

# ADAMANTINE EPITHELIOMA OF THE LOWER JAW\*

CLINICAL REPORT OF TWO UNUSUAL CASES

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IN order to comprehend the etiology and structure of the intrinsic tumors of the jaw whose origin is connected with the development of the teeth, it is necessary to review the embryology and the histology of the teeth. There is

a rather striking resemblance between the origin of the hair and the teeth. Scudder has called attention to this analogy (Scudder, C. L., *Tumors of the Jaw*, 1912, pp. 163 and 165). In the first stage of specific development of both hair and teeth, the earliest change noticed is that the deep layers of the epidermis form buds and grow into the tissue beneath it, dipping into the mesenchyma of the alveolar processes. At the same time, a papilla arises from the mesenchyma which grows up into the epithelial cord.

One of the early stages in the embryology of the teeth is represented by Fig. 1. In a 40-mm. embryo about the beginning of the third month, a shelf of epithelial tissue, a development of the original bud or cord from the under surface of the epidermis, pushes down. This is called

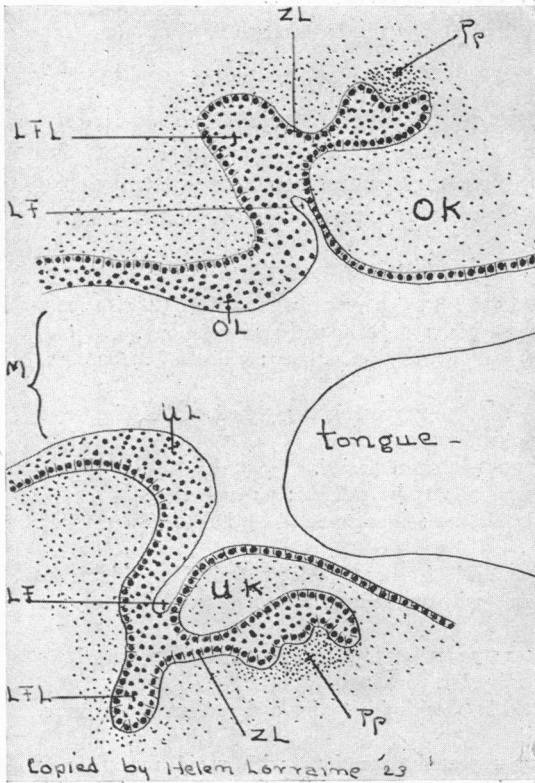


FIG. 1.—Dental anlagen from a human fetus, 40 mm. long. (Jordan, Fig. 312.) LF, dental groove; M, oral cavity; OK, mesoblast of upper jaw; UK, anlagen of lower jaw; OL, epithelium of primitive upper lip; and UL, of the lower lip; ZL, dental lamina; LFL, labial lamina; Pp, dental papilla. (X 60.) (After C. Röse.)

the dental lamina. During the third month of foetal life the anlagen of all the temporary teeth are laid down. About the same time a thin portion of this dental lamina buds out and forms the anlagen of the permanent teeth. A papilla from the mesenchyma pushes up into the cord of epithelium as shown

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in Fig. 2. This epithelial cord is converted into an isolated closed sac, the enamel organ, by the development of a membrane, the dental sac, which severs the epithelial bud from its original stalk and at the same time surrounds the papilla from below. The primitive tooth thus lies within the dental sac which develops about the end of the third month (Fig. 3).

The enamel organ sits upon the papilla as a cap and the papilla pushes into this closed sac. Within this sac is a rather soft material called the enamel pulp. In the earlier stages the sac of the enamel organ reaches almost to the base of the papilla.

From the papilla is developed the dentine, which is the hard ivory substance of the tooth, and upon the dentine in that portion of the tooth which is above the gum rests the enamel, consisting of very hard prisms. These arise from the layer of the enamel organ in contact with the papilla, and are formed by the cylindrical cells nearest the papilla called ameloblasts, into which calcium salts, chiefly calcium phosphate, are deposited, making a substance of extreme hardness. The outer layer of the root of the tooth, or the cementum, is developed from mesenchyma and is formed by ossification of that portion of the wall of the dental sac which covers the base of the dental papilla and the root of the tooth. The cementum has no epithelial connection.

The dentine, on which the enamel and the cementum rest, is developed from cells called odontoblasts, and is penetrated by an abundant system of canals containing branches from the odontoblasts and nerves.

The enamel pulp consists of soft material within the enamel sac and at first seems to resemble syncytial tissue. As the tooth develops, the cells take form and become stellate with long anastomosing processes. No blood-vessels penetrate the enamel organ, though blood-vessels, nerves and lymphatics communicate freely with the papilla through the dental foramen below. (Fig. 4.)

