ON

BLOOD TUMOURS (ANGEIOMATA AND ANGEIOSARCOMATA) OF BONE.

BY

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BLOOD tumours of bone are of great rarity, but nevertheless of considerable interest and importance, as their nature is very little understood, their diagnosis very difficult, and their treatment consequently not directed upon any definite lines.

The disease has received various names from different authors, having been called osteo-aneurism, capillary aneurism, hæmatoma of bone, sarcoma of bone, &c., these names expressing the different views which have been held with regard to its nature.

Its almost exclusive seat is the cancellous ends of the long bones, especially the head of the tibia and the condyles of the femur. On examination the tumour is found to be composed of a cyst-wall and contents. The cyst-wall is usually formed by a thin expanded shell of bone covered by thickened periosteum, to which the surrounding tissues are more than usually adherent. On opening the tumour it is found to contain little else but blood, partly fluid, partly

coagulated. Sometimes a thin layer of tissue is found lining the interior of the cyst-wall, and sometimes the bony shell is quite destitute of any lining. In some specimens the bone is so destroyed that there is complete loss of continuity between the portion of bone above and below the disease (see Fig. 1). In the last edition of one of the standard text-books of pathology¹ I read that these tumours "are now known to be in the majority of cases soft round-celled or spindle-celled sarcomata. . . . They are exceedingly malignant, and hence the recognition of their sarcomatous nature is all-important." That this statement is not true of all cases is clearly proved by the following case which I had the opportunity of observing very carefully.

Lilian C—, æt. 4, was admitted into St. Bartholomew's Hospital on May 26th, 1886, under the care of Mr. Langton, to whom I am indebted for permission to publish the notes. The only history the mother could give was that ten months previously the child had fallen and hurt her leg. Since then she had noticed that her gait had altered somewhat, and that her left leg was gradually swelling just below the knee.

On admission her general health seemed excellent, and there was no evidence of any constitutional disease: the left leg was swollen below the knee, the circumference of the limb opposite the tubercle of the tibia being an inch and a half greater than that of the other limb. The swelling seemed to involve the upper three or four inches of the tibia and to expand it, for on pressure egg-shell crackling could be easily detected. There was complete absence of pain, tenderness, redness, and œdema; the knee-joint was natural, and there was no glandular enlargement to be discerned anywhere. The thigh was slightly wasted, probably from disuse. It will be gathered from the above account that the cause of the swelling was something inside the head of the tibia expanding it; that was sufficiently evident, but the nature of the "something

¹ Green's 'Pathology,' 1889.

inside" could only be guessed at. Thinking that the case was probably one of myeloid sarcoma, it was resolved to make an exploratory incision, and deal with the case according to the conditions found. This was done on June 14th, 1886, and it was discovered that the upper end of the tibia contained a cavity capable of holding about two ounces of fluid.

The wall of the cavity was composed of bone destitute of any lining, and in front, where the opening had been made, was not more than one tenth of an inch thick. Very little bone seemed to intervene between the cavity and the knee-joint. The contents consisted of a dark red fluid, looking like altered blood and serum, and a very little solid matter, which proved on microscopic examination to be blood-clot. The cavity was stuffed with oiled lint. In a few days it became lined by granulation tissue, and began to contract.

On August 18th the child developed scarlet fever, which necessitated her removal from the hospital; the cavity, however, continued to contract slowly, and by March, 1887, was completely obliterated, a healthy scar remaining in the site of the operation wound.

The patient is now perfectly well. There is a healthy scar in the site of the operation wound, and the bone seems quite consolidated. All the measurements of the two limbs are so nearly equal that no difference can be detected.

This case presented features so peculiar, and so unlike everything I had hitherto believed about such tumours, that I was induced to peruse the literature of the subject to see if I could discover anything like it. Although I could find no record of a similar case, yet I found accounts of others which I think throw very great light upon the nature of these tumours. I propose, therefore, to give short notes of those cases I have been able to find which have most bearing upon the subject.

The most malignant type of blood tumour of bone is nothing more or less than an endosteal sarcoma, in which

blood-vessels have burst and caused extravasation of blood. In this variety there still remains enough solid new growth to be at once recognised, even by the unaided eye, and on microscopic examination the nature of the tumour is sufficiently evident.

