FROM THE RADIO-PATHOLOGIC DEPARTMENT OF RADIUMHEMMET IN STOCKHOLM, SWEDEN; CHIEF: PROFESSOR OLLE REUTERWALL, M.D.

ADAMANTINOMA OF THE LONG BONES

Report of a Case of Tibial Adamantinoma

By

JOHN HERTZ

> There are more things in heaven and earth, Horatio,
> than are dreamt of in your philosophy–
>
> (*Shakespeare:* Hamlet, Act I, Scene V)

Adamantinomas very rarely occur in the long bones; the author has been able to trace only 24 instances of this location in the entire literature. In 22 of these cases the growth appeared in the tibia, in one instance in the femur (*Bell*) and in one case in the ulna (*Anderson & Saunders*).

Of 32 cases of adamantinoma referred to the radio-pathologic department of Radiumhemmet in Stockholm, Sweden, during the thirty years period 1921–1950, one was located in the tibia.

CASE RECORD

History. A male, 49 when first seen, had noticed a lump on the front of the right tibia four years before entry. The lump, which had increased slowly, was much exposed to traumas, especially when the patient was working. Apart from a slight weakness of the leg and some tenderness of the lump there had been no symptoms.

Examination showed a slightly tender node, the size of a hen's egg, on the medial surface of the right tibia; the node was felt as a firm ring with a soft central area and it was not fixed to the skin. There was no atrophy of the leg. The routine examination was essentially negative. The Wassermann test was negative. The x-ray report was positive for a polycystic growth in the right tibia (Fig. 1); x-ray examination of other parts of the skeleton was negative.

Treatment. Excochleation with chiselling was performed and a solid, firm

The surgical department of the hospital in Falun, Sweden, has kindly placed the clinical data at my disposal.

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brownish tumour with a few cystic cavities was revealed. The compact bone of the medial surface of the tibia was perforated by the tumour; the bony shell presented small depressions but the tumour appeared to be well demarcated. The limb was put in a plaster-of-Paris casing.

The postoperative course was uneventful and *irradiation* was instituted.

Subsequent progress. When he had a trauma 18 months later, a recurrence was revealed which required *resection of the tibia,* with grafting. Apart from a later

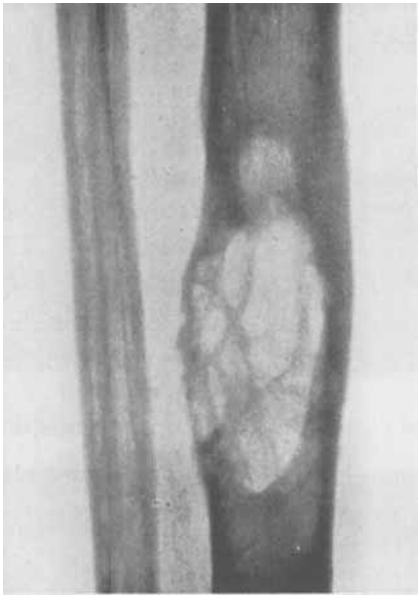

Fig. 1.
Tibial adamantinoma.

fracture of the graft the postoperative course was uneventful, and the patient has been without relapse for a postoperative observation period of 8 years.

Pathology. Grossly, the tumour was grayish-white, like marrow, with yellow-brownish streaks; the pieces were finely papillomatous.

Microscopically, the tumour was composed of a stroma of connective tissue of varying abundance and content of cells supporting irregularly anastomosing epithelial strands which were rich in cells. At the periphery of the epithelial areas the cells were in places cylindric, forming palisades, while in the central parts of the areas they were irregular in outline, polygonal, and in places stellate (Figs. 2 and 3). In other places the cells were large, ovoid, with nuclei rich in chromatin, showing no particular polymorphism or remarkable amount of mitotic figures. Elsewhere the cells were rather small and compact, with deeply staining nuclei and relatively scant protoplasm (Figs. 4 and 5). The epithelial vegetations were sharply outlined against the stroma. In some places vacuolization was observed, with formation of cysts. In other parts slight hemorrhages as well as some dark pigment were seen.