The outer layer of the enamel organ which lies on the interior of the

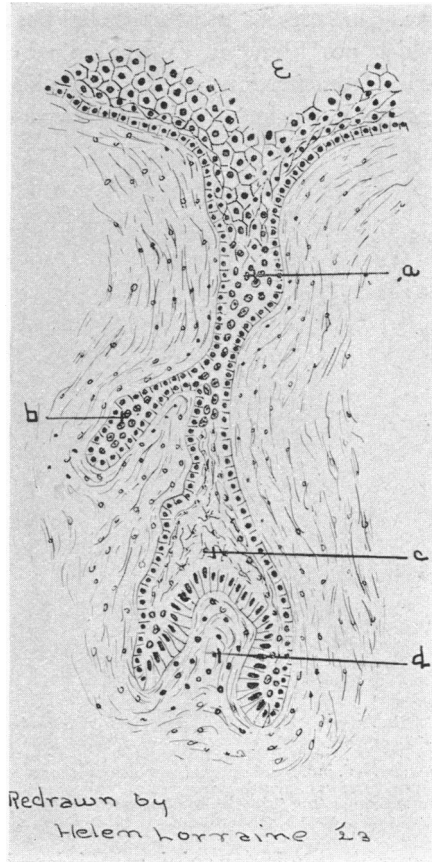


FIG. 2.—Vertical section of cuspid of human fetus, showing the budding of the primitive epithelial cord (X 70) (Marshall's "Operative Dentistry") (Scudder, Fig. 152). A, primitive epithelial cord; B, budding of the primitive epithelial cord; C, enamel organ; D, dentin papilla; E, epithelial cells.

dental sac except at the base of the tooth forms several layers of flattened epithelium. According to Jordan (Jordan, H. E., Text-book on Histology, 2nd edition, 1920, page 335), "Remnants of this cell layer frequently persist in relation to the inner margin of the bony alveolus." The bony alveolus is formed by ossification of the connective tissue around the embryonal dental sac. Such remnants of epithelial tissue can give origin to the epithelial tumors which are found in connection with the teeth. The dental root cyst may arise from portions of this epithelium attached to the root of the tooth and carried down into the jaw by the development of the root. The dentigerous

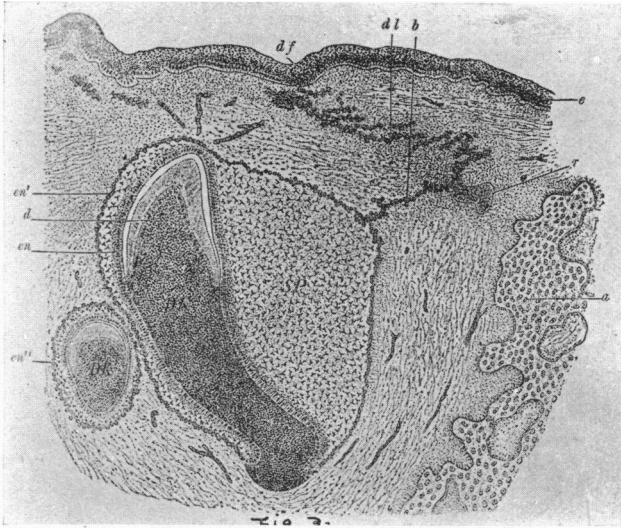


FIG. 3.—Section of first milk incisor of a human embryo 30 cm. long. Frontal section through lower jaw. (X 30.) (Röse.) (Schaffer, Fig. 739.) DK, tooth pulp; d, odontoblasts; a, bone of alveolar process of jaw; en, en', inner and outer layers of enamel organ; SP, enamel pulp; df, dental furrow; e, mouth epithelium; dl, remains of dental lamina; b, cell bridge connecting this with tooth germ; r, reserve germ for permanent tooth; en'', germ of 2nd milk incisor cut obliquely.

cyst comes from the abnormal development of the dental sac.

One of the most interesting tumors of the jaw is the adamantinoma, or adamantine epithelioma, to which the excellent work of Bloodgood has called attention. This tumor, while unusual, is not rare. In its development, columnar cells are conspicuous. These are supposed to be analogous to the ameloblasts of the enamel organ and often form what

resemble tubules or acini, containing, however, a structure like the enamel pulp with poorly differentiated star-shaped cells, or, in some instances, apparently squamous epithelial cells. Not infrequently this material forms a cyst which may enlarge, probably by coalescence with adjoining cysts, and make a cavity of considerable size. Some authors, as Broders, A. C., and MacCarty, W. C. (Epithelioma, Surgery, Gynæcology and Obstetrics, vol. xxvii, August, 1918, pp. 141-151), and Buchtemann and Kolaczek, think that the adamantinoma develops directly from the epithelium of the gum and not from the enamel organ. The accompanying two photomicrographs, kindly given by A. C. Broders (Figs. 8 and 9), are very suggestive of such an origin. The columnar cells that are found in adamantinoma resemble somewhat the basal cells of the epidermis of the gum.

