A SPECIAL FORM OF CHONDROMA OF THE LUNG

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Tumors composed of cartilage occur in the substance of the lung but are infrequent. They are much rarer than the chondromata of the bronchi and still less frequent than malignant tumors of the bronchi or the lungs. A number of cases have been reported, all in adults.

The type of tumor about to be described is of a somewhat different nature. It is characterized by the presence of epithelial in addition to cartilaginous elements. It was not possible to find analogous tumors described in the literature—with perhaps the exception of a case reported by Hart as follows:

Age of patient 67 years. Cause of death, diabetes. At the periphery of the lower right lobe there was found a lobulated tumor. Microscopic examination showed, besides cartilage, connective tissue in part myxomatous, and gland formation. There is central calcification. Long tubules with many lateral branches are frequent. They are lined with single or multiple layers of low cylindrical cells. These tubules apparently pass through the ground substance in all directions. In addition there are many strands of cells arranged in single and double rows. Diverticula, sac-like in part, filled with desquamated epithelium and occasionally forming cysts, are to be seen. Minute gland-like structures are present, often lying in fat tissue, with a narrow lumen lined with high cylindrical epithelium.

We shall now discuss our own cases which by their uniformity seem to permit the conclusion that they are an unusual form of chondromatous neoplasm of the lung. For microscopic study the tumors were fixed in formalin and imbedded in paraffin. Sections were stained with hemalum and eosin, van Gieson’s, Weigert’s elastic stain, and muchematin.
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CASE 1

Autopsy Record

The patient was 51 years of age. The clinical diagnosis was auto-intoxication, delirium tremens. The anatomical diagnosis was atrophic cirrhosis of the liver; chronic passive congestion of the spleen; edema of the lungs; chronic catarrh of the stomach and intestines; pyelonephrosis; hemorrhagic and purulent cystitis; aspiration lobular pneumonia in lower right lobe; chondroma of lung.

Macroscopic Description of the Tumor

The chief tumor was situated about three millimeters below the lung surface, is approximately two cm. in diameter and of chondromatous appearance and consistency. It shows an indistinct lobulation, the lobules varying in size. A few smaller tumors of the same appearance lie in the neighborhood, a little deeper in the lung parenchyma.

Microscopic Description

The tumor shows microscopically a more pronounced lobulation than in the gross, so that the single roundish units are frequently connected, if at all, by thin bridges of chondromatous tissue. The units are separated by a vascularized stroma containing abundant epithelial elements.

The largest masses show regressive metamorphosis in their centers. This degeneration is in the form of areas in which the ground substance takes no color and in which are to be seen only a few fibrillar structures and cartilage cells in varying number and in irregular forms with indistinct nuclear staining. These areas give the impression of some sort of hydropic change or even liquefaction. In other places similar areas instead of appearing unstained show a pale tint from the hemalum stain.

The cartilage cells, comparatively few in number, are scattered through the ground substance more or less uniformly. In a few places only, and as a rule near the periphery of the single units, do they form little groups, which might be interpreted as centers of more rapid growth.
In several places the periphery of the tumor shows a gradual transition to connective tissue, staining intensely pink with eosin. The fibers of this connective tissue appear first singly and then in bundles arranged parallel to the periphery and interrupting the continuity of the ground substance. Finally they form a continuous mass resembling a true perichondrium. The cartilage cells in these peripheral areas may preserve their type even in the neighborhood of this perichondrium, but a part of them gradually lose the type so that if the transitions were not visible the latter would scarcely be recognizable as former cartilage cells.

In general the cartilage cells are contained in comparatively large cavities or capsules, which often contain two or three cells. The nucleus is very poor in chromatin and possesses a very small amount of protoplasm. The chondromucin fills the space between cell and capsule and stains bluish with hemalum. It frequently shows a sort of vacuolar structure. In many capsules only chondromucin is seen, no cartilage cells being visible. The chondromucin takes the mucin stain intensely, the ground substance in a very weak and quite diffuse way, the areas of regressive metamorphosis not at all.

With Weigert's elastic stain fibres are found to be abundant in the ground substance, especially in the previously mentioned areas where the cartilage cells form little groups. In the peripheral areas they distinctly show a radial course, but in the outermost peripheral parts their course runs parallel to the periphery. In the central part they form a network whose meshes vary in size. The intensity with which they take the elastic stain shows great variation; in this case it seems that degenerative changes also affect the fibres.

