft tissue sarcoma.
17. A number of studies have tried to link agricultural pesticides with soft tissue sarcomas. Some of these studies are technically and statistically unsound. No causal association has been proved.
18. For many years a possible association between virus exposure and soft tissue sarcomas has been postulated. With respect to Kaposi's sarcoma some scientists consider it to be a single disease of viral aetiology and varying in clinical site and course as a reflection of the patient's age at the time of infection and his immune status. This theory remains speculative.
19. Soft tissue sarcomas are not caused by climatic extremes, trauma, physical or mental stress or lowered resistance arising from hardship or other diseases. Its progress is independent of external factors other than medical treatment.

CONCLUSION

20. Soft tissue sarcomas are rare malignant tumours of connective tissue. Constitutional and environmental factors play a part in their aetiology. The course of the conditions is unaffected by environmental factors other than those involved in their treatment.

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