According to Broders and MacCarty, the stellate cells, which are supposed to correspond to the enamel pulp, may resemble the prickle cells of the

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epidermis. Then, too, they assert that cysts similar to those usually found in adamantinomas are also seen in squamous-cell cancers as a result of the degeneration of cells. As the enamel organ arises from the under surface of the epidermis of the gum, it is quite possible that the adamantinoma may come directly from this tissue. There are many basal cell cancers in which this same type of columnar epithelium of an adamantinoma, corresponding to the basic or germinal layer, is found. (Figs. 10 and 11.) The clinical course of adamantinoma, however, is quite different from that of basal-cell cancer.

Of the two cases reported here, the first seems a direct descendant of remnants of the enamel organ. In the other, which contains what appears to be epithelial tissue, the origin is more obscure.

CASE I.—Mrs. J. E. W., aged twenty-nine, referred by Dr. B. F. Royal, of Morehead City, N. C., entered St. Elizabeth's Hospital on January 16, 1919. She had ptosis of the stomach and transverse colon, cholecystitis, and a cyst in the right lower jaw. This was thought to be a dentigerous cyst. The cyst was first noticed in 1917, though there had been pain in the lower jaw for the past eleven years. The pain was frequently rather severe. About nine months ago some operation was done by another surgeon, and a few months later operation was performed upon the cyst. The technic of these operations could not be determined, but it was doubtless an incision from within the mouth. No permanent relief was obtained. On

January 17, 1919, I operated, plicating the gastrohepatic and gastrocolic omentum and removing the gall-bladder, which was very adherent. After completing the abdominal operation, a transverse incision, on a level with the lower portion of the ear, was made over the cyst in the jaw. The cyst seemed to be about two inches in diameter. It appeared bluish in color, and there was no bone covering this portion of its exposed wall. The cyst was incised, and more than an ounce of slightly muddy fluid was evacuated. The cyst wall was removed as far as possible by curetting. The cavity was filled with the Moorhof iodoform plug, and the wound was closed in layers with tanned catgut. Convalescence was satisfactory except that some of the Moorhof plug worked through the incision and the scar was very prominent. On February 12, 1919, the scar was excised under local anæsthesia,

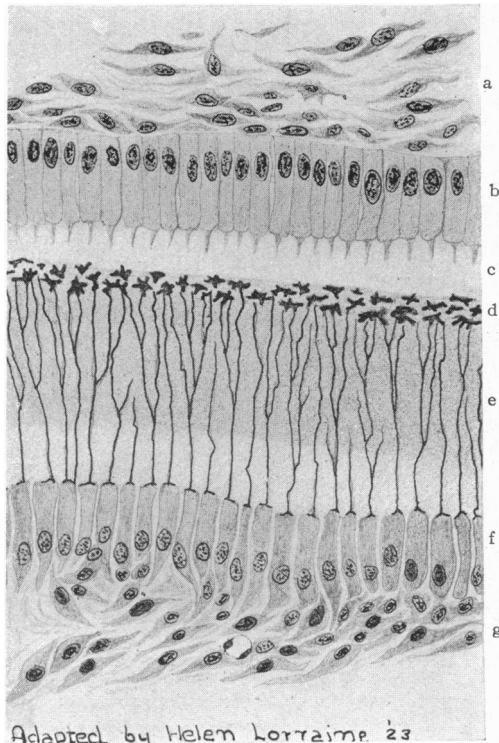


FIG. 4.—Section of developing tooth through junction of enamel and dentine. (X 400) (Modified from Piersol.) a, intermediate layer of enamel organ; b, ameloblasts; c, young enamel with Tomes' processes; d, granular layer Tomes; e, dentine; f, odontoblasts; g, embryonal pulp-tissue.

and the wound united with a subcuticular stitch of silkworm gut, over which was placed an epidermal suture of fine silk. She was discharged February 17, 1919.

The patient returned on May 31, 1923. The growth on the jaw had increased in size. After leaving the hospital in February, 1919, she felt well for several months; then pain of a dull aching character began in the left lower jaw, and occasionally extended down her neck and to the back part of her head. During the last six months there has been a marked increase in the size of the growth

