AN INSTANCE OF ADAMANTINOMA OF THE JAW WITH METASTASES TO THE RIGHT LUNG*

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Since Guzack [1] in 1826 described the first case of cystic tumor of the lower jaw,† and Falkson [2] in 1879 gave the first full account of this condition, at least 130 cases of adamantinoma have been recorded.[3-20]‡ Adamantinomas are tumors probably arising from the so-called "paradental epithelial débris," first described by Malassez.[21] The enamel organ develops by a downward growth of the gingival epithelium. All of the epithelium of the enamel organ, except its internal layer, undergoes atrophy and is absorbed. Occasionally these cells persist and it is believed that they are the histogenetic precursors of adamantinomas. The structure of the adamantinoma closely resembles the embryonal enamel organ.

Most investigators consider the adamantinoma a benign tumor.[22] It may, however, undergo malignant transformation, and where this occurs metastases having the histological structure of the malignant growth have been reported (Aschoff,[23] Ewing,[24] Kaufmann,[25] Heath,[26] Krompecher,[27] and others). Such malignant transformation develops usually after several local recurrences, following repeated excisions. From an analysis of the literature it is apparent that metastases from an adamantinoma are extremely uncommon. Eve [28] in 1883 described an unusual instance of a cystic tumor of the lower jaw of thirteen weeks' duration in a woman 60 years of age, who died of postoperative bronchopneumonia. Metastases to the lumbar lymph nodes were noted. Simmons [29] in 1927 reported two cases of adamantinoma with metastases. In one instance the metastases were in the regional lymph nodes, appearing fourteen years after the onset of the disease. In the second instance metastases were present in the glands of the neck and in the lungs. In both instances the patients

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† Sculter, cited by Albarran, J., *Rev. de chir.*, 1888, **8**, 429, is said to have described cysts of the jaw in 1654.

‡ Coryllos (ref. 3), gives a complete bibliography of the literature on this subject up to the year 1910.

had undergone operations for resection of the primary tumor. Unfortunately Simmons does not describe the histological structure of the nodules in the lungs. In the instance reported by Ewing [24] of an adamantinoma with metastases to the lung, the histological structure of the metastatic deposits was that of a malignant growth.

The following instance of an adamantinoma is reported because of the unusual metastatic lesion in the lung.

Case Report

An adult of 38 years was admitted to Montefiore Hospital on Sept. 3, 1930, with the chief complaint of a productive cough. Twenty-one years prior to the present admission, he first noted a lump in the right upper jaw. At that time excision of the tumor was attempted and five subsequent operations were performed, due to recurrence of the local lesion. In 1925 a small, soft, reddish tumor of the right upper jaw in the scar of a previous operation was removed at the Haggard Clinic in Nashville, Tenn. The area was cauterized and radium needles were implanted in the surrounding tissues. Pathological examination at that time showed the tumor to be an adamantinoma. A few months later a radical excision was performed. At the time of the operation the antrum was filled with a brain-like material which penetrated into the nasal cavity and involved the septum. Intensive radium treatment was given and the local condition healed. For a period of five years following this operation there was no evidence of recurrence locally. Two years ago, however, after an attack of "dry pleurisy," which confined the patient to bed for three weeks, he developed a productive cough with occasional blood-streaked sputum. About a year ago he complained of progressive weakness, anorexia, marked loss of weight and pain in the right hip. An X-ray of the chest taken at the Lebanon Hospital revealed a dense mass in the lower lobe of the right lung, extending from the sixth rib to the diaphragm and obscuring the right half of the latter. In the base of the left lung a circumscribed, moderately dense nodule the size of a large marble was also noted. Nothing abnormal was revealed in the X-rays of the right hip, pelvis and vertebral column. He was transferred to Bellevue Hospital where he received X-ray therapy for the lung condition.