The bony channels in the adjacent parts were filled with epithelial tumour

elements and a dissolution of the bony structure was in evidence with the formation of osteoid masses and fascicles of collagen in between which the tumour cells penetrated. Further, there were unmasked protoplasmic areas and areas of "granular crumbling" (*John Hertz*).

Where the epithelial tumour elements invaded the tissue surrounding the bone, a comparatively dense fibrous tissue was seen in which the epithelial elements

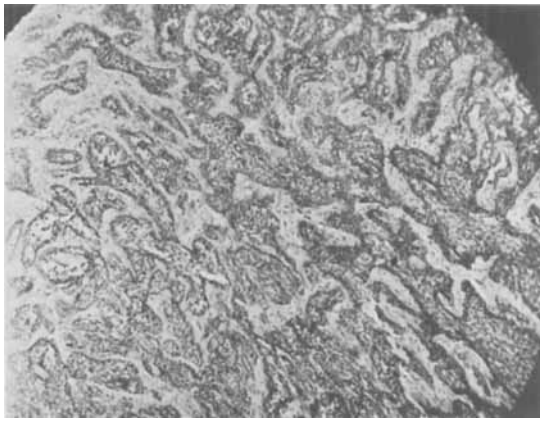

Fig. 2.
Tibial adamantinoma; fully differentiated type.

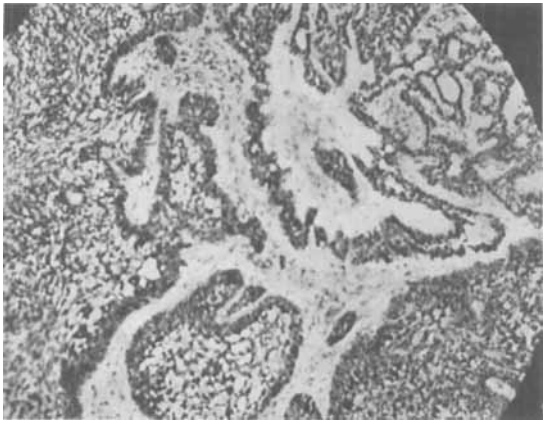

Fig. 3.
Tibial adamantinoma; fully differentiated. Note the enamel organ-like structures.

were in evidence; these elements were, however, few and scattered. In no place have the epithelial proliferations been found to grow in the loose connective tissue (Figs. 6 and 7). An infiltrative and destructive growth into the surrounding structures as in a true carcinoma was not observed anywhere.

Classification according to *John Hertz:* Fully differentiated adamantinoma, in places resembling a basal-cell carcinoma and containing areas with large solid epithelial elements with less differentiated cells.

The clinical card is typical. A slowly growing, only slightly tender, swelling was submitted to conservative surgery, curettage. The patient had a relapse which required a radical surgical procedure, resection. The histologic pattern is likewise typical, revealing a fully differentiated adamantinoma, exactly like that seen in adamantinomas of the jaws.

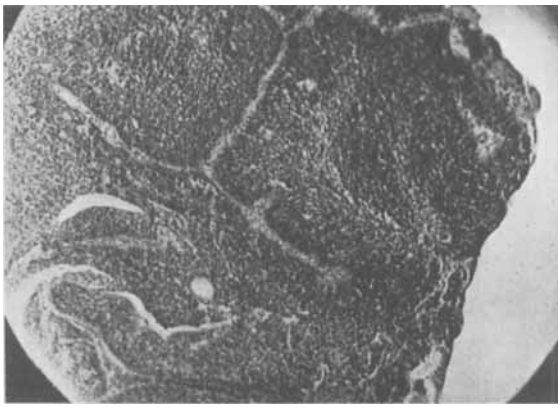

Fig. 4.
Tibial adamantinoma, suggestive of a basal-cell carcinoma.