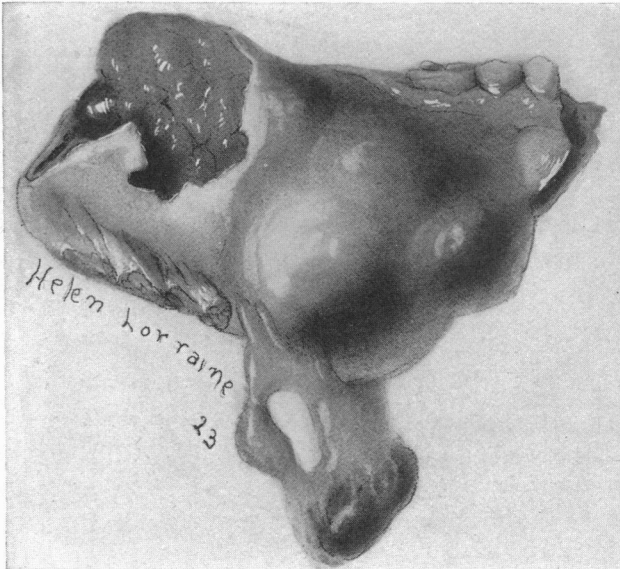


FIG. 5.—Drawing of specimen removed from Case I, external view. On the right is a small margin of healthy bone with the alveolar process containing three teeth. On the left the tumor has been exposed during removal of the growth, and below attached enlarged lymph-nodes which on section were merely hyperplastic.

in the lower jaw, which for a time after the operation was hardly observable. The pain has also increased.

On inspection there was a large growth in the right lower jaw. Röntgenographic report by Dr. Fred Hodges follows: "There is an extensive cystic involvement of the bone extending from the first bicuspid tooth back beyond the angle of the jaw on the inferior border; and on the superior border, practically back to the con-

dyle of the jaw. The jawbone is practically entirely destroyed, and there are a good many bony septa dividing this area into many different parts."

On June 1, 1923, the patient was operated upon under gas-ether. An incision was made from about the middle of the right mastoid process downward, forward and inward, along the border of the lower jaw to the middle of the neck. The tissues were dissected down, and the facial vessels were doubly clamped and divided. The whole mass of tissue was dissected up, taking care to keep close to the jawbone. The wall of the tumor was exceedingly thin, and at one point anteriorly was opened. A small amount of thick, clear, yellowish fluid containing some flocculent particles was evacuated. This was washed off with sterile water. The dissection was carried beneath the jaw, and the attachment of the muscles to the right half of the jaw was severed. Anteriorly there were a few enlarged lymph-nodes, and these were left attached to the tumor. The incision was carried through the lower lip and the bone was divided anteriorly, about one-half inch in front of the diseased tissue. The growth was then retracted outward, and the dissection was made internally, preserving as much mucosa as possible and packing the mouth with gauze to prevent aspiration of blood. The bone was severed posteriorly with bone forceps. The growth involved the anterior portion of the ramus of the jaw. A small fragment was left at the upper part, and this was removed separately. The raw surface at the stump of the bone was cauterized with

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the electric cautery. The mucosa of the mouth was sutured with a continuous suture of tanned catgut. The skin wound was closed with a continuous suture of silkworm gut, leaving a drainage tube through the posterior end of the wound. No effort was made to fill in the defect left by removing the jaw, as the mouth cavity had been widely opened and it was felt that a graft would not take at this time. The operation lasted one hour and twenty minutes.

The specimen consists of the tumor from the lower jaw, together with one lymph-node which is enlarged and soft. The tumor is three and one-half inches long, by two inches wide. On the upper surface is the alveolar process with three lower teeth anteriorly. They seemed quite firmly imbedded in the gum. Externally most of the growth consists of a bulging out and thinning of the bone until at most places the bone is merely the thick-

ness of parchment and over some areas has disappeared. Both anteriorly and posteriorly there is about one-half inch of healthy jawbone, except at the upper portion posteriorly, where the section was made partly through diseased tissue and the small remaining fragment of diseased tissue removed with bone forceps. (Fig. 5.)

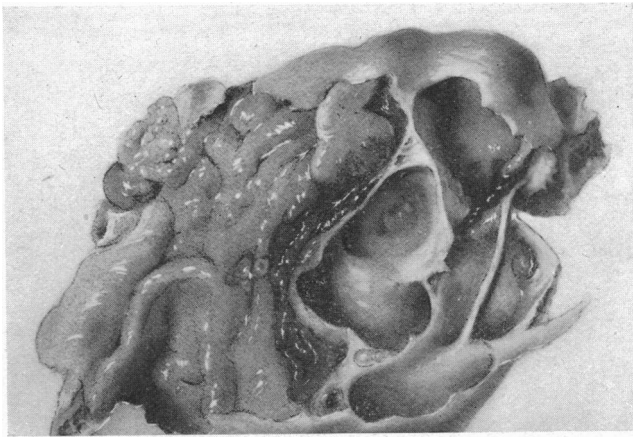


FIG. 6.—Drawing of specimen shown in Fig. 5. An incision has been made along the outer wall. Note the thin capsule of the growth, and in the right side a cyst containing smooth walls with imperfect partitions. Through about the centre of the specimen is a complete partition, and on the left is soft content. To the left of the central partition the material is a dark purplish color. In other portions the solid material is yellowish.

