The Natural History of Aneurysmal Bone Cyst

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ANEURYSMAL bone cyst may be defined as a cystic lesion of bone containing clefts or spaces filled with blood which is in continuity with the general circulation. It often increases in size very rapidly and may expand the bone to an extreme degree. This expansion is frequently confined to one aspect of the bone.

The clinical and radiological appearances of the lesion closely resemble those of osteoclastoma, and the solid parts of it may appear similar histologically, so that it is sometimes classified as a "variant" of the giant cell tumours. Aneurysmal bone cyst, however, frequently commences in childhood when osteoclastoma is very rare.

The purpose of this paper is to present the microscopic appearance of the epiphysis related to a typical cyst. This shows clear evidence of infiltration by the cyst lining and suggests that the cells are neoplastic. The findings explain the frequent recurrence of the lesion after curettage and the occasional spread to adjacent bones. It seems likely that the cells of the cyst wall are of vascular origin and that the lesion is a form of hæmangioma, probably a hamartoma. Six other cysts are described which illustrate the probable natural history of the lesion, which is seen to resemble that of hæmangiomata in other tissues.

Aneurysmal bone cyst was first described as a clinical and pathological entity by Jaffe and Lichtenstein (1942). Several theories regarding the ætiology of the lesion have been proposed. Ewing (1940), Cruz and Coley (1956) and Lichtenstein (1957) all considered that the condition was caused by increased venous pressure, but Barnes (1956) suggested that it was an abnormal response to injury. Hadders and Otterdoom (1956), after a thorough pathological study, concluded that aneurysmal bone cyst was an angioma of bone. Parkes Weber (1956) also holds this view.

Case I.—A 3-year-old girl attended hospital in December 1956 with a swelling at the upper end of the left fibula: no history of injury or other causative factor. The swelling was slightly tender, but not red or hot.

Radiographically the upper end of the diaphysis of the fibula was distended by an apparently multilocular cyst (Fig. 1). The epiphyseal line and bony epiphysis were poorly defined, but apparently separated from the cyst by a plate of bone.

The whole upper end of the fibula, containing the cyst and epiphysis, was excised. Functional recovery in the leg was complete.

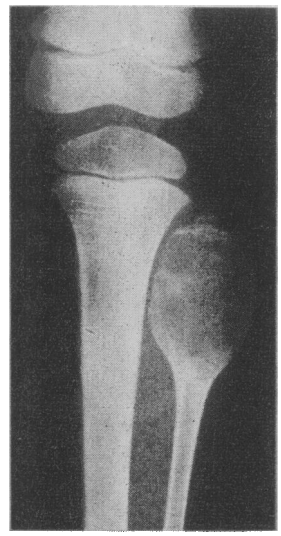

FIG. 1 (*Case I*).—Radiograph showing the expanded upper end of the fibula, the irregular line of dense bone adjacent to the epiphyseal plate and the small, rarefied ossific centre of the epiphysis.

Pathology.—On section the cyst was seen to be unilocular, but the bony wall was roughened by low projecting ridges which had caused the radiological appearance of a multilocular cyst. The cyst cavity was filled with fluid blood except for one corner which contained a wedge of cellular tissue. Microscopically, the cyst wall was seen to contain many blood-filled spaces lined by endothelium and separated by cellular fibrous tissue, composed of numerous dark spindle-shaped cells and occasional groups of multinucleate giant cells, the picture typical of aneurysmal bone cyst.

The epiphysis.—Three full-width sections of the epiphysis and related cyst were examined. The cyst wall was seen to be closely opposed to the cartilage in many parts, especially near the perimeter of the plate, but in others they were separated by a zone of provisionally calcified cartilage up to 1 mm. in width.

At the edge of the epiphyseal plate the blood spaces and cells of the cyst wall infiltrated between the cartilage columns of the plate and the periosteum, probably along the course of epiphyseal blood vessels (Fig. 2). Where blood vessels penetrated the plate, the cyst extended along them to pass through the zone of cartilage-cell columns (Fig. 3), and at one point, not illustrated, it reached the bony epiphysis. This infiltration of the cyst tissue along blood vessels, and the presence of blood-filled spaces in the infiltrating tissue suggests that the cyst cells are of vascular origin. It also explains the invasion of the epiphysis which is frequently seen