The interstitial areas are of interest because of the presence of the epithelial structures, these areas appearing in the section in the form of long, thin tongue-like projections or septa. They contain, in addition to the epithelial structures, connective tissue in comparatively small amount, and also blood vessels. In some places the connective tissue is so scarce that these septa seem to be formed chiefly by the epithelial structures.
The epithelial structures appear chiefly as longitudinal axial sections of ducts with a lumen of more or less constant width and lining epithelium on both sides. These duct-like double rows of epithelium are to be seen frequently extending all along the spaces separating the chondromatous units. In places where these septa contain the least amount of connective tissue the two rows of epithelium stand in so intimate connection with the surface of the two adjacent cartilaginous units that the row of epithelium on the right side of the lumen simply covers the surface of the chondromatous unit to the right, the epithelial layer on the left side covering the surface of the one to the left. In no place between the cartilaginous units are round cross sections of these ducts to be seen. It would be quite impossible that in all the sections the knife could have hit those epithelial structures axially without exception, if they were ducts. Therefore these structures can not be ducts, but must be flat sacs or pockets with complicated ramifications. In fact we have here a conglomeration of closely packed chondromatous bodies, the surface of each covered by a thin epithelial layer. Hence the facets of contact must show double rows of epithelium in whatever way the plane of the cut passes the cleft between two surfaces of contact.

A peculiar picture is seen at the outer surface of the tumor where it is in contact with the surrounding lung tissue. In some places a thin layer, mostly infiltrated with lymphoid and polymuclear cells, separates the whole chondromatous tumor from the surrounding lung parenchyma; but in many places this separation is maintained by the same epithelial structures visible in the interior of the tumor. At the periphery the inner epithelial lamella covers the surface of the tumor, the outer lamella covers the connective tissue of the lung parenchyma and may frequently be identified with the wall of the adjacent extended and compressed lung alveolus. This outer epithelial lamella often becomes defective so that a cleft separating tumor and lung parenchyma may be lined on its inner side with regular epithelium; but on its outer side no epithelium is present (See Fig. 1). In these places little branching ducts are
visible in oblique and cross section, lined with the same epithelium that lines the sacs and pockets.

Fig. 1. Showing cleft separating tumor and lung parenchyma, the tumor bearing epithelium. On the side of the cleft bordering lung parenchyma epithelium is absent.

This epithelium is cylindrical and in a single layer, except where in a somewhat atrophic condition its form approaches that
of a cubical epithelium; but its true nature is to be seen only in areas near the center of the tumor where it appears in form of a typical ciliated epithelium.

The lumina of these epithelial structures are as a rule empty, but in a few places they contain a thickened product of secretion which appears in form of sharply outlined glistening masses taking the eosin intensely, the staining resembling somewhat that of colloid. The true nature of the secretion is demonstrated by the mucin stain. In some places a thin thread-like layer of mucin covering the surface epithelium or even filling the lumen of the narrow pockets is to be seen, especially in the more central parts of the tumor.

A transition between this epithelium and the epithelial structure of the lung is nowhere demonstrable. And nowhere in the lung parenchyma are epithelial structures found that might be brought into relation with the tumor epithelium, except in one place at some distance from the tumor, where a small focus of conglomerated tubercles is visible. Some of these tubercles are caseated and between them fibrous changes in the lung parenchyma occur, in which are to be seen small groups of the gland-like structures which form so frequently in all sorts of lung fibroses as proliferating remnants of bronchioles and alveoli. But this epithelium shows a marked difference from that of the epithelial structures of the tumor. It occurs only in cubical forms, the nuclei nearly filling the cells and taking the stain comparatively poorly. The epithelium in the tumor shows the characteristic cylindrical or ciliated type with oval nuclei staining intensely with hemalum. This tuberculous area is separated from the tumor surface by a broad layer of unchanged lung tissue.

CASE 2

Autopsy Record

The patient was 75 years of age. The anatomical diagnosis was healed dysentery of the rectum; marasmus; spontaneous fracture of neck of right femur without tendency to heal (fracture occurred seven months before death); cartilaginous
tumor of spherical form, one and one half centimeters in diameter, situated in lower lobe of left lung near pleura next to the hilum and extending into the depth of the lung parenchyma. Chronic diffuse bronchitis; hemorrhagic gastritis.