At the time of admission to Montefiore Hospital, examination of the patient revealed a transverse and vertical scar on the right cheek. There was evidence of a partial resection of the hard palate and the right upper jaw, with a fairly large cavity in the roof of the mouth on the right side, measuring about 3 by 3 by 3 cm. opening into the nasopharynx. There was no evidence of recurrent growth. On examination of the chest there was a slight retraction of the fourth, fifth and sixth intercostal spaces over the right anterior wall. Expansion was slightly limited on the right side. Vocal fremitus was increased anteriorly from the third intercostal space downward and markedly diminished from the level of the eighth dorsal vertebra to the base. There was flatness posteriorly from the apex of the axilla to the seventh dorsal vertebra. Posteriorly the breath sounds were entirely absent below the seventh dorsal vertebra. Examination of the rest of the body revealed no abnormalities.

Nine months after admission the patient developed a septic temperature and signs of an empyema on the right side. A thoracotomy was performed and three

subpleural abscess cavities were drained. Following this his temperature dropped to normal. On histological examination of the tissue removed at operation a diagnosis of metastatic adamantinoma was made. Three months later he had a hemoptysis of 5 oz. of bright blood and continued to have blood-streaked sputum. A bronchoscopic examination, which was performed at that time, revealed a growth blocking the bronchus leading to the right lower lobe, from which bright red blood was oozing. Section from the tumor revealed the structure of an adamantinoma. Two days later he developed chills and a septic temperature, and for the first time a systolic and diastolic murmur were heard over the apex of the heart. There was no evidence of cardiac enlargement. An acneiform eruption then appeared over the chest and back. A culture of *Streptococcus viridans* was isolated from the blood stream. The patient became irrational and died.

Laboratory data showed no important findings other than the positive blood culture. Roentgenological examination of the maxillae and mandibles revealed no gross pathological changes other than those caused by the operative excision of the right upper maxilla. The mass in the right lung decreased slightly in size following a course of radiotherapy but subsequently again increased in size. The small nodule in the left base, after slightly increasing in size, completely disappeared. Roentgenological examination of the skull and skeletal system revealed no abnormalities.

Autopsy Report

Anatomical Diagnoses: Adamantinoma of the right upper jaw (postoperative) with metastases to the lower lobe of the right lung; polypoid thrombus attached to the auricular surface of the mitral valve with perforation of the leaflet; *Streptococcus viridans* septicemia with localization of the streptococci on the thrombus; splenomegaly; chronic passive congestion of the viscera; bronchopneumonia of the right lower lobe.

The body was that of a well developed, well nourished, adult white male about 165 cm. in length. There were a few petechiae in the inferior conjunctival sac of the right eye. There was anemia of the conjunctivae and lips, and cyanosis of the nail beds. There were no palpable lymph nodes in the neck, axilla or groin. Extending backward from the right angle of the mouth for a distance of about 2 inches there was a healed scar. The right cheek sagged inward as most of the maxilla and antrum wall had been removed from the right lateral incisors backward. The mucous membrane of the right cheek presented a healed scarred appearance. There was no evidence of ulceration. On opening the chest and abdomen, the dome of the diaphragm reached the third intercostal space on the right side and the fourth rib on the left. There was no fluid or air in either pleural

cavity. The right lung was closely adherent over the lower lobe posteriorly, laterally, and at the base.

The heart was not enlarged, and weighed 260 gm. The parietal pericardium adjacent to the lower lobe of the right lung was closely adherent to it. There was a moderate amount of epicardial fat which was well defined from the musculature. The myocardium was reddish brown in color and firm in consistence. The right auricle and ventricle presented no abnormalities. An occasional small, yellowish patch was seen through the endocardium in the right ventricle. The endocardium of the left auricle was somewhat opaque and grayish. Attached to the auricular surface of the aortic cusp of the mitral valve, by a broad base which measured 1.5 cm. in diameter, was a lobulated mass about the size of an olive. It had a somewhat smooth, greenish gray surface to which was adherent some dark red, clotted blood. The mass was fairly firm, somewhat elastic and quite friable. Most of the chordae tendineae of the aortic leaflet were markedly thickened and on the aortic surface of the aortic leaflet directly opposite the polypoid mass there was a punched out ulceration about 1 cm. in diameter. Its edge was fairly smooth and was covered in places by a small amount of grayish, friable material. The base of this ulcer around the periphery could be probed for a distance of about 8 to 10 mm. into the polypoid mass. In the central portion at a depth of 2 to 3 mm. a greenish gray mass similar to that of the polyp was present. This material was adherent to the lower margin of the ulceration. Clotted blood was removed from the ulcerated region. The tips of the papillary muscles were firm, and grayish white in appearance. There were many grayish streaks seen in the endocardium of the left ventricle.