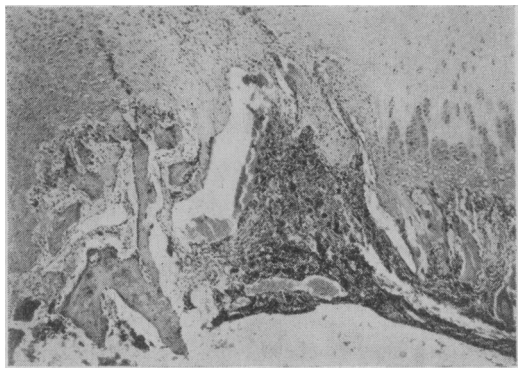

FIG. 2 (*Case I*).—Photomicrograph of part of the perimeter of the epiphyseal plate where the cyst extends up to the cartilage-cell columns and infiltrates between the edge of the plate and the periosteum (× 28).

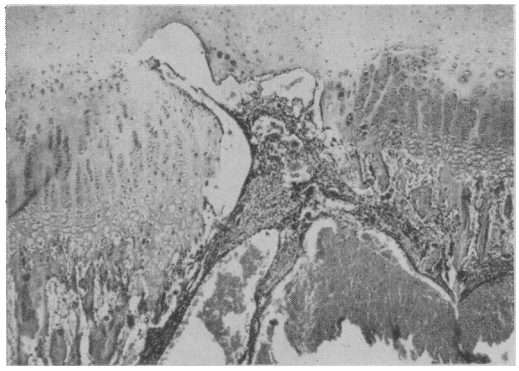

FIG. 3 (*Case I*).—Photomicrograph of the epiphyseal plate where it is pierced by a blood-vessel. The cells of the cyst lining can be seen spreading along the channel toward the bony epiphysis (× 28).

radiologically in aneurysmal bone cysts, and the occasional spread to adjacent bones in the spine and tarsus described by Lichtenstein (1957).

The cartilage surrounding these penetrations was covered with a thin plate of woven bone. Where the invasion reached the bony epiphysis, it was apparent that this plate of bone was, in effect, a rigid tie across the plate, which would interfere with growth. The passage of blood vessels across the normal epiphyseal plate was studied by Ham (1953), who concluded that such penetration does occur and that the normal vessel walls must possess some special property to prevent ossification of the adjacent cartilage. Such a property is apparently not present in the vessels of the aneurysmal bone cyst.

In the central part of the epiphysis, where the cyst was separated from the plate, a curious change was noted (Fig. 4); the swollen cartilage cells were not arranged in columns but were

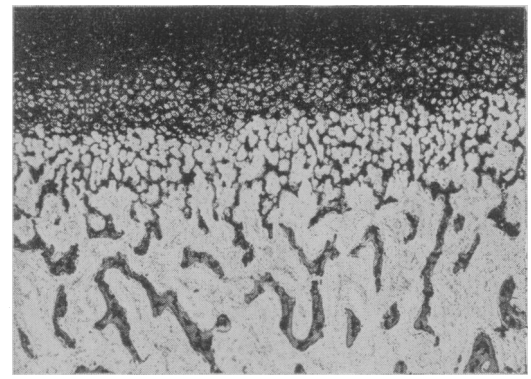

FIG. 4 (*Case I*).—Photomicrograph of the central part of the plate where the cartilage-cell columns and provisionally calcified trabeculæ are irregular.

scattered irregularly, and the provisionally calcified cartilage was in irregular branching fragments, quite unlike the straight bars in the remainder of the metaphysis. The cause of this was obscure until the work of Gelbke (1951) was studied. He showed almost identical changes resulting from restriction of epiphyseal growth by wire bands. In this case a similar restriction could have been caused by the bone layers crossing the epiphyseal plate.

Discussion.—The study of this specimen suggests that the lesion was a neoplastic condition affecting the blood vessels. The radiographic and microscopic appearances are, however, very different from those of the hæmangiomata which are frequently found in the vertebræ and occasionally in other bones (Bucy and Capp, 1930). In these cases the radiographs of the flat bone and clavicle tumours showed a well-marked radial trabeculation, while the long-bone tumours produced a polycystic appearance. Microscopically the blood spaces retained their circular shape and the interstitial tissue resembled loose connective tissue with very few giant cells. The behaviour of hæmangiomata is, however, very variable. Winston Evans (1956) describes active invasion in some cases, mostly children, while others undergo thrombosis and fibrosis. The study of the other cases in this series shows that all these processes may occur in aneurysmal bone cyst and account for its variable natural history.