But sometimes the amount of sarcoma tissue is so remarkably small that it may easily escape detection. As an excellent example of this, I might quote the following case recorded by Max Oberst.¹

A man twenty-one years old had noticed a swelling upon the inner side of his knee for three months. When first seen by Oberst the tumour was as large as two fists, and was fluctuating in some places. An incision was made into it and blood escaped, partly fluid and partly coagulated. The finger introduced through the aperture impinged upon the internal condyle of the femur, greatly destroyed. The limb was immediately amputated. On examining the limb the sac of the tumour was composed partly of the cancellous tissue of the lower end of the femur, and partly of a thin shell of bone greatly expanded and covered by thickened periosteum. The cyst-wall was lined inside by a thin layer of tissue, most marked in the interior of the femur. On microscopic examination this was found to be sarcoma tissue rich in large round-cells. There were a few giant-cells containing from five to fifteen nuclei. Only a very few layers of cells next to the periphery of the sac were intact; all the others were more or less disturbed and separated by effusion of blood.

The patient died subsequently with metastatic deposits of soft and vascular myeloid sarcoma, containing true ossifications in the periphery, and having a tendency to apoplexy.

Here, then, is a case presenting all the clinical characters of a sarcoma, but being peculiar in that the tendency to effusion of blood was far greater than the power of the tissue itself to grow, and hence the naked-eye appearances of the tumour on dissection.

¹ 'Deutsche Zeitschrift für Chirurgie,' Band xiv, 1881.

The following is a case in which the progress of the disease was arrested for seven years by tying the main artery of the limb. I have made the following abstract from Breschet,¹ who quotes the case from the practice of M. Dupuytren in the Hôtel Dieu.

Clement Nicholas R—, æt. 32, suffered from a pulsatile tumour expanding the upper end of the right tibia. He had noticed it for a year before his admission to the hospital on February 9th, 1819. Dupuytren regarded it as a case of osteo-aneurism, and tied the femoral artery on March 16th. The next day the tumour diminished in size, and on the sixth day the pulsations ceased. The patient left the hospital on April 30th, the "aneurism" having disappeared, leaving only a little tumefaction in the site formerly occupied by it. A long time afterwards the tumour grew again, and assumed a considerable size. On August 1st, 1826 (seven years after the operation), he again presented himself at the hospital, the tumour having attained such dimensions that the leg measured thirty-two inches in circumference. Dupuytren amputated above the knee, and the patient made a good recovery. The specimen was examined by Breschet. The limb was enormous owing to the extraordinary development of the upper end of the tibia, the condyles of which were expanded and divided by compartments into numerous cells like a pomegranate; the walls of the cavity were lined with a vascular network distended by injection, which had been forced into the arteries of the limb. Some of the cells contained a yellowish-black substance, others contained strata of coagulated blood. The cartilages, almost intact, were loosened from the osseous surfaces, and moveable in the middle of the disease.

The following case related by Roux² is even more remarkable.

A man, æt. 25, suffered from a pulsatile tumour ex-

¹ 'Repertoire Générale d'Anatomie et Physiologie Pathologique, et de Clinique Chirurgicale,' tome ii, Paris, 1826.

² 'Quarante années de pratique chirurgicale,' tome ii, p. 456.

panding the upper end of the tibia. After ligature of the femoral artery the tumour disappeared, and the bone returned to its normal condition. The patient was seen twenty years afterwards, and was then in good health. In this case it would seem that the inherent vitality of the new growth was so slight, that after a certain time it became arrested either by pressure of the extravasation of blood, or by arterial starvation following the ligation of the femoral artery, or both.

The following case related by Dr. Lagout d'Aigueperse¹ confirms the preceding case, which if it stood alone might be open to doubt.

The "aneurism" was situated in the upper end of the tibia. It diminished greatly in size, and its pulsation ceased after ligature of the femoral artery, but the bone did not return to its natural condition. It was in the same state eight years afterwards.

I will now quote a case in which the tendency to progress was so slight that it was arrested without any operative interference.

Dr. McDonnell,² in reading a paper on pulsating tumours of bone before the Royal Academy of Medicine in Ireland, detailed the case of a lady who had been sent to him by Mr. Erichsen nearly five years previously. She then suffered from a pulsating tumour over the upper part of the fibula, which he and Mr. Erichsen agreed in regarding as probably a hæmatoid sarcoma of bone. It continued for some time to increase in size. Operation was deferred on account of the lady's pregnancy, but she was directed to wear an elastic stocking. She suffered after delivery from phlegmasia of the other limb, which caused her to remain in bed for nearly six months—still, however, wearing the elastic stocking. When she came again under Dr. McDonnell's care some time after her parturition the tumour was found to have disappeared.

In the absence of any pathological examination of the

¹ 'Bulletin de la Société de Chirurg.,' t. ix, p. 258, 1858-9.

