VASCULAR TUMOURS OF THE MUSCULO-SKELETAL SYSTEM

ESSAY

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by

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Introduction

INTRODUCTION

A classification of vascular tumours of bone should differentiate between those tumours characterized by a mass of vascular element (Group I) and those that have a specific tissue pattern and are characterized by increased vascularity (Group II). Group I tumours are further subdivided into tumours of "certain vascular origin", and tumours of "uncertain origin". **(Formasier, 1984).**

Vascular tumours of bone arising from blood vessels still represent a debatable and nebulous group of lesions. When confronted with a patient with such a tumour, both the clinician and the pathologist are interested, because of the unique nature of the lesion, but are usually made uncomfortable by the problems of staging, asigning a prognosis, and developing a treatment protocol. One reason is the rarity of such tumours, which limits the experience of even large institutions to few cases. Another reason is that the spectrum of vascular tumours in bone encompasses a wide range, varying from active but benign endothelial growths to tumours of borderline malignancy to low-grade malignant and to very high grade malignant neoplasms. Finally, while in clearly benign and in very high-grade malignant haemangioendotheliomas, the grading is usually relatively straightforward, in those that fall in between the histologic definition may be difficult, and has not yet found unanimous acceptance **(Evarts, 1990).**

Proliferations of newly formed vessels that differentiate and mature into relatively normal structures are *angiomas*, and depending on whether they arise from blood vessels or lymph vessels, they further develop into *haemangiomas* or *lymph-angiomas*. It is often not always possible to tell whether such lesions are malformations, hamartomatous lesions, reactive disorders, or true neoplasms, but they are clearly benign, and do not undergo malignant degeneration. Neoplastic growths of endothelial cells (angioblasts) that cannot differentiate into mature endothelium and vessels are called *haemangioendotheliomas*. The more malignant (high grade) haemangioendotheliomas are also called *haemangiosarcomas* or *angiosarcomas*. Some of these tumours are so poorly differentiated that the production of blood or lymph vessels cannot be assessed. The tumours arising from pericytic cells are appropriately named *haemangio-pericytomas* **(Evarts, 1990)**.

Aneurysmal bone cyst is a primary tumour-like lesion of bone, but the pathogenesis and the tissue of origin remains unresolved and uncertain and, in spite of its histologic resemblance to many other bone lesions, it is currently recognized as a separate and distinct entity. This tumour constitutes approximately 6% of the primary tumours of bone **(Huvos, 1991)**

Disappearing bone disease or massive osteolysis is a rare disease of unknown aetiology and is considered a benign vascular tumour of uncertain origin.

Review of Literature

THE BLOOD SUPPLY OF BONE

1. LONG BONES:

The osseous circulation supplies the living bone tissue, the marrow, perichondrium, epiphyseal cartilages in young bones and, in part, the articular cartilages **(Williams, 1989)**.

Modern researches have emphasized a centrifugal flow of blood through cortical bone in shafts of long bones, in contrast to an earlier concept of substantial centripetal arterial flow into the cortex from periosteal vessels **(Williams, 1989)**.

The vascular supply of a long bone depends on several points of inflow, feeding complex and regionally variable sinusoidal networks within it, which in turn drain to venous channels. These vascular patterns are summarized in Figure (1) **(Williams et al., 1989)**.

One or two main *diaphyseal nutrient arteries* enter the shaft obliquely through *nutrient foramina*, leading into *nutrient canals*. Their sites of entry and angulation are almost constant and characteristically directed away from the dominant growing epiphysis. This is the basis of the *growing-end hypothesis* to explain the positions and orientations of nutrient foramina and canals **(Williams, 1989)**.