The Natural History

The onset of symptoms in aneurysmal bone cyst is frequently during childhood. Case I is the youngest example published so far, but all the cases described by Taylor (1956) and Barnes (1956) were under 21 years of age, as were 6 of 7

cases by Lichtenstein (1953) and 11 of 20 by Cruz and Coley (1956). In many of the other cases it is probable that the cysts had been present for many years before producing symptoms.

In most cases the lesion arises on the metaphyseal side of an epiphyseal plate, but the lesion may extend across the plate into the epiphysis at an early stage, and cysts presenting toward the end of growth frequently involve the epiphysis when first seen. This process is well illustrated by the next two case reports.

Case II.—A 12-year-old girl first noticed a swelling below her right knee one month before attending hospital in March 1957; radiographic examination revealed a smooth-walled cyst in the upper metaphysis of the tibia (Fig. 5). At exploration the cyst contained

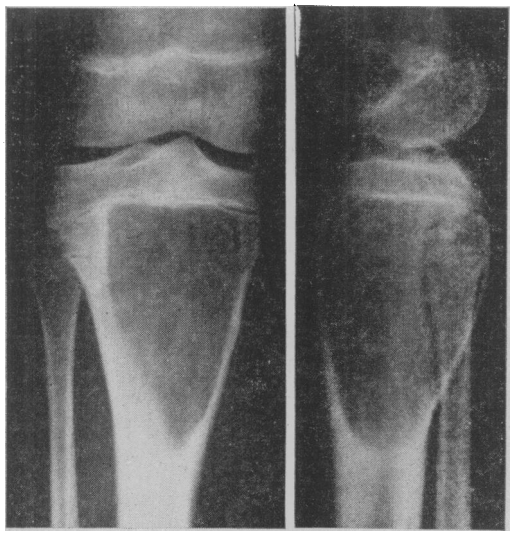

FIG. 5 (*Case II*).—March 1957. A unilocular cyst in the upper end of the tibia, slightly eccentric and producing marked thinning of the medial wall.

glairy fluid, and the soft-tissue lining, which was thick posteriorly, was removed by curettage. The epiphyseal cartilage was exposed in the wall of the cyst. Cancellous bone chips were packed into the cavity.

Three months after the operation radiographs showed the grafts apparently consolidating and the wall becoming thicker. After six months, however, the grafts had been absorbed and the cyst was obviously extending both laterally and upward through the epiphyseal plate (Fig. 6).

Treatment.—In view of the unsatisfactory response to curettage and grafting, further treatment was by radiotherapy alone. A dose of 1,500 r was delivered to the tumour, and the follow-up for the first year has shown a gradual recalcification and no further extension of the cyst.

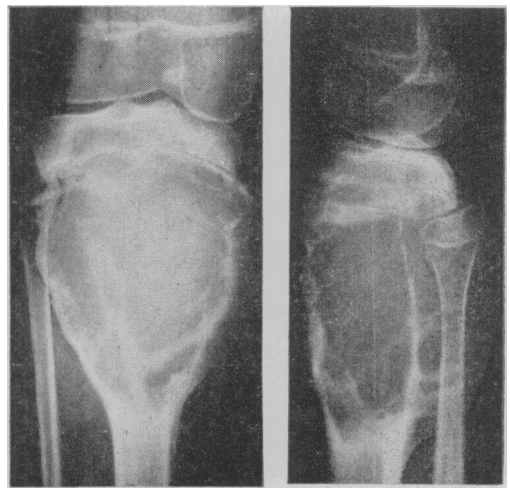

FIG. 6 (*Case II*).—September 1957. The cyst has enlarged and is extending into the epiphysis. The absence of bone from the antero-medial wall is shown.

Histology.—The curettings removed at the time of operation showed the typical endothelium-lined vascular spaces of aneurysmal bone cyst with a very cellular interstitial tissue containing many spindle-shaped cells and occasional giant cells, some of which contained pigment granules. The bone of the cyst wall was composed of newly formed woven bone.

Case III.—A 19-year-old girl attended hospital in 1951 with aching in the left ankle and swelling on the anterior surface of the lower end of the tibia for eight months; no injury or other causative factor. There was no limitation of ankle movement. The radiographic appearances are shown in Fig. 7.