² 'Lancet,' Dec., 1888, p. 1130.

tumour it is impossible to be absolutely sure of the nature of the case ; yet, taking into consideration the facts of the case, and bearing in mind that it was under the care of excellent observers, one can, I think, only infer that it was an innocent blood-cyst of bone, in which the newly formed tissue had such little power of growth that it was arrested by the pressure of an elastic stocking.

Here, then, we have a series of cases of blood tumour of bone presenting very varying clinical characters. Arranging them in order of malignancy, they may be briefly recapitulated thus :

1. Max Oberst's case. A highly malignant endosteal sarcoma.

2. Dupuytren's case. Arrested for seven years by tying the main artery of the limb.

3. Roux's case. Permanently cured by tying the main artery.

4. Lagout's case. Similar to the preceding.

5. The case recorded by myself, in which the growth was arrested by simple incision.

6. McDonnell's case. Cured by the pressure of an elastic stocking.

We must now turn to the microscopical characters of these tumours in order further to elucidate their nature.

In Oberst's case the microscope showed that the tumour was a myeloid sarcoma in which a large number of blood-vessels with very weak walls had burst, and caused extensive extravasation of blood, which had, so to speak, swamped and destroyed most of the solid tissue. It is to be regretted that the other cases in which the specimens were examined occurred before the days in which myeloid tumours had been recognised.

The piece of bone removed from the wall of the cyst in the case of Lilian C— was examined microscopically by my friend Mr. Bowlby, and by him and other authorities pronounced to contain myeloid sarcoma tissue. The section presents different appearances in different places. In some parts trabeculæ of bone are seen being eroded by small round

nucleated cells. In other places large giant-cells containing many nuclei are clearly seen, whilst the bulk of the tissue appears to consist of spindle-shaped cells very much like those of spindle-celled sarcoma. The blood-vessels of the tumour, however, are the most interesting and important. Many of them appear to be only spaces in the tissue with no wall of any sort, and from them red blood-corpuscles may be seen exuding into the surrounding tissues and crowding out the cells of the tissue itself (Fig. 3). Others have a very thin wall, still allowing considerable exudation. Others, chiefly the smallest, have a wall of considerable thickness, and containing many large nuclei crowded together. These blood-vessels are so numerous and large that one is at once led to inquire why they are present in such large numbers. Surely not because the tissue requires a great deal of nourishment, for we see plenty of examples of other tumours growing much more rapidly, and yet being much more poorly supplied with blood-vessels. I am, therefore, forced to conclude that the blood-vessels are an essential part of the tumour. But then it may be asked, how is it that these blood-vessels are mixed up with other structures which are evidently in many cases, at any rate, decidedly sarcomatous, both in microscopical appearance and in clinical deportment? I think the answer to this question is that these blood-vessels grow mainly, if not entirely, from the giant-cells of the bone-marrow, and may present every degree of developmental perfection, from the most imperfectly formed vessels incapable of containing blood up to the perfectly formed vessels of an ordinary angioma. This statement I am unable to prove, but there are many considerations tending to confirm my view.

Thus, firstly, Heitzmann¹ has described the formation of blood-vessels from the giant-cells found in the marrow of healthy cancellous bone. I am not aware that those observations have been confirmed by other observers, but

¹ Heitzmann, "Rück- und Neu-bildung v. Blutgef. im Knochen," 'Wien. med. Jahrb.,' 1873.

Dr. Klein tells me that although he has not actually observed the process himself, yet he thinks it highly probable on *a priori* grounds that it does occur; certainly, blood-vessels are developed from similar cells in other situations: thus in the area vasculosa of the chick large multinucleated cells may be seen becoming vacuolated and forming blood-vessels, and in the subcutaneous tissue of rats large multinucleated connective-tissue corpuscles may be observed to be undergoing the same changes. Although one cannot actually see blood-vessels developing from myeloid cells in the sections of the tumour I have described, yet there are appearances very suggestive of it, especially the capillary vessels with richly nucleated walls already described.

Secondly, these tumours only occur in those parts of bone where myeloid cells are found in health.

Thirdly, in several cases of blood tumour of bone, of which I have read the notes, great stress is laid upon the fact that the sac of the tumour contained a vast number of thin-walled vessels. Thus Breschet, in describing the tumour amputated by Dupuytren, says "the walls of the cavity are lined with a vascular network greatly developed. Over the membrane which lines some of the cells are seen vascular networks distended by the injection forced into the arteries." Scarpa,¹ in describing a blood tumour of the tibia for which he amputated the limb, says the "aneurismal sac was quite covered with arterial vessels of a much greater size than those of the proper arteries of the cellular substance and those of the periosteum. . . . After cleaning thoroughly the inside of the aneurismal sac it was wonderful to see from how great a number of arterial orifices the wax injected into the popliteal artery had been effused into the cavity of the aneurism." Richet² observed the same appearances, and thought that these tumours were pure vascular tumours of bone, and denied that they contained any sarcomatous elements.