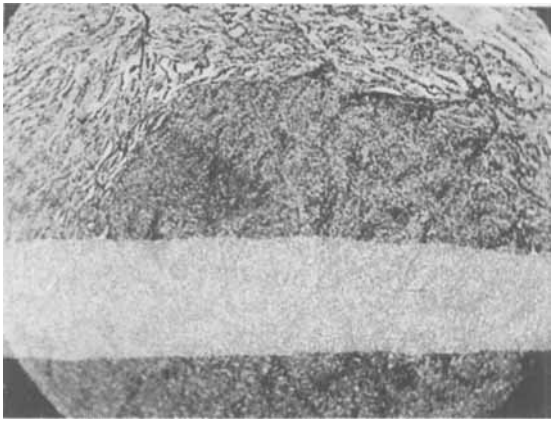

Fig. 5.
Tibial adamantinoma, suggestive of a basal-cell carcinoma.

CLINICAL SYMPTOMS

Of the instances on record 12 patients were males and 12 females. The patients were between 12 and 57 years old. In no less than 16 cases of the 23, in which it has been possible to obtain sufficient information, the site of the tumour had been exposed to trauma.

It is a slow growing swelling rather than pain which in a number of cases has brought the patient under treatment. In half of the cases the lump has, however, been painful; in one instance pain was the first symptom (*Holden & Gray*). The patient reported here complained of some weakness of the afflicted limb.

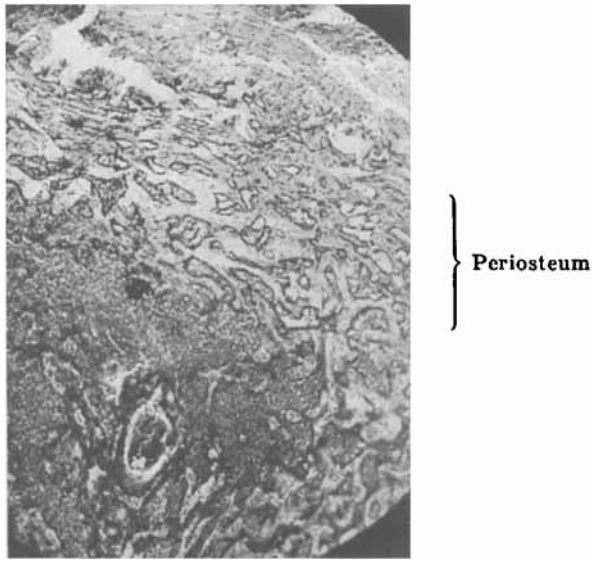

Fig. 6.
Tibial adamantinoma. Small epithelial strands are present in the periosteum.

In one case on record an ulcer of a considerable size was present and the condition occurred in a previous osteomyelitis focus (*Casini*).

For the establishment of a decisive diagnosis a biopsy must be taken in addition to the x-ray examination.

PROGNOSIS

In the prognosis the most striking feature is a marked tendency to recure. This holds good for adamantinomas of the long bones as well as for adamantinomas of the jaws. Most of the patients tell about a condition of long standing which has been submitted to numerous surgical procedures over several years. In all the cases on record which were first submitted to conservative surgery, excision or curettage (14), a recurrence occurred. In one case a "wide excision" effected a cure for the 8 month period during which the patient was

followed-up (*Rehbock & Barber*). In one instance where irradiation was the only therapeutic procedure (*Dunne*) a recurrence occurred. The case of adamantinoma of the femur recorded by *Bell* is peculiar in that the patient, a 16 year old native from Nigeria, died 5 weeks after an incision. The tumour was considered a deep intramuscular abscess and treated accordingly. The patient died with symptoms of bronchopneumonia but autopsy was not performed, and the possibility

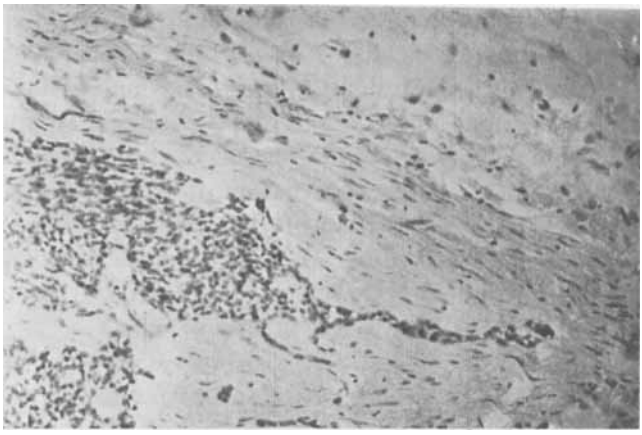

Fig. 7.
Tibial adamantinoma. Representative picture of the peripheral part outside the bone. Note the appearance of epithelial vegetations in comparatively dense fibrous tissue and abutting on this, a loose connective tissue, without tumour infiltration.

cannot be excluded that the condition may have been of a metastatic nature; anyhow, the diagnosis of adamantinoma had been verified microscopically.