Treatment.—This lesion was not recognized as an aneurysmal bone cyst. At biopsy the very thin cortex over the tumour with areas of hæmorrhagic discoloration under the periosteum suggested possible malignancy, so the whole of the lower end of the tibia was excised and the defect bridged by cortical

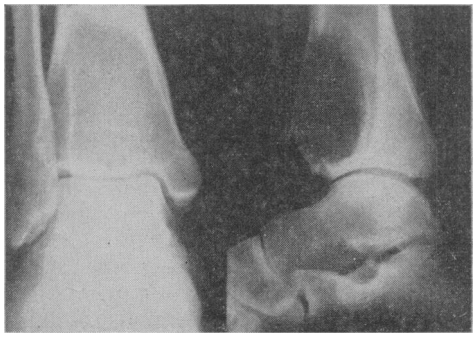

FIG. 7 (*Case III*).—Radiograph showing the cyst bulging forward from the lower end of the tibia and raising the periosteum from the adjacent normal shaft.

and cancellous bone grafts fusing the lower ends of the tibia and fibula to the talus.

The patient now has sound fusion of the ankle, a painless leg and no shortening.

Pathology.—The cyst was intersected in many places by thin grey to light brown membranes. The cyst wall consisted of delicate strands of connective tissue, covered by flattened endothelial cells. Giant cells in the wall of the cyst adjacent to lamellar bone were grouped around and often contained hæmosiderin. There was no histological evidence of resorption of bone. The appearance is very similar to that in Case I.

Another possible mode of onset has been described in the literature. Some cysts, pathologically indistinguishable from aneurysmal bone cyst, have been described arising from the midshaft region of long bones (Van Arsdale, 1893; Thompson, 1954; Jaffe, 1950; Lichtenstein, 1950; Geschickter and Copeland, 1949). These frequently occur in adults and are often related to trauma. Most of the early cases were diagnosed as ossifying subperiosteal hæmatomata. The next case is an example of this type of cyst.

Case IV.—A 43-year-old housewife first noticed a painful swelling in her right upper arm in September 1958, but did not attend hospital until January 1959 when a minor injury caused a great increase in the pain. No previous trauma.

There was a hard swelling deep to the muscles on the outer side of the upper arm, tender to deep pressure, but with no limitation of shoulder or elbow movement (*see* radiograph, Fig. 8).

The serum calcium and alkaline phosphatase were normal.

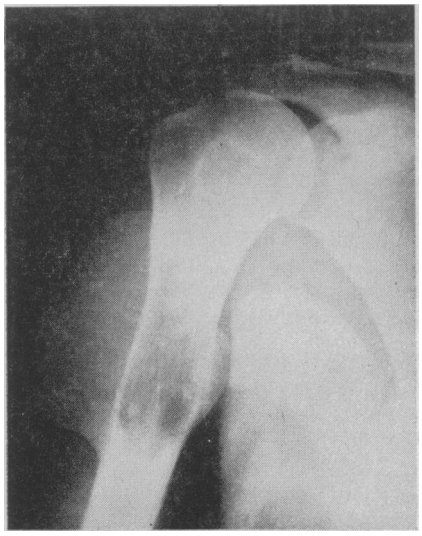

FIG. 8 (*Case IV*).—Radiograph of the right humerus showing a thin-walled cyst arising from the shaft of the bone.

A needle biopsy revealed sheets of fibroblasts with occasional giant cells and, in some parts, an endothelial covering.

The tumour was excised. It separated easily from an intact fibrous capsule over most of its extent, but was attached to the bone. It contained fluid blood and some grey tissue. The cyst lining was curetted from the bone, leaving a saucer-like defect on its antero-lateral aspect which was filled with bone chips. Recovery was uneventful.

Histological section of the cyst wall showed a typical aneurysmal bone cyst.

The subsequent progress of untreated cysts has not been fully elucidated in previous publications. That extension of the cyst may continue after skeletal growth stops is clear from cases in Lichtenstein's series, and several reported examples have reached a large size. It is probable, however, that some cysts remain undiagnosed throughout the life of the patient, having ceased to grow before they interfered with any vital structure. If these cysts are in fact hæmangiomata, thrombosis and fibrosis may be expected to occur in some. In a small cyst this may lead to complete resolution and recalcification; in a larger one some residual deformity would be inevitable. The next case probably illustrates an early stage in the resolution of a small cyst.

