

(Osteogenic sarcoma, Ewing's tumour, chondrosarcoma)**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 metastasises (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. **Sarcoma of bone** is a malignant tumour arising in a bone. The tumours are of three main types, **osteogenic sarcoma**, **Ewing's tumour** and **chondrosarcoma**.

CLINICAL MANIFESTATIONS

4. These may arise from local effects where bone pain, a lump or pathological fracture may occur. In the spine, local tissue spread may cause vertebral collapse, spinal cord or nerve root compression. There may be characteristic radiological findings although these are not always present. Malignant bone tumours are of variable growth rate and malignancy but all have the potential to spread by blood particularly to the lungs. Untreated local expansion may cause an infected malignant ulcer.
5. **Osteogenic sarcoma** occurs in young patients, arises from normal bone and predominantly from the metaphyseal regions of long bones. In older people the tumour arises in bone which is the site of Paget's disease.
6. **Ewing's tumour** occurs in childhood or early adolescence, affects the middle of a long bone and presents like an acute osteomyelitis with systemic features including pyrexia.
7. The incidence of **chondrosarcoma** increases with age. In the older age group it is usually secondary to a previous benign osteochondroma or diaphyseal aclasis. It may arise in any bone, the flat bones and ribs being common sites.

AETIOLOGY

8. 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 as an individual disease with its own specific aetiology.

9. 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.
10. 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.
11. 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 eyes, which will cause carcinoma of the bladder in 100% of cases following sufficient exposure.
12. 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.
13. 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).
14. 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.
15. **Environmental factors** play a part in the aetiology of some types of cancer. The following groups of factors have been identified:
 - 15.1. **Chemical**, for example aniline eyes and carcinoma of the bladder.
 - 15.2. **Physical** agents, for example asbestos and mesothelioma.
 - 15.3. **Ionising radiation** which when a certain dose is exceeded will cause cancer in some, but not all, tissues.
 - 15.4. **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.
 - 15.5. 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.

- 15.6. 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.
16. Sarcoma of bone is a rare tumour, accounting for about 0.5% of all malignancies. There is a relative lack of geographical variation in its incidence. South America, Pakistan and certain parts of Western Europe are said to show a higher incidence. Males are more commonly affected than females.
17. Bone tumours have been extensively studied in animals and a number of causal associations have been described including viruses and chemical agents. No such association has been corroborated in humans.
18. There is some evidence of a familial tendency for bone tumours. Certain skeletal malformations and dysplasias predispose to bone malignancy and, in older adults, Paget's disease predisposes to osteogenic sarcoma.
19. Trauma has been ascribed as a cause of bone cancer but the association has not been confirmed. The consensus of medical opinion is that the apparent association is in fact observer recall bias following the discovery of a tumour after a local injury.
20. Ionizing radiation exposure is conclusively linked to bone cancer. Classic studies of occupational radiation exposure in the manufacture of luminous watch dials and the medicinal use of thorium in radio-scanning are proof of this association.
21. Alkylating drugs in childhood may produce bone tumours in later life.
22. Sarcoma of bone is not caused by climatic extremes, 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

23. Sarcoma of bone is a malignant tumour arising in bone. Constitutional and environmental factors play a part in the aetiology. The course of the condition is unaffected by environmental factors other than those involved in its treatment.

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