**Macroscopic Description**

The peripheral surface of the tumor lies adjacent to the pleura. It is whitish in color. On the whole the tumor represents a single unit but is divided by deep fissures containing vessels into single areas. Small areas of calcification are present.

**Microscopic Description**

The lobulation of the tumor is marked. Sometimes a layer of connective tissue consisting of a few layers of broad fibres, showing hyaline change in some places, may separate the cartilaginous islands. This connective tissue may also extend over greater or small parts of the surface of the single units as a perichondrium.

The chief cause of the marked separation of single units is to be seen in all clefts, with a distinct, but at times narrow, open space. These spaces are lined by a single layer of epithelium which often shows ciliated forms (See Fig. 2). In compressed areas flat forms occur, and between these two extremes may be seen cubical and cylindrical types. In some places the two opposite layers of flattened cells have completely united as the result of compression of the cleft by the growth of the two corresponding islands of cartilage.

The cleft form is the same as that seen in the previous case. In only a few areas at the periphery of the tumor, especially in the angles between two adjacent units, more complicated forms of proliferation occur, and, by the form of their cross and oblique cuts, show slightly ramified duct-like structures.

In a number of places the covering epithelium and the surface of some of the cartilaginous units do not stand in immediate contact, but a thin layer of fat tissue, often containing numerous vessels, is interposed in varying thicknesses (See Fig. 2). The cartilage in these chondromatous units shows no great tendency
to regressive metamorphosis. Only in a few places areas of degeneration are to be seen together with absence of cartilage
cells and a fibrillar change of the ground substance. Very small areas of calcification are present.

Another form of degenerative change appears in larger areas containing no cartilage cells. In such areas long thin cells of the
connective tissue type occur in great numbers and between them
the remnants of ground substance show in form of a network,
formed apparently out of the ground substance as a result of the
appearance of numerous holes or defects. These vacuole-like
areas are neither stained by hemalum and cosin nor by the
mucin stain, although the latter stains the network itself deeply;
in some places at the periphery of such areas the denser grouping
of the spindle-shaped cells and their radial position make it
possible that the degenerative change of cartilage first took
place and then the young elements entered. The ground
substance of the cartilage contains no elastic fibres.

CASE 3

Autopsy Record

The age of the patient was 53 years. The clinical diagnosis
was chronic fibrous myocarditis with sclerosis of coronary
arteries. The anatomical diagnosis was aneurism 10 cm. in
diameter in the anterior wall of left ventricle near apex, with
marked thinning of that part of heart wall showing marked
fibrosis; arteriosclerosis with partial stenosis of the anterior
descending branch of the left coronary artery; marked thickening
of intima at point of origin of vessels arising from descending
aorta. The arteries of the extremities were free from sclerotic
change; recent thrombosis of the portal vein; infarcts of liver;
general passive congestion; supernumerary spleen; a hard
subpleural tumor in lower lobe of left lung, the tumor being
about three centimeters in diameter and situated in the anterior
inferior area of the mediastinal surface of the lobe.

Macroscopic Description

The tumor, which is spherical in form, is separated from the
pleura by a thin layer of lung parenchyma. Its color is light
blue and the mass is translucent. The tumor is composed of a
conglomeration of roundish units separated either by vascular-
ized connective tissue or in a few places by quite thin layers of
compressed lung tissue. The single units vary in size, the largest
being three to four millimeters. The tumor is quite sharply
outlined against the adjacent lung tissue, which shows no pathological change.

**Microscopic Description**

Microscopically the tumor shows clearly the conglomeration of single units, the smallest of which has a diameter of two millimeters. Some of these units are connected by thinner or broader bands of cartilaginous tissue. The smaller units show a regular roundish form of hyaline cartilage.

The cartilage cells are distributed in a fairly regular way through the ground substance and the quantitative relation between cartilage cells and ground substance is about that seen in epiphyseal cartilage. The cartilage cells do not differ much from the common type. The cavities in which they lie average a little wider than normal. In some areas calcium deposits occur along the inner surface of these cavities as a result of which the cavity appears surrounded by a bluish ring (See Fig. 3). These changes occur in circumscribed areas in which the cartilage cells are diminished in number and stain poorly, thus indicating an area of degeneration.