Case V.—A 40-year-old man attended for a routine chest radiograph in September 1956. This showed a

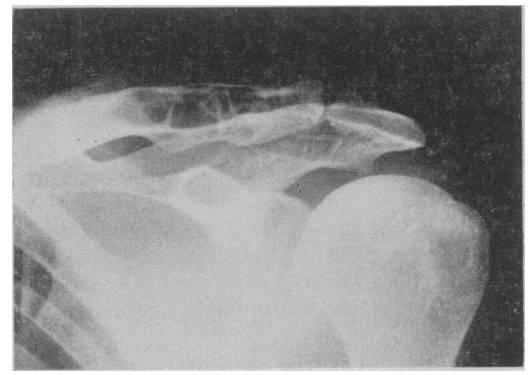

FIG. 9 (*Case V*).—Radiograph showing cystic distension of the outer end of the clavicle.

cystic condition in the outer end of the left clavicle (Fig. 9). He had no symptoms and there was no history of injury.

The cyst contained blood-stained serous fluid. The wall presented several projecting bony ridges and was curetted. Recovery was uneventful.

Histological examination showed the cyst lining to consist of delicate, almost myxomatous, connective tissue containing very few giant cells and covered with flattened endothelium. The lining was split by

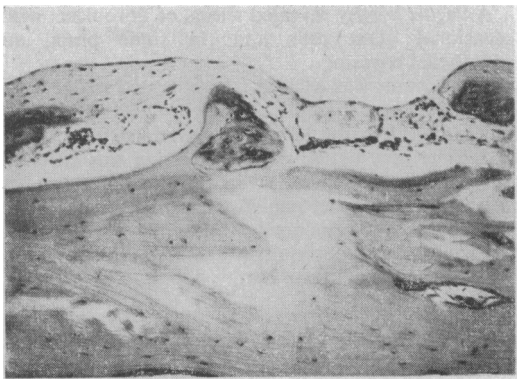

FIG. 10 (*Case V*).—Photomicrograph of the bony wall of the cyst showing the mature fibrous tissue containing b'ood-filled spaces and covered with endothelium (× 75).

several endothelium-lined clefts containing red blood corpuscles (Fig. 10).

Comment.—This cyst presented none of the active cellular tissue seen in the lining of the typical cyst, but the endothelium-lined cavity with blood-filled clefts in its walls retained the basic pattern of aneurysmal bone cyst.

The deformity left by thrombosis or fibrosis of large cysts has not previously been described and there are no established criteria for the diagnosis of such a lesion when found at necropsy.

Two cases were described, in each unconnected with the cause of death. A large cyst involving the petrous temporal and middle fossa was found in a woman of 72, and another in the left ilium of a woman of 75. These two cases presented a recognizable lesion characterized by a fibrous wall with plaques of calcification, surrounding an endothelial-lined cavity containing thrombus, the whole arising from a bone, usually near the site of a cartilaginous growth plate. Aneurysmal bone cyst appears, at present, to be the most likely cause of these lesions.

Treatment.—On the basis of this natural history, a rational basis for treatment can be formulated. In the growing child the cyst wall will probably be infiltrating the epiphyseal plate at the time the diagnosis is made, so that curettage, or local excision which does not remove the epiphyseal cartilage, is likely to be followed by recurrence. The ideal treatment is therefore complete excision of the cyst and the related epiphysis when this can be done without loss of function, as in Case I. When this is not feasible, radiotherapy is indicated, and is usually effective in small doses from which there is very little risk of post-irradiation sarcoma.

In adults, cysts in accessible sites can be cured by curettage or local excision, and this is the treatment of choice. Radiotherapy is only indicated when an inaccessible tumour is pressing on vital structures, such as the spinal cord, or is increasing in size.

Ætiology.—This is still uncertain. While this study suggests a neoplastic process affecting vascular tissue, it is apparent that aneurysmal bone cyst bears very little resemblance to the established cases of cavernous hæmangioma of bone. Some parts of the cyst lining, however, do resemble closely the cellular lining of the large cavities in certain hæmangiomata of the soft tissues. These tumours are in fact so variable in their structure that different types of presentation would be expected.

Summary.—The epiphysis related to an aneurysmal bone cyst showed active infiltration of the epiphyseal cartilage by the cyst wall, reaching through to the bony epiphysis. The appearances suggest that the lesion is a form of hæmangioma.

The natural history may be summarized thus:

Onset, usually in childhood, as a proliferation of vascular tissue in the bone adjacent to a cartilaginous growth plate or, in some cases, in a subperiosteal hæmatoma.