¹ 'Sull' Aneurisma,' fol., Patav., 1804.

² Richet, 'Archiv. Gén. de Med.,' 1864 and 1865.

Fourthly, pure angeiomata of bone are occasionally met with. Dr. Mapother¹ has recorded a case in which a blow upon the shin was followed by the development of a tumour, the size of a walnut, in the tibia; there was distensile pulsation, thrill, and bruit. He removed the cuticle by *potassa fusa*, and then applied a cautery. After an interval of ten days intense hæmorrhage occurred: in a few days a "nævoid matter" came away, leaving a granulated surface which rapidly healed. The patient was well sixteen years afterwards. A similar case also occurred to Dr. Bickersteth, of Liverpool. In these cases there was no blood tumour because the blood-vessels were sufficiently strong and fully developed to hold blood without bursting.

From the above facts and arguments I think it follows that not only do these blood tumours present different degrees of malignancy, but that they also differ in structure, some having very embryonic blood-vessels, others more fully developed ones; and it would appear that the more embryonic these vessels are, the more malignant are the clinical features of the tumour in which they occur; and conversely, the more the vessels approach to the type of fully developed structures, the less malignant the tumour. This notion is fully in accord with what is so well known about fibrous tumours, spindle-celled and round-celled sarcomata.

My conclusion, then, with regard to the nature of blood tumours of bone is that they are tumours of blood-vessels, some innocent and some malignant in nature. I would therefore suggest that they be called angeiomata and angeiosarcomata of bone.

Turning now from the pathological to the clinical aspect of these cases, we are still met by great difficulties. Owing to their great rarity they are seldom suspected, and hence usually unrecognised until they are subjected to surgical treatment. Their general symptoms have been sufficiently referred to in the preceding cases to need no further description. The most important thing

¹ 'Lancet,' Dec., 1888.

is to distinguish between innocent and malignant cases, for on this diagnosis must depend to a great extent our prognosis and treatment. On comparing the innocent and the malignant cases, the only difference discoverable between the two is that in the former the progress of the disease is slower than in the latter. As far as I can ascertain, there is no other difference which will help us to decide this very important point. Age seems to have no influence, nor do the characters of the tumour or its constitutional effect on the patient afford the least clue to its degree of malignancy.

How then should such cases be dealt with? The golden rule of cutting into the tumour before removal of the limb or affected bone should certainly be followed. If the tumour be found to contain a large quantity of solid sarcomatous-looking tissue there will be no chance for the patient except from very free removal, and this usually necessitates amputation. If, however, the contents of the tumour consist entirely of blood, or if only a very thin lining of tissue be found, I would suggest that the latter be scraped carefully away, and the cavity stuffed and allowed to granulate up. Of course there is a risk of dangerous hæmorrhage from the interior of the cyst-wall. In two cases, however, which I have seen opened (the one related above, and the other under the care of Mr. Cripps in St. Bartholomew's Hospital) no hæmorrhage occurred. Should it occur, one would not anticipate great difficulty in arresting it.

Should the future progress of the case indicate that we have to deal with a malignant growth we can still resort to amputation, and with scarcely less favorable prospect than if we had resorted to it in the first instance, while we have the satisfaction of having given the patient the chance of recovering with a sound limb.

Although ligature of the main artery of the limb has been practised in a few cases with success, I should not be disposed to try it, as, having regard to the great difficulty in distinguishing between an innocent and a malignant

tumour, we may be wasting valuable time in performing a useless operation which is not devoid of dangers peculiar to itself.

The other methods of treatment which have been used, such as injection with coagulating fluids, compression, &c., need only be mentioned to be condemned.

(For report of the discussion on this paper, see 'Proceedings of the Royal Medical and Chirurgical Society,' Third Series, vol. ii, p. 42.)

DESCRIPTION OF PLATE I.

On Blood Tumours (*Angeiomata* and *Angeiosarcomata*) of Bone
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FIG. 1.—Blood-cyst of tibia (from the Museum of St. Bartholomew's Hospital).

FIG. 2.—Blood-cyst of lower end of femur.

a. Thin layer of sarcomatous tissue.

b. Cavity filled with blood.

(After Max Obërst.)

FIG. 3.—Blood-cyst of head of tibia (the case of L. C—).

Section of cyst-wall, showing blood-corpuscles exuding from thin-walled blood-vessels into surrounding tissues.