Longitudinal incision with a knife shows in the lower half of the specimen several cavities whose walls are lined with a thin membrane, beneath which is a very thin layer of bone. The bone can be readily dented with the finger. There is a small bridge of bone along the alveolar process containing the teeth. Just in front of what corresponds to the angle of the lower jaw and internally the wall of the tumor is very thin. The cysts communicate, and there are incomplete partitions between them. The wall is smooth. In the upper posterior portion of the specimen, there is a solid mass of tissue which fills a compartment. This is rather soft and most of it is a pale yellow, though anteriorly and below the tissue is purplish in color. All of the tissue can be easily separated from what appears to be the capsule, which resembles very much the wall of the cysts that contained fluid. (Fig. 6.) The lymph-node on section is oblong, about an inch in diameter, and soft. It seems to be hyperplastic. Microscopic sections in celloidin show typical structure of adamantine epithelioma, or adamantinoma. The columnar cells are abundant, and are arranged somewhat irregularly as tubules or as acini. In portions of the growth there is a myxomatous-like material with stellate cells resembling enamel pulp. (Fig. 7.) The patient made a satisfactory recovery from the operation, and left the hospital on June 20, 1923.

Under date of December 6, 1923, in a letter from Doctor Royal, the patient's family physician, he says that soon after returning home the patient had a small

sinus in the chin, which led down to the bone. He dilated and curetted this sinus, and it promptly healed. He has not seen her very recently. He says the deformity is inconspicuous, being only a slight flattening, and the scar is hardly visible as it lies largely in the fold of the neck.

Under date of December 8, 1923, the patient writes me that so far as she can tell there is no recurrence, but that the jaw pains her some, and "swells when I eat or worry." The swelling may be due to the action of the parotid gland.

CASE II.—This patient, Mrs. A. R. T., aged forty-one, referred by Dr. J. Bolling Jones, of Petersburg, Va., entered St. Elizabeth's Hospital, on May 19, 1918. The family history was of no significance. She first noticed some swelling

under her tongue on the right side of the mouth about twenty years ago. This, however, did not trouble her until the last few years, when it enlarged and the jawbone apparently was affected. She had been operated upon about two years before this by another surgeon, who had removed a portion of the alveolar process from the lower right jaw.



FIG. 7.—Photomicrograph of the tumor shown in Figs. 5 and 6. The columnar cells of which this growth is largely composed are arranged somewhat irregularly as tubules or acini. In the lower right corner is part of a mass of tissue resembling somewhat enamel pulp. Areas of this kind are common in the section, and often are of large proportions. (X 150.)

There was a recurrence of the growth. It involved the lower jaw on the right side, extending from a short distance

from the symphysis back to near the angle of the jaw. It was rather sharply circumscribed, though it apparently infiltrated the immediate tissue along the adjacent portion of the floor of the mouth.

An incision was made along the lower border of the right side of the jaw, extending up over the chin in the midline. The bone near the midline was divided with a wire saw. The growth was dissected out with the electric cautery, taking care to include the adjoining adherent tissue. The jaw was sawed posteriorly at the angle. A graft was taken from the tibia. The ends of the graft and the stumps of the bone of the jaw were drilled and the graft was fastened in position with kangaroo tendon. An effort was made to suture the mucosa of the mouth over the graft. There was some tension in the suturing, and the mucosa could not be satisfactorily approximated. The wound was closed and a small drain of catgut was inserted at the posterior angle.

The specimen consists of the right side of the lower jaw extending from near the symphysis to a point just anterior to the angle of the jaw. The specimen

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measures two and one-half inches in length. At the two extremities there appears to be healthy normal bone. Between these there is a tumor which bulges the bone in all directions. On the internal surface of the tumor a small amount of soft tissue is adherent. This presents a charred appearance resulting from the dissection with the electric cautery. There is a very thin capsule of bone around most of the growth. At other points the bone seems to be entirely missing, and is replaced by a fibrous capsule. Where the bone is present it is exceedingly thin and can be easily dented with the finger. (Fig. 12.) On section the tumor seems to be solid without any cystic formation. The jawbone has been expanded and absorbed, so that the thin capsule can be readily cut with a knife. The tumor has a yellowish appearance with occasional red specks. It is moderately firm, but elastic. It is not encapsulated, though it is distinctly circumscribed. It appears



FIG. 8.—Photomicrograph of adamantinoma showing apparent origin of growth from the epithelium of the gum. (X 50.) (From A. C. Broders.)

to displace the tissue as it grew, though there is some infiltration at certain areas. This, however, is limited. (Fig. 13.) A block is taken for celloidin section. (Fig. 14.)

Infection followed, and the graft had to be removed. The wound gradually healed, and the patient left the hospital on June 21, 1918.