The upper and middle lobes of the right lung were crepitant throughout. The pleura was smooth and glistening and presented no abnormalities. On section, the surface was reddish gray, mottled with anthracotic pigment with an occasional darker red area of congestion. A large amount of well aerated serous fluid and blood exuded on pressure. Bronchi and pulmonary vessels of these lobes presented no abnormalities.

The entire posterior part of the lower lobe was torn on removal, with marked destruction of the parenchyma. There was a large area in the upper anterior portion which was firm and non-crepitant. On section, the lung pattern was completely distorted and the alveolar

portions were markedly diminished in amount and obscured. The bronchi were dilated and very prominent. In the lower portion of the lobe they formed large bronchiectatic cavities with smooth-ribbed walls and with bands running through them. The whole bronchial tree, wherever it was intact, was plugged with a cast composed of fairly firm, somewhat friable, grayish white tumor tissue. In some areas this plug measured 2 to 2.5 cm. in diameter. Although for the most part this cast lay free in the bronchi, in some areas the walls of the bronchi seemed to be invaded. The cartilages of the larger bronchi were somewhat thickened in some areas and in others thinned out. There was an occasional large patch of the remaining alveolar portion which was completely replaced by tissue similar to that found in the bronchi. This tissue seemed to invade the bronchial wall and in places the mucosal surface was raised by irregular, grayish white patches. The upper anterior portion of this lobe was completely consolidated. On section the surface was yellowish gray and translucent, with areas of necrosis and many small cavities surrounded by reddish zones.

The left lung was similar to the upper and middle lobes of the right lung, with the exception of a small nodule found in the middle of the lower lobe. This nodule was about 1.5 cm. in size, well defined from the rest of the parenchyma, and seemed to be composed of firm yellowish material. There was a small amount of fibrosis around it.

The spleen weighed 220 gm. and measured 14.5 by 9 by 3.5 cm. The organ was enlarged, soft and somewhat flabby in consistence. The capsule was bluish gray in color and presented no abnormalities. On section the surface was purplish red in color with many irregular areas of hemorrhage and congestion. The corpuscles were indistinct but the trabeculae were quite prominent. The pulp scraped with ease on the edge of the knife.

There were no other abnormalities found in the rest of the viscera, other than chronic passive congestion.

Histological Examination

On microscopic examination the heart shows fragmentation of muscle fibers, cloudy swelling, small areas of perivascular edema and round cell infiltration, foci of cellular infiltration between muscle fibers composed of round cells and occasional polymorphonuclear

leukocytes. Occasionally in some areas the polymorphonuclear leukocytic infiltration predominates.

The thrombus on the auricular surface of the aortic leaflet of the mitral valve shows tissue composed of necrotic material. Large masses of bacteria and fibrin are present and the base of the polypoid mass contains very dense cellular connective tissue which is partly hyalinized. The tissue is markedly vascularized with small capillaries which are congested. There are numerous round cells and polymorphonuclear leukocytes.

Sections through portions of the lung uninvolved with tumor show extensive edema and congestion of the vessels. In some areas alveoli are filled with large mononuclear cells which contain brown pigment. Occasionally these cells are fused to form giant cells with nuclei arranged eccentrically.

An area of consolidation of the upper portion of the right lower lobe shows extensive fibrosis, atelectasis and atypical epithelial proliferation in some of the infundibula, and alveoli in the fibrotic areas. The wall of a bronchus shows very marked dilatation of the lumen (bronchiectasis), striking inflammation of the wall with extensive infiltration of round cells, polymorphonuclear leukocytes and newly formed blood vessels. One of the large blood vessels shows some thickening of the wall with adherent thrombotic material in the lumen. Areas of bronchopneumonic exudate are present.