In all the cases treated with radical surgery, the period of observation has been too short to allow any evaluation of the final outcome. In the case of adamantinoma of the ulna a relapse occurred 5 years after resection and required amputation (*Anderson & Saunders*).

Clinically, adamantinomas of the long bones thus behave exactly as adamantinomas generally. My series shows that in adamantinomas in general, conservative surgery has failed to cure the patient in 100 % of the cases, whereas radical surgical procedures give a five year cure rate of appr. 70 % of the cases.

Consequently, conservative surgical procedures are insufficient in cases of adamantinomas in the long bones as well as in the jaws and only invite disaster. Radical surgery must be the treatment of choice.

PATHOGENESIS

Several theories have been advanced concerning the pathogenesis of the adamantinomas of the long bones. *Fischer* (1913), to whom the first published case of tibial adamantinoma is credited, arrived at the conclusion that the tumour begins as a downgrowth of the ectoderm, extending to the bone. According to *Fischer,* the differentiating capacity of the oral ectoderm is shared by the ectoderm generally in the manner that a rest laid down at the time of formation of the enamel organ retains a specialized growth capacity and in neoplasm assumes the structure of an adamantinoma. *Rankin* also holds responsible for the tumour development a cell rest from foetal life which becomes activated in later life. *Baker & Hawksley* are of the opinion that in the case which came under their observation the epithelium was subperiostal from embryonic life; it would be difficult to account for its deposition subsequently by means other than a cutaneous wound with implantation of a fragment of skin, or metastasis from a primary tumour elsewhere. There was no evidence of this.

Another theory explains the origin of these tumours entirely on the basis of a trauma with implantation of cutaneous appendages. *Ryrie* speaks of a "thwarted repair". It would require a very considerable trauma to have the epithelium implanted.

Dockerty & Meyerding claim that tibial adamantinomas are merely composed of modified squamous cells which vary considerably in their differentiation into ameloblasts. *Richter* has called this type of tumour a particularly differentiated bone endothelioma.

Hebbel has arrived at a discouraging conclusion; he eliminates trauma as an aetiologic factor in tibial adamantinomas, "leaving its origin from an ectodermal rest as an insecurely established alternative". The theory is by no means unattractive, but information concerning the occurrence of ectodermal rests in relation to skeletal structures is wholly lacking.

The present case fails to give an answer to the question about the aetiology and the pathogenesis; it cannot be definitely established whether the epithelial proliferations have originated in the periosteum or have arisen in the bony tissue and secondarily invaded the periosteum as small epithelial strands (Figs. 6 and 7); the latter explanation is the most acceptable when taking the relationship to the adjacent structures, as described above, into account. Further, it seems most reasonable to hold embryonic anomalies responsible for the development of adamantinomas of the long bones.

No.	Year	Author	Age	Sex	Symptoms	Trauma	Latent period	Treatment	Subsequent course
1	1913	*Fischer*	37	M	Pain – Swelling	Contusion	5 months	Resection + grafting	8 months' cure
2	1930	*Richter*	12	M	Swelling	Fall		Amputation	
3	1931	*Baker & Hawksley*	46	M	Pain – Swelling	Contusion twice	8 months – 6 weeks	Resection + grafting	2 months' cure
4	1932	*Ryrie*	52	M	Swelling	Contusion	8 years	Curettage	Relapse – Amputation 2 months later
5	1933	*Petrow & Glasunow*	22	M	Swelling for 3 years			Resection + grafting	One year cure
6	1934	*Holden & Gray*	36	F	Pain	Contusion		Excision	Relapse after 2 years – Re-excision + irradiation
7	1935	*Casini*	55	M	Swelling – Ulcer	Run-over 7 years old – Osteomyelitis		Amputation	
8	1937	*Bishop*	22	M	Swelling	Fracture	8 weeks	Curettage + chips graft	Relapse – Amputation 14 months later
9	1938	*Rehbock & Barber*	24	F	Pain – Swelling	Sprained ankle		Wide excision	8 months' cure
10	1938	*Oberling, Vermes & Cheverau*	54	F				Excision	Relapse – Amputation 7 months later