Nutrient arteries do not branch in their canals, but divide into ascending and descending branches in the medullary cavity. These approach the epiphysis, dividing repeatedly into smaller

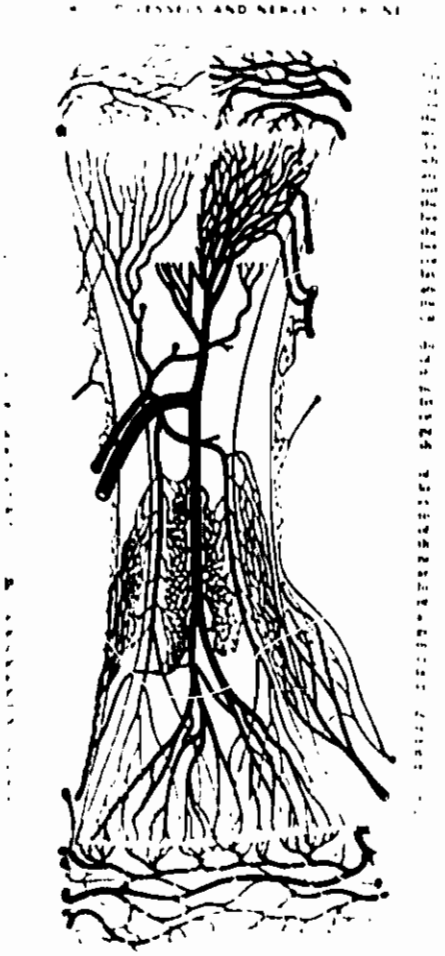

Figure (1)

Blood vessels of bone

rami, which pursue helical courses in the juxta-endosteal medullary zone. Near the epiphysis, they are joined by terminals of numerous *metaphyseal* and *epiphyseal arteries*, the former are direct branches of neighbouring systemic vessels, the latter from periarticular vascular arcades formed on non-articular bone surfaces. Epiphyseal and metaphyseal arteries quantitatively exceed the diaphyseal supply, which they can complement, e.g. when the latter is experimentally destroyed **(Williams et al., 1989)**.

Medullary arteries of the shaft give off:

a) *Centripetal branches*: to a hexagonal mesh of medullary sinusoids, draining into a wide, thin-walled central venous sinus.

b) *Cortical branches:* passing through endosteal canals to feed fenestrated capillaries in Haversian systems. The central sinus drains veins retracing the paths of nutrient arteries, sometimes piercing the shaft elsewhere as independent emissary veins **(Williams et al., 1989)**.

Cortical capillaries conform in their pattern to the Haversian canals. At bone surfaces, cortical capillaries make capillary and venular connections with the periosteal plexuses, which are formed by arteries from neighbouring muscles, constituting vascular arcades with longitudinal links to the fibrous periosteum. From this external plexus, a capillary network permeates the deeper osteogenic periosteum. At muscular attachments, periosteal and muscular plexuses are confluent and the cortical capillaries, then drain into interfascicular venules **(Williams, 1989)**.

The large nutrient arteries of epiphyses form many intra-osseous anastomoses, branches passing towards the articular surfaces within trabecular spaces of the bone. Near the articular cartilages, these form serial anastomotic arcades from which spring end-arterial loops often piercing the thin hypochondral compact bone to enter, and sometimes, traverse the calcified zone of articular cartilage before returning to the epiphyseal venous sinusoids **(Williams et al., 1989)**.

In immature long bones, the blood supply is similar, but the epiphysis is a discrete vascular zone; epiphyseal and metaphyseal arteries enter on both sides of the growth cartilage, anastomosis between them being few or absent. Growth cartilages probably receive a supply from both sources, and also from an anastomotic collar in the adjoining periosteum **(Williams et al., 1989)**.

2. SHORT LONG BONES:

Short bones receive numerous fine vessels from the periosteum at non-articular surfaces, supplying their compact and cancellous bone and medulla.

Study of 848 metacarpal and 811 metatarsal bones showed that, apart from a few with double or no foramina, over 90% had a single nutrient foramen in the middle third of the shaft. All foramina, single or double, slanted away from the epiphysis **(Williams et al., 1989)**

CLASSIFICATION OF BONE TUMOURS

Tumours of bone may arise from the various mesenchymal elements present, in and around bone, and have been assumed to recapitulate the biologic and histomorphologic characteristics of the tissue of origin of the tumour cells. This histogenic concept is the basis of all classifications of bone tumours **(Formasier, 1984)**.