On October 1, 1920, she was admitted to the hospital, and a sebaceous cyst of the scalp was removed and a small enlargement on the alveolar process of the left side of the jaw was removed with a curette and the tissues were thoroughly cauterized. There was no evidence of any growth on the right side. Microscopic examination did not show this tissue to be a recurrence.

On September 23, 1921, she was admitted with a small recurrence, and operated upon. The anterior part of the incision in the neck which was made in the first operation was incised and the end of the jawbone was exposed. A small portion was cut away with bone forceps. The bone appeared to be normal. There was a nodule in the floor of the mouth on the right side about an inch from the bone. This was dissected out without opening the mucosa and the tissue was examined with frozen sections. The nodule was firm and about one-quarter inch in diameter. A frozen section showed malignancy. The tissues around the nodule were thoroughly cauterized with electric cautery without penetrating to



the mouth. The wound was closed with silkworm gut, leaving a rubber tube for drainage. A small nodule on the exterior surface of the bone about two inches from the midline was exposed and curetted. There appeared to be no evidence of malignancy. This wound was thoroughly cauterized with the electric cautery. The operation lasted fifty minutes.

The specimen consists of the small piece of tissue described in the operation. A gross section shows a firm, yellow growth that is not encapsulated. There is a small amount of muscular tissue attached. Microscopic section shows the growth of the same nature as the original. The histologic appearance of the original tumor removed by me was so unusual that I was unable to make a definite diag-

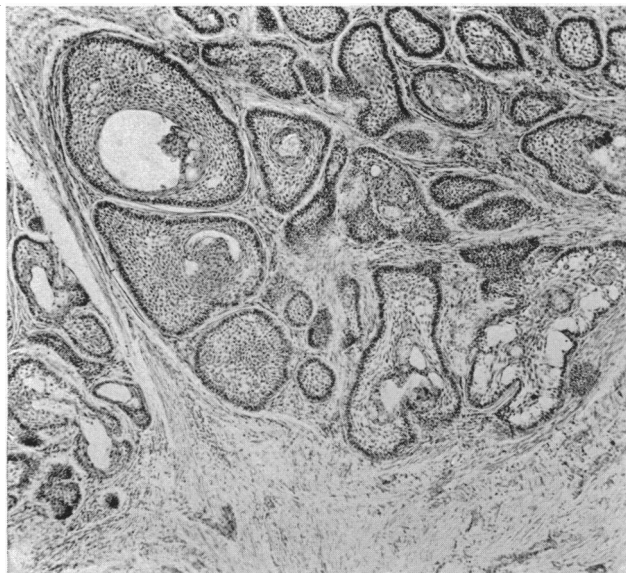


FIG. 9.—Section from the central portion of the tumor shown in Fig. 8, which gives a more characteristic appearance of adamantinoma. (X 50.) (From A. C. Broders.)

nosis except that it was malignant. Sections or blocks of tissue were submitted to several prominent pathologists.

Dr. J. C. Bloodgood, under date of August 2, 1919, said: "This section shows tubular alveoli of cells whose nuclei take the hæmatoxylin stain, imbedded in a very fibrous stroma. These alveoli are not all tubular; some are irregular masses and the question is what these cells are. They do not resemble adamantine epithelium. They show more pearly body formation. Many of the cells are vacuolated. They do not appear to be a type of sarcoma. I am inclined to think that the diagnosis rests between a carcinoma arising in the gum, or one of the rare forms of adamantine epithelioma, in which the peculiar degeneration is not well marked. I shall diagnose it *jaw adamantine epithelioma* until I hear more definitely from Doctor Horsley."

Dr. A. C. Broders, of the Mayo Clinic, under date of July 15, 1918, says: "From the tissue that you sent from Mrs. T. some time ago, I have made a number of slides, staining some with hæmatoxylin and eosin and also some for fat. Those stained for fat showed the tumor cells to be free from it. It is extremely difficult to say whether this is a sarcoma or carcinoma. At any rate, it is a malignant tumor, though apparently of a low grade." Later, after another study of

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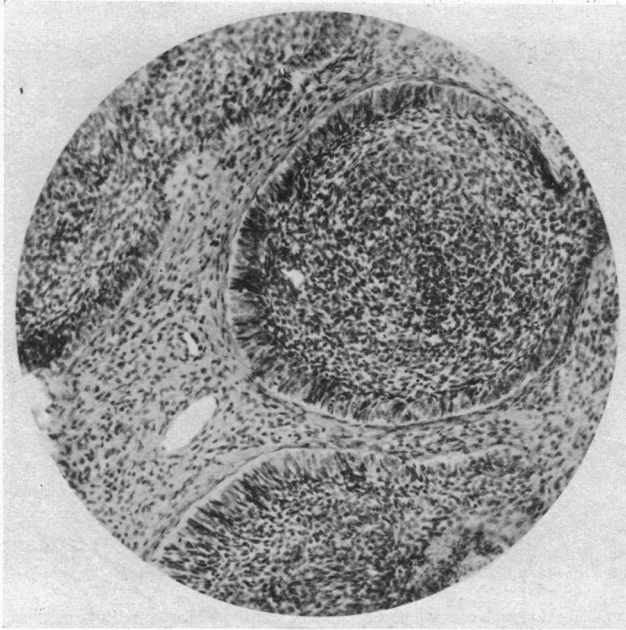


FIG. 10.—Basal cell carcinoma from the skin over the eyebrow showing columnar epithelium similar to that found in adamantinoma, though the cells surrounded by the columnar epithelium are of a different type from the enamel pulp. (X 150.)