The microscopical picture shows that the interstitial spaces between the cartilage units do not contain lung tissue as assumed from the macroscopical aspect. These spaces contain either a cellular connective tissue, of a more or less edematous type, and frequently fat tissue. As a rule the interstitial spaces show a comparatively rich vascularization.

In this interstitial tissue numerous duct-like structures occur. These ducts are either long and narrow or short, roundish and dilated to cyst-like forms by a liquid that contains some mucoid substance. The mucin stain shows a few shreds of mucin, especially along the surface of the epithelium. The epithelium is cylindrical or more cuboid and occurs in a single layer. Ciliated types were not found. Weigert's elastic stain shows fine fibres, sometimes in loose bundles all through the cartilaginous ground substance. The interstitial tissue shows elastic elements, chiefly related to the periphery of vessels and in the wall of the larger vessels. The lung tissue surrounding
the tumor area shows nothing worth mentioning. The outline of the tumor against the lung parenchyma is very sharp. The van Gieson's stain shows distinctly a continuous thin layer of connective tissue comparable to a capsule extending all around the tumor area and bearing no relationship with the immediate adjacent lung parenchyma.

Fig. 3. Showing calcium deposits along inner surface of cartilage cell cavities.
CASE 4

Autopsy Record

The age of the patient was 41 years. The clinical diagnosis was embolus of left femoral artery; dry gangrene of left leg.

The anatomical diagnosis was mitral stenosis and mitral insufficiency; slight insufficiency of the aortic valve following healed endocarditis of the mitral valve; complete thrombosis of left auricular appendix; slight excentric hypertrophy of both ventricles; diffuse fibrinous, purulent peritonitis following gangrene of intestinal loop due to embolus of superior mesenteric artery; numerous fresh and old infarcts of both kidneys; complete embolic occlusion of left renal artery; dry gangrene of left foot and leg following embolus of femoral artery; beginning gangrene of thigh due to embolus; chondroma in lower lobe of left lung.

Macroscopic Description

The tumor was about 2 cm. in diameter, of distinct cartilaginous appearance and consistency. In some places fissures are to be seen which contain small blood vessels. The contour of the tumor is of irregular form. In the central part is a cavity about nine millimeters in its greatest diameter. The inner surface of this cavity is smooth, except where remnants of tissue in form of threads and fine membranes are seen projecting into the lumen. The surface of the tumor is separated from the pleura by a thin membrane.

Microscopic Description

The chondroma is made up of hyaline cartilage. The ground substance takes a light blue stain with hemalum, this blue changing to pink (eosin) in the periphery of the single units composing the tumor. The cartilage cell cavities (capsules) vary somewhat in size without attaining excessive size. An arrangement in pairs is not infrequent, but they sometimes occur, lying close together, in small groups. The cartilage cells mostly appear shrunken and deformed, a great or the greater part of the cell cavity being filled with chondromucin which
appears, as a rule, in form of a fine network staining bluish with hemalum and dark violet with muchematin.

Frequent small areas of the ground substance are free of cellular elements and take a transparent pale stain, with a rudimentary sort of fibrillar structure which is also pink, indicating retrograde metamorphosis. The difference between these areas and the central cavity mentioned in the macroscopic description seems to be only one of quantity. The borders of this cavity show more extensive changes in the ground substance identical with those just mentioned. The cavity itself is the result of liquefaction. These small areas of degeneration show especially well in sections stained for mucin, the muchematin staining the ground substance in a very characteristic way. Next to the cell capsule is a concentric zone nearly unstained by muchematin, then follows a pale homogenous violet stain of the ground substance in fine streaks or reticular structures and a shade darker, between two adjacent cell cavities.

The periphery of the single units is marked by a pink marginal stain. In some places the outline becomes especially sharp, due to the fact that adjacent units are separated by strands of connective tissue containing blood vessels. Elastic stain shows the absence of elastic fibres in the cartilage itself. These are to be found only at the periphery and occasionally in the interlobular areas. The outer surface of the tumor does not show a true perichondrial covering.