No.	Year	Author	Age	Sex	Symptoms	Trauma	Latent period	Treatment	Subsequent course
11	1938	*Dunne*	32	M	Swelling	Contusion	9 months	Irradiation	Progression – Amputation 7 months later
12	1938	*Wolfort & Sloane*	57	F	Swelling – Pain	Contusion		Excision	Relapse – Resection 16 months later – Relapse – Amputation one year later
13	1938	*Wolfort & Sloane*	18	F	Pain	Abrasion	4 years	Curettage – irradiation	Relapse – Resection 13 months later – Relapse – Amputation one year later
14	1939	*Thoma*	19	F	Swelling			Curettage + grafting	Relapse – Amputation 8 months later
15	1939	*Rankin*	25	M	Pain – Swelling	Puncture wound	One year	Curettage	Relapse – Resection 15 months later – 2 years' cure
16	1939	*Hebbel*	14	F	Pain – Swelling			Curettage	Relapse – Recurettage 15 months later – Relapse – Amputation 2½ years after onset

17	1942	*Dockerty & Meyerding*	24	F	Pain – Swelling			Removal	Relapse – Amputation 15 months later
18	1942	*Dockerty & Meyerding*	27	M	Pain – Swelling	Bruise – Recurrent fractures		Excisions + irradiation	16 months' cure
19	1942	*Anderson & Saunders*	45	F	Swelling of the *ulna*	Abrasion – 2 previous fractures	Several months	Resection	Relapse – Amputation 5 years later– 6 years' cure
20	1942	*Bell*	16	M	Pain – Swelling of the lower end of the *femur*			Incision (The lesion was regarded as a deep intramuscular abscess)	Died in pneumonia 5 weeks later (after a rapid deterioration)
21	1943	*Cagnoli*	14	F	Pain – Swelling			Curettage	Suspicion of relapse 6 months later
22	1943	*Meffley & Northup*	16	F	Swelling	Bicycle accident	8 years	Excision + cauterization	No observation
23	1947	*Halpert & Dohn*	24	F	Painful swelling	Contusion 2 years previously – later fracture		Amputation	6 months' cure
24	1948	*Possati*			(not available)				
25	1952	*J. Hertz*	49	M	Swelling			Curettage + chiselling	Resection 18 months later – 8 years' cure

SUMMARY

A case of tibial adamantinoma is recorded, in which the histologic pattern typifies that of adamantinomas of the jaws. 24 instances of adamantinomas of the long bones have been traced in the available literature. Prognosis and treatment are outlined and a plea is made for a biopsy in the establishment of the diagnosis and for radical surgery as the therapeutic procedure of choice. Different theories concerning the pathogenesis are mentioned.

RESUME

Un cas d'adamantinome tibial est rapporté dans lequel on a retrouvé le tableau histologique typique de l'adamantinome maxillaire. 24 cas d'adamantinomes des os longs ont été retrouvés dans la littérature. Le pronostic et le traitement sont retracés et il est plaidé en faveur de la biopsie dans l'établissement du diagnostic et pour une intervention chirurgicale radicale comme étant le procédé thérapeutique de choix. Différentes théories concernant la pathogénèse de cette maladie sont mentionnées.

ZUSAMMENFASSUNG

Ein Fall eines Adamantinoms der Tibia wird berichtet, in dem der histologische Aufbau in typischer Weise dem eines maxillaren Adamantinoms gleicht. 24 Fälle von Adamantinom der langen Röhrenknochen wurden in der Litteratur gefunden. Die Prognose und Behandlung werden kurz besprochen und die Wichtigkeit der Probeexcision für die Diagnose und für den radikalen chirurgischen Eingriff, als die Behandlung der Wahl, wird eindringlich hervorgehoben. Verschiedene Theorien über die Pathogenese der Tumoren werden erwähnt.

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