Section through tumor of lung shows large irregular islands of tumor cells separated by dense fibrous connective tissue. These tumor masses have an outer layer composed of elongated cylindrical epithelium arranged in a vertical orientation to the surface. Occasionally there are two layers of cells of this type. The rest of the tumor masses are composed of spindle-shaped and round cells in a very loose meshwork of fine fibrillar connective tissue. The cytoplasm of the cells within the core of the nodules is scanty. An occasional fine process can be seen definitely radiating outward, communicating with similar processes in other cells (star-shaped cells). In the center of some of the islands within the core of the tumor mass the cells are arranged in a whorl resembling the beginning of epithelial pearl formation. An occasional small, cyst-like space is present within the tumor masses. There is no evidence of a malignant transformation. There is a striking similarity between the histological structure of this tumor tissue and that of the enamel organ of a four month fetus.

Other portions of lung where the tumor fills the bronchus and invades the surrounding tissue show that the tumor within the lumen of the bronchus is identical with that described above. In addition there are small areas of hemorrhage and necrosis. In other areas tumor is seen in the surrounding pulmonary tissue, invading the wall of the bronchus from within and encroaching on the cartilage within the bronchial wall. At the points where the tumor tissue invades the cartilage the tumor cells maintain their original adamantinomatous structure.

The nodule in the left lower lobe is composed of a very necrotic hyalinized material surrounded by cellular granulation tissue which is well vascularized. No evidence of tumor is present in the necrotic nodule. The surrounding lung tissue is congested and in places atelectatic and fibrotic.

The spleen shows evidence of marked congestion. The follicles are small, arterioles thickened. There are many cellular elements in the sinuses, some of which are plasma cells and round cells. An occasional erythrophagocyte is observed.

Discussion

From the review of the literature it is apparent that adamantinomas rarely metastasize. The presence of a large, secondary focus in the lung showing the histological structure of an apparently benign adamantinoma, in the instance reported in this communication is, therefore, unusually interesting.

The peculiar structure of the tumor in the lung was striking. The tumor filled the bronchial tree of the right lower lobe in a cast-like form and markedly distended the lumina of the bronchi. The bronchiectasis resulting from the inner pressure was extreme. The parenchyma, in places, and the pleura were extensively invaded. In view of this unusual manner of growth of the tumor it is suggested that tumor cells were aspirated from the primary focus in the jaw. In favor of this hypothesis are the following facts.

1. Such aspiration of tumor tissue was possible since the primary tumor in the upper jaw extended into the antrum and impinged on the nasopharynx. Furthermore, the patient had undergone several operations in which a general anesthetic had been administered.

2. The secondary tumor in the lung was present only in the right lower lobe. Jackson [30] found that aspirated foreign bodies lie most frequently in this site.

3. The growth of the tumor in the bronchial tree with the development of distention bronchiectasis further supports the theory of intrabronchial spread.

4. Both the gross and microscopic appearance of the metastasis closely resemble the primary benign growth in the jaw. Histologically, the section of the tumor in the lung is that of an ordinary adamantinoma.

5. There was no local lymph node involvement, and no metastases were found elsewhere in the body. This suggests some other method of spread of the secondary deposit than via the blood stream or lymphatics.

If the tumor was transplanted by aspiration it is necessary to assume that the tumor cells grew in the bronchial mucus secretion and in the mucosa. Histogenetically, adamantinoma cells arise from cells which are the precursors of enamel-forming tissue. While no similar or analogous instance of growth of tumor cells in such a tissue culture medium *in vivo* is known to the authors, it is probable that these cells are particularly hardy in their capacity to withstand adverse conditions.

Summary

An instance of adamantinoma of the jaw with metastases to the lung is reported. The bronchi of the lower lobe were markedly dilated and their lumina filled with a cast of the tumor tissue. In places the parenchyma of the lung was invaded. It is suggested that the tumor tissue was aspirated into the lung from the primary tumor, via the trachea and bronchial tree, and grew primarily within the lumina of the bronchi. No similar metastatic lesion of an adamantinoma in the lung was found reported in the literature.

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DESCRIPTION OF PLATES

PLATE 81

FIG. 1. Tumor cast in a bronchus. × 30.

FIG. 2. Section through tumor cast showing the histological appearance of adamantinoma. × 120.