The tumor shows at its periphery a contour broken by numerous sharp angles projecting into the tumor mass, each angle forming an area between two units. In these angles, besides connective tissue and vessels, numerous little ducts with narrow, sometimes invisible, lumina are to be seen lined with a cubical epithelium in a single layer. The ducts give the impression of atrophying structures (See Fig. 4). Only in a few places is a transition of these ducts to lung alveoli visible. These structures seem to be due to the growth of the tumor mass, especially between tumor surface and pleura. The majority have undergone the change which is so frequently seen in fibrous lung changes in which remnants of alveolar epithelium
undergo the alteration to cubical epithelium, forming duct-like structures. These distinctly atrophic structures are not to be mistaken for the characteristic tubular structures of a proliferating type so characteristic of the adenochondromata.

Fig. 4. Showing connective tissue, vessels, and small ducts lined with cubical epithelium lying between two cartilaginous units.
SUMMARY

It is easy to recognize that the first three of these four cases are of identical nature. The fourth, although having quite a number of points in common, differs in one detail,—that the epithelial layers between the cartilaginous units are lacking.

However, the common features are so striking that it must be taken into consideration whether this fourth tumor is not of the same nature as the other three, but as having lost its epithelial component in the course of its development. The whole literature of chondromata of the lung does not mention tumors of a similar conglomerated type (with the exception of Hart’s case in which the tumor was in the right lower lobe) and this seems to be a point in favor of the hypothesis explaining this fourth tumor as analogous to the other three, only differing in the non-development of the epithelial part.

Among other important characteristics in common is the localization. In three out of four cases (unfortunately the localization of one is not stated in the record) the site of the tumor is the lower left lobe, and in the two cases where a more exact localization is given in the record, the site near and below the hilum is mentioned. All show the sub-pleural localization and all are sharply outlined by connective tissue, which in three cases shows such an intimate connection with the surface of the cartilage as to resemble perichondrium.

The single cartilaginous units are separated by connective tissue and not by lung parenchyma. The presence of fat tissue in the intracartilaginous clefts in all four cases is remarkable.

The tumors caused no functional disturbance and therefore all were accidental findings.

It scarcely needs to be discussed that these tumors did not form by neoplastic proliferation of physiological elements, that is, from cartilaginous structures in the wall of the bronchi, in post-embryonal life. In fact the tumors have no connection with physiological structures, neither in their cartilaginous nor in their epithelial part, but give the impression that they arise from aberrant structures.

The uniformity of localization raises the possibility that the
material out of which these tumors are formed corresponds to a structure which disappeared in phylogenesis, especially of something that from the standpoint of human anatomy would have to be classified as a supernumerary bronchus, and it would be an interesting investigation to try to find this bronchus by a study of the comparative anatomy of the lung in lower animals.

**NOMENCLATURE AND CLASSIFICATION**

A new type of tumor like this warrants a name. Considering the two most important components of the tumor, adenochondroma is justified. This would harmonize with adenomyoma uteri.

Of importance is the classification of the tumor. In the old nomenclature it would be a mixed tumor in the sense in which Virchow used the term, but modern views would not be satisfied with this. According to E. Albrecht's terminology it seems that the tumor would deserve the title "hamartoma" on new growths which are to be explained as tumor-like malformations formed by an abnormal mixture of the constituent parts of the organ in which they originate.

Perhaps one detail of structure deserves special mention, i.e., the presence of fat tissue. A recent article by A. Feller discusses the presence of fat in a lung tumor and refers to Hart who quotes Chiari as explaining that fat may develop from the small amount which is sometimes present in the submucosa of the large bronchi.

We believe that this might explain the presence of fat tissue, especially in tumors lying near the hilum and originating from a supernumerary bronchus. Such a bronchus developing from the trachea in a direction from the mediastinum to the lateral parts may carry with it elements from the mediastinal interstitial tissue destined to become fat tissue. The same occurrence might be an accidental finding—fat as an accompanying tissue in the other structures, such as nerves and vessels developing in the same direction, passing the hilum and entering the lung area, especially under circumstances of disturbed development.

In conclusion I wish to express my thanks to Professor
C. B. McGLUMPHY

Stoerk for the opportunity of reporting these cases and for the invaluable aid given me in this work.

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