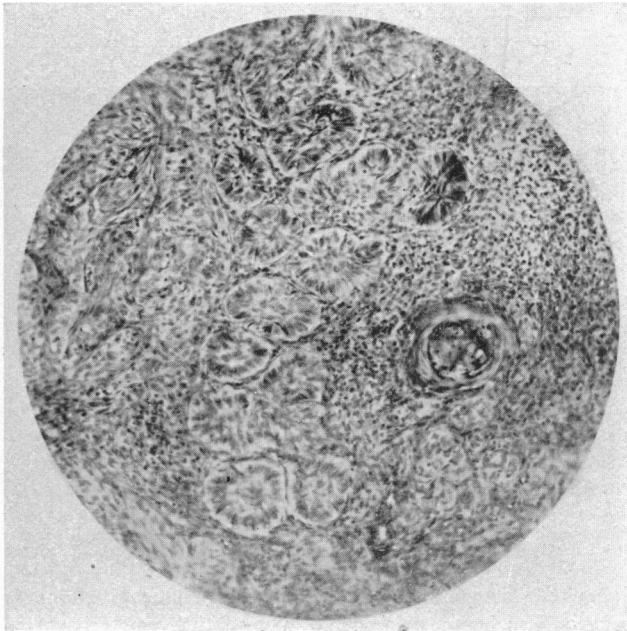


FIG. 11.—Photomicrograph of small growth removed from the skin near the inner canthus of the eye. It shows in the upper portion of the photograph spinous cell type of cancer with a "pearl". In the central portion it is a basal cell type, with columnar cells resembling those found in adamantinoma. (X 150.)

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this tissue in August, 1923, he said the tissue showed "epithelioma belonging to the squamous-cell family of adamantine type, of low-grade malignancy," and

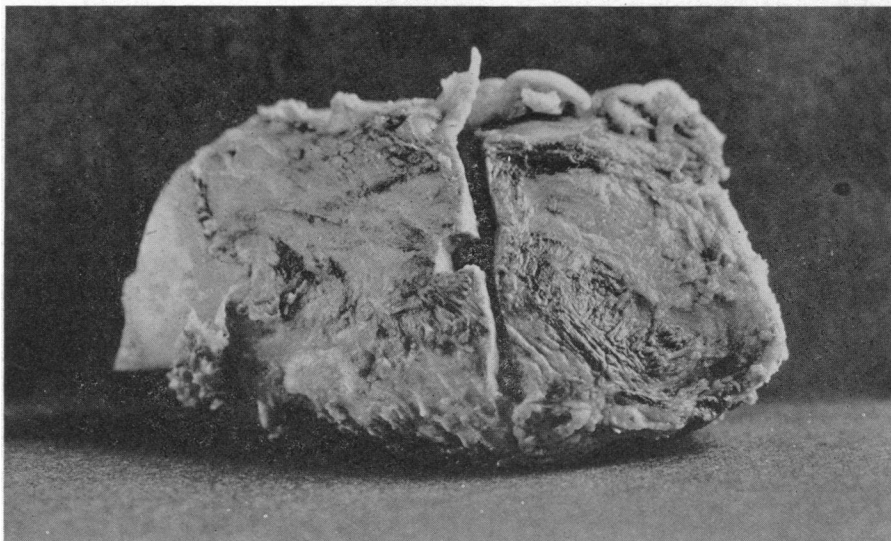


FIG. 12.—Photograph of specimen removed from Case II. At both ends there was apparently healthy bone.

that a section removed from the last operation showed a recurrence of the original growth.