1

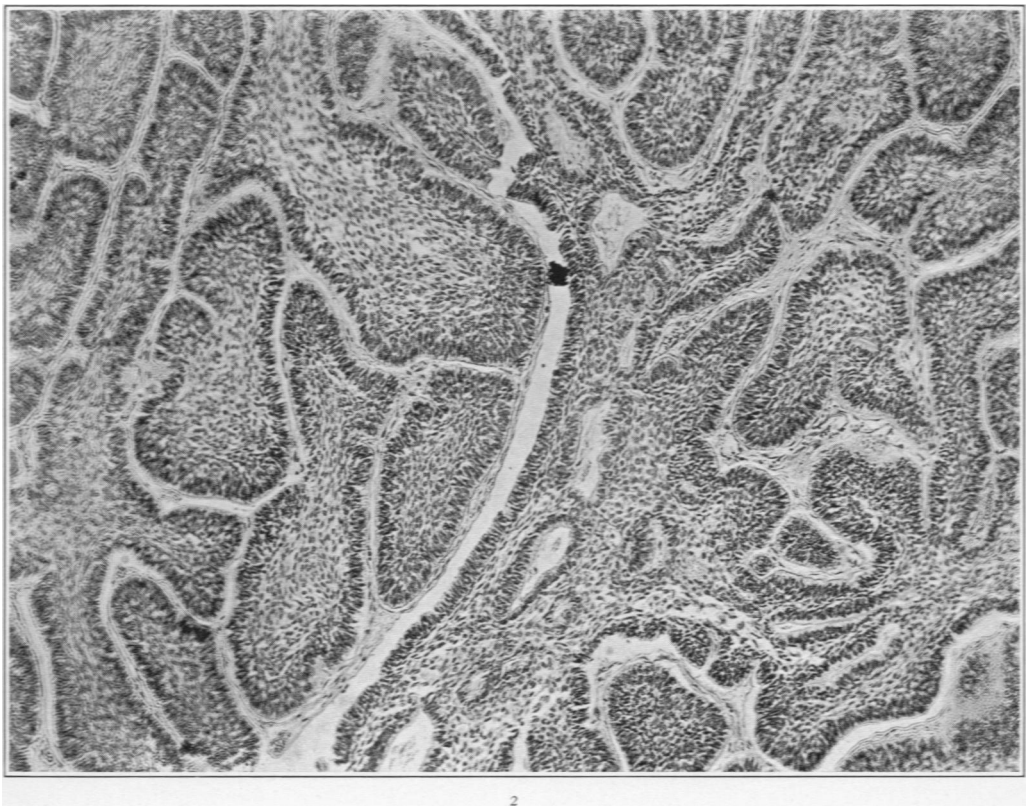

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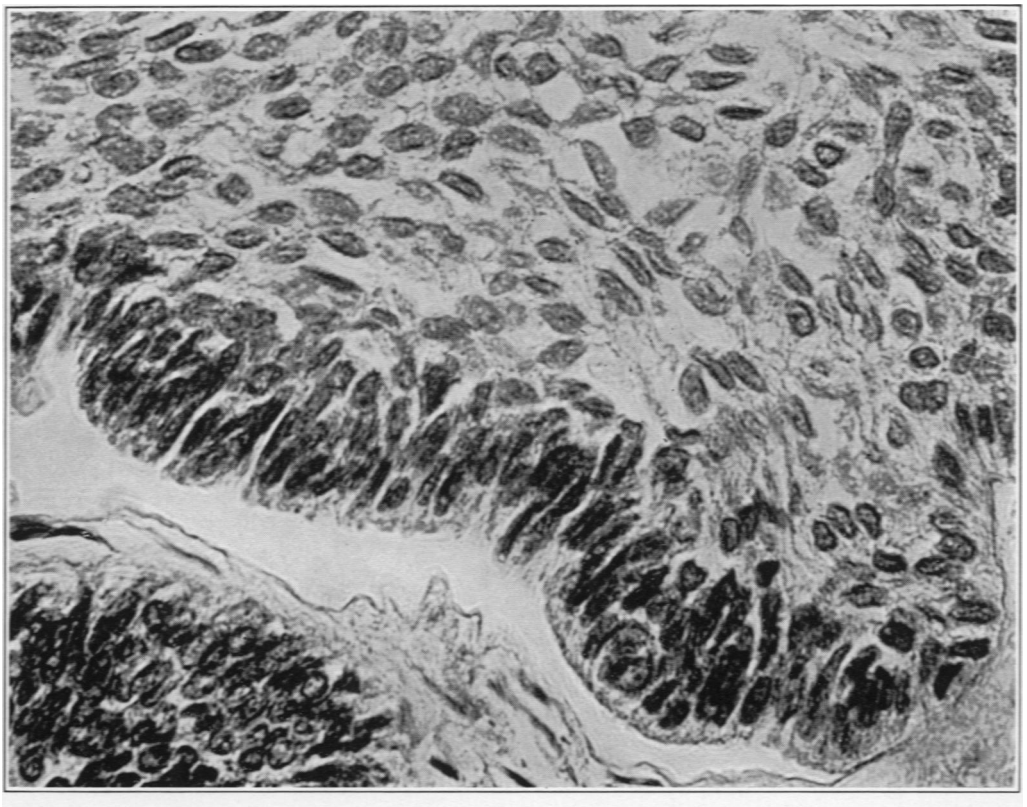
PLATE 82