Historically, Virchow made the first attempt at a classification of bone tumours in 1865. He divided bone sarcomata into three categories: the spindle-celled, the round-celled, and the giant-celled types Individual categories of bone tumours were really not categorized until chondrosarcoma was identified as a specific entity by Phemister in 1930 **(Formasier, 1984)**.

The giant cell tumours of bone were first separated from other lesions with giant cells by Coley and Higinbotham in 1938. They were also the first investigators to arbitrarily divide giant cell tumours of bone into a benign and malignant category, in accordance with the clinical behaviour exhibited by their patients.

Parker and Jackson (1939) introduced a classification of malignant lymphoma. They were the first to identify a category of primary lymphoma of bone, distinct from other malignant lymphomas and from Ewing's sarcoma.

Ewing (1939) produced one of the first classifications of bone tumours. He applied the histogenetic concept and produced a categorization of bone tumours which has been the basis of most classifications still in use today **(Formasier, 1984)**.

Geshickter and Copeland (1949) classified bone tumours according to those derived from osseous tissues and tumours composed of non-osseous tissues **(Turek, 1984)**.

Lichtenstein (1952), Johnson (1953) and Jaffe (1958) classified bone tumours according to the basis of Ewing's classification **(Formasier, 1984)**.

The World Health Organization (W.H.O.), in 1972, classified the bone tumours on the basis of histological criteriae, particularly the type of differentiation shown by the tumour cells and the type of intercellular material they produce **(Formasier, 1984)**.

According to Aegerter and Kirkpatrick (1975), bone tumours are best divided into four groups, each group arising from one of the four stem cell types that are derived from the mesoderm. Accordingly, the bone tumours are divided into osteogenic, chondrogenic, collagenic and myelogenic groups **(Aegerter and Kirkpatrick, 1975)**.

Mirra (1980) produced another recent classification based on the fundamental cell(s) and tissue(s) of origin (Table 1). Fundamental cells or tissues are those that are produced by the tumour itself, i.e. those cells or tissues that are directly related to the origin of the tumour. This definition excludes any host bone

cells or tissues that form as reparative or reactive response to the tumour. In most tumours, the blood vessels, osteoclasts, osteoclast-like giant cells and host reparative bone are not of fundamental origin of the lesion. The blood vessels of most tumours merely serve to nourish the lesions and are not of fundamental origin, with the exception of the haemangioma and haemangiosarcoma of bone **(Mirra et al., 1980)**.

Formasier (1984) produced a recent classification, which is a modification of that of Dahlin, incorporating some of the aspects of the classification presented in the histologic classification of bone tumours by the W.H.O (Table 2).

Most recent, one practical classification of primary bone tumours is based on the tissue of origin of the tumours, considering both benign and malignant counterparts **(Moser et al., 1987)**.

According to this classification, bone tumours are classified into:

(1) CHONDROGENIC ORIGIN:

- *Benign:*

1. Enchondroma.
2. Osteochondroma.
3. Chondroblastoma.
4. Chondromyxoid fibroma.

- *Malignant:* Chondrosarcoma.

(2) OSTEOGENIC ORIGIN:

- *Benign*:

1. Osteoma.
2. Osteoid osteoma.
3. Osteoblastoma.

- *Malignant*: Osteosarcoma.

(3) COLLAGENIC ORIGIN:

- *Benign*:

1. Desmoplastic fibroma.
2. Cortical dermoid.
3. Fibroxanthoma.
4. Bone cyst

- *Malignant*:

1. Fibrosarcoma.
2. Malignant fibrous histiocytoma.

(4) MYELOGENIC ORIGIN:

- *Benign*: Intraosseous lipoma.

- *Malignant*:

1. Myeloma.
2. Ewing's sarcoma.
3. Liposarcoma.

(5) VASCULAR ORIGIN:

- *Benign*:

1. Haemangioma
2. Lymphangioma.