Dr. Alfred Stengel, of Philadelphia, was interested in the growth and was good enough to go over the sections. His report, under date of December 11, 1921,

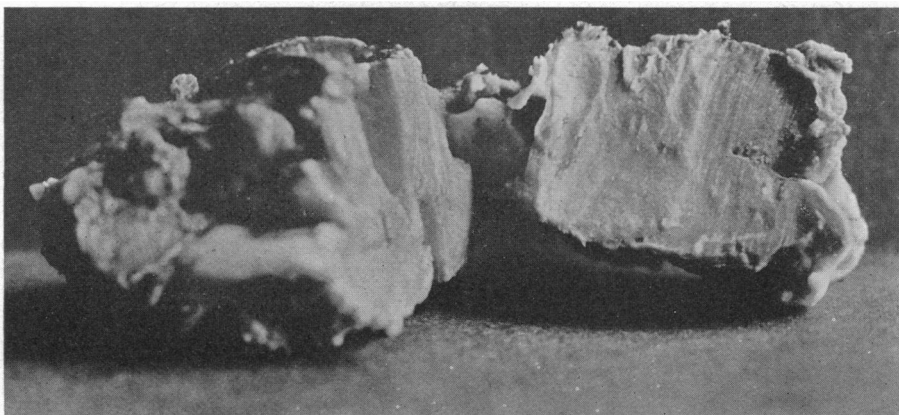


FIG. 13.—Photograph of a section of the tumor shown in Fig. 12. The capsule is thin. The bone in some areas has entirely disappeared. The growth infiltrates as well as expands the surrounding tissue.

is as follows: "The microscopic slide of the very interesting tumor regarding which we spoke at Lynchburg reached me in good time and I have been examining it with several of my colleagues here who were as much interested as I. Dr. Herbert Fox and I first looked it over and concluded that it was not an adamantinoma, but

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we could not quite agree what we did think it. Both of us were impressed with the curious resemblance of the cell groups to those of certain hypernephromata. I was rather of the opinion that the cells have an epithelial appearance, the stroma being, of course, a very curious feature as well. Doctor Fox, I think, was rather inclined to believe it a dental tumor of some sort, though he did not express nor was he willing to express a positive opinion. We then submitted it to Dr. Hopewell Smith, the histologist of the Dental School, and Doctor Ivy of the Dental School, both of whom examined it carefully and stated that they did not believe it a dental tumor of any sort.

Neither of them, however, was willing to commit himself to a positive opinion as to what the nature of the growth might be. All of us, of course, agreed that it was a form of malignant tumor. I confess that I have never seen anything like it and am no more able to reach a conclusion than were the others. The epithelial or endothelial character of the large cell nests impressed me, but I hesitate to say what I would call the thing if I were pinned down to an expression of a positive opinion.

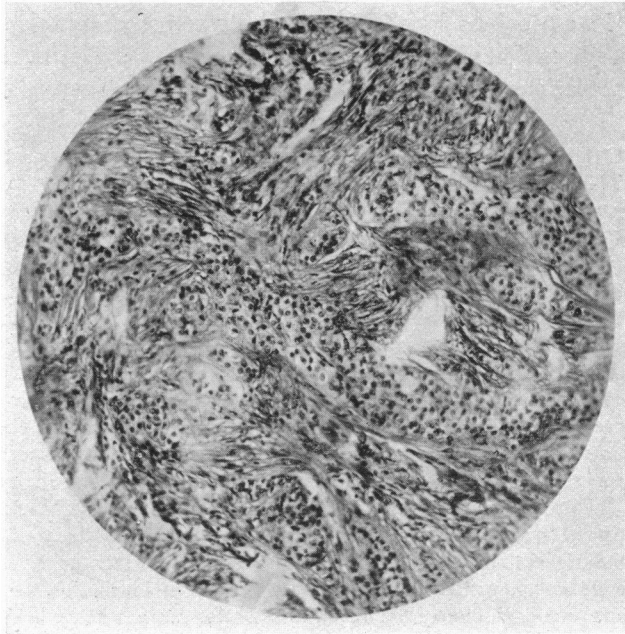


FIG. 14.—Photomicrograph of tumor shown in the preceding figures. The portion from which the block was removed is shown in the photograph. The cells seem fairly well differentiated. The cytoplasm is clear. The cells are arranged irregularly in columns or are grouped together in masses. The stroma is well organized but delicate connective tissue. The growth is evidently malignant, though of a grade of malignancy. Most of the pathologists are inclined to think it springs from the tooth ramus. (X 140.)

I believe that Dr. Joseph McFarland also looked at it, but did not come to any conclusion regarding its nature."

The exact nature of the growth seems difficult to determine. Most of the pathologists feel that it is an epithelial growth, probably of adamantine origin, though an endothelial origin can not be entirely discarded.

Under date of December 6, 1923, Dr. J. Bolling Jones, of Petersburg, reports that he has examined Mrs. T. and finds the condition satisfactory, except for "A distinctly enlarged gland the size of a cherry at the posterior inferior angle of the scar on the neck. It is unattached to the skin or scar, and is freely movable and apparently has very little deep attachment. It is in very close proximity to the common carotid, yet apparently not attached to it." Doctor Jones says that according to the patient's statement this lymph-node has been present for eighteen months, and she does not think it has changed in size. In his opinion it would be impossible to say whether this is a recurrence without a microscopic examination.