Growth by: (1) Active invasion, mainly along blood vessels causing early transgression of the epiphyseal plate. (2) Passive distension of the vascular spaces in the tumour by the pressure of the contained blood.

Inactivation by fibrosis or thrombosis leading to: (1) Absorption and recalcification of small cysts. (2) The formation of a chronic, thrombus-filled cavity surrounded by endothelium, fibrous tissue and bone in the site of a large cyst.

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The Management of Fractures of the Tibial Spine in Children

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THE object of this paper is to define the place of conservative and operative treatment for fractures of the tibial spine in children. It is based on experience of 10 patients, 5 treated conservatively, 4 operatively and 1 untreated until he was first seen four years after injury.

Clinical features.—The children's ages ranged from 4 to 14 years. They had suffered either a rotational strain at the knee, or, more commonly, a blow applied to the front of the thigh with the knee flexed, as by the handle of a wheeled toy that had tipped back. The clinical findings were characteristic—a flexed, painful knee, signs of hæmarthrosis and possibly a bruise or abrasion on the front of the thigh. A lateral radiograph of the knee showed a flake of bone raised from the tibial plateau, not easily visible on antero-posterior films (Fig. 1).

Pathology.—The injury was always much more extensive than the radiographic evidence suggested. With the flake of bone, a large area of surrounding articular cartilage of the upper surface of the tibia had been separated and displaced upwards and backwards by the anterior cruciate ligament (Fig. 2). The fragment, unlike that in adult fractures, was always attached posteriorly by a hinge of articular cartilage. In 3 out of 5 knees that were explored, the area of cartilage detached from the medial articular surface of the tibia was greater than from the lateral surface, and extended beneath the medial meniscus almost to the anterior or medial margins of the bone. The edges of the fragment were entangled with the medial meniscus, so that, when attempts were made to reduce it, the fragment "locked" over the upper surface of the meniscus. It was in these 3 patients that manipulation under anæsthesia before operation had revealed a block to the last 20 degrees of passive extension of the knee.

In one patient, in whom no such limitation was found, the fragment was smaller, did not involve much of the articular surface, and was

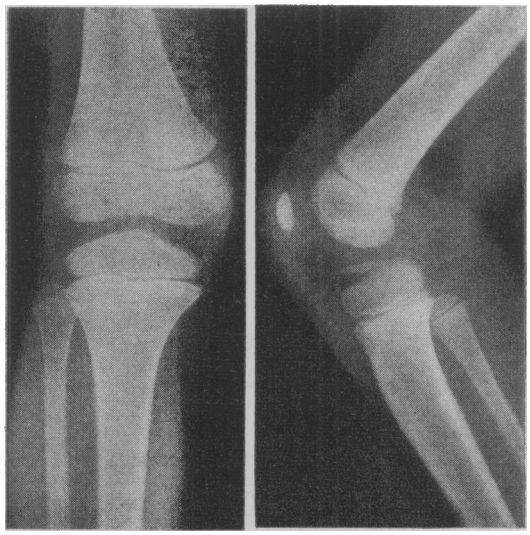

FIG. 1.—Fracture of the tibial spine in a child aged 8. The fragment did not reduce with simple extension of the knee, and there was some residual disability at follow-up examination.

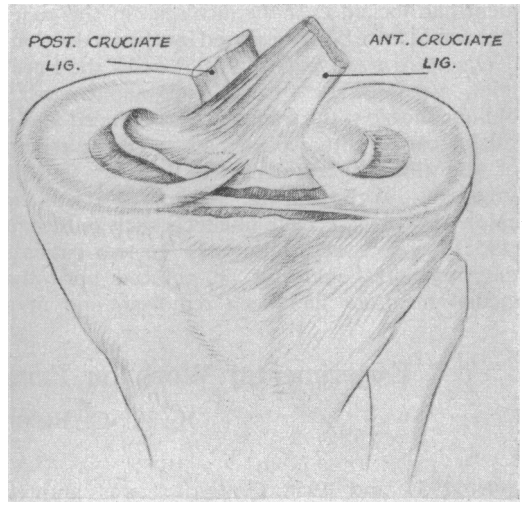

FIG. 2.—Diagram to illustrate the findings in fracture of the tibial spine. A large osteocartilaginous fragment is hinged posteriorly and is pulled upwards and backwards by the anterior cruciate ligament. Attempts to reduce the displacement may cause the fragment to lock on to the upper surface of one or other meniscus.