FIG. 3. Same as Fig. 2. Higher magnification. × 440.

FIG. 4. Adamantinomatous tissue invading the cartilage of a bronchus.

FIG. 5. Nodule in left lung showing hyaline degeneration with connective tissue capsule. × 30.



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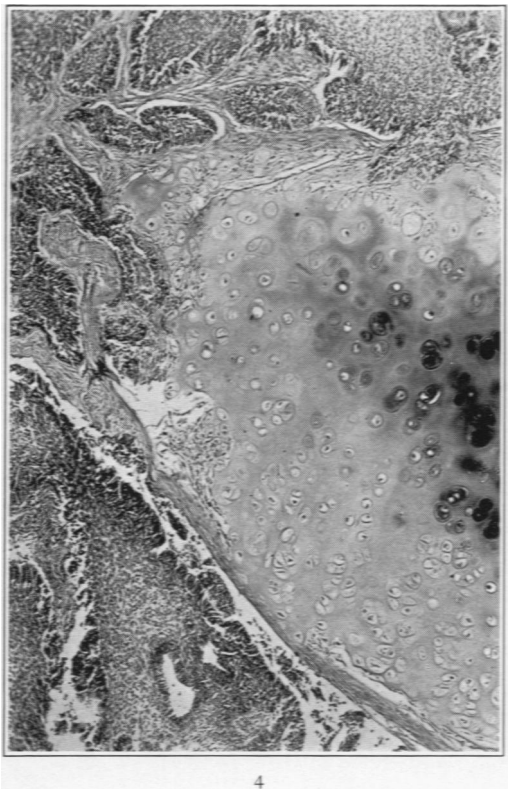

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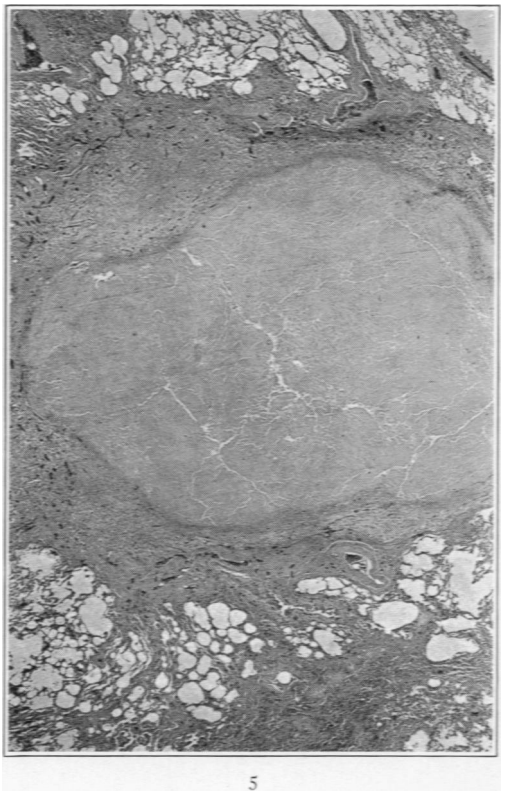

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Adamantinoma of Jaw with Metastases to Lung