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ORIGINAL COMMUNICATIONS.

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RHABDOMYOMA OF THE NOSE.

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Rhabdomyoma (Myoma Striocardiale, Rhabdomyoma Sarcomatodes, or Rhabdosarcoma) has been observed in the various organs of the body, most frequently in the kidney, uterus, rectum, stomach, testicle, once in the parotid gland and once on the root of the nose between the eyebrows. A single case like the following becomes interesting only on account of its extreme rarity, and, believing that all unique cases should go on record, I beg to report it. So far as I can ascertain it is the first case of its kind on record.

The patient, Mrs. L., first consulted me regarding the present trouble May 7th, 1906. She gave the following history:

Six weeks ago had the "grip," affecting the respiration apparatus and accompanied by considerable stoppage of the nose with twinges of pain in the ears. Was treated by her family physician, who, meeting with little success in relieving her of her nasal symptoms and recognizing that a change of climate would be beneficial in a general way at least, advised her to go to Atlantic City. She followed his advice, staying two or three weeks, and returned feeling much better as regards her general condition, but still complaining of nasal stenosis on the left side. She called to see me regarding the symptom, which was not painful, but annoying.

Status Præcens: Patient a spare but well nourished lady of fifty-eight years of age, presenting no external evidences of ill health and giving no symptoms other than complete stoppage in the left nares.

*Read at the 14th annual meeting of the American Laryngological, Rhinological and Otological Society, held at Pittsburg, May 28-30, 1908.

Examination: Right naris normal; left revealed no breathing space whatever, the turbinal body being in tight contact with the septum, and the attempt to introduce pledgets of cotton saturated with cocaine and adrenalin solution met with failure. The stenosis began just within the vestibule of the nose and seemed to be entirely due to a swollen inferior turbinate. The middle fossa could not be seen. Post-rhinoscopic examination revealed the choana of the left side to be completely filled with a mass of pale tissue, light gray in color, which felt like a nodulated mass of tough tallow. The transillumination test was negative, the hearing unimpaired and the eye normal in all respects. My associate, Dr. Lamb, examined the case with me and we agreed that it "looked suspiciously like a sarcoma," although it lacked the reddish color of the sarcomata and did not bleed when wounded. We advised removal of the growth and gave a guarded prognosis.

The Operation: On June 4th, 1906, I attempted to cocaine the left naris and made more or less futile attempts at getting solution of adrenalin within the nasal cavity, but very little, if any, could be crowded along the septum on account of the impacted mass of tissue. Scissors could not be passed without puncturing and tearing the specimen. A fine straight nasal saw was passed under the growth along the floor of the nose and an attempt was made to engage the inferior turbinate in the cutting stroke. Meeting with no resistance that felt like anything, the saw was withdrawn. A nasal snare loaded with a large loop of No. 4 piano wire was insinuated along the septum until it passed the growth and the wireloop could be felt in the pharyngeal vault with the finger. The tip of the index finger of the left hand was passed through the loop of wire in the pharyngeal vault and made to pass up over the summit of the growth, and then by drawing the wire home, it slid along the finger and thus entire tumor mass was engaged. I expected to meet with considerable resistance by this large mass of tissue engaged in the wire-loop, but on drawing the wire it was delivered in the canula of the instrument without any apparent resistance. On attempting to extract the mass, I encountered considerable difficulty. I could not extract it forwards; it was too large and any attempt to grasp the front end of the mass met with utter failure for the reason that the tissue was so friable that it could not be grasped. I succeeded finally in getting it out piecemeal by the various means at

my command, and extracted a wineglassful of fragmentary masses of the tumor. There was no bleeding and inspection seemed to show a thorough removal.

The specimen was hardened and sectioned by Dr. Lamb and was thought to be a large spindle-cell sarcoma by those who examined it. This information was given to her brother, who desired the specimen sent to any expert pathologist I might choose with a view of getting a positive diagnosis. Two pieces of the growth were sent to Dr. Wm. H. Welch, of Johns Hopkins University, Baltimore, and I beg to read his report in full.

Examination of Tumor from Nasal Cavity, for Dr. D. T. Vail.

The specimen is a solid grayish white elongated mass of tissue, about $1\frac{1}{2}$ ctm. long and $\frac{5}{8}$ mm. broad, and of irregular shape. A part of the free surface is smooth as if covered with mucous membrane. The cut surface appears smooth and uniform. The tissue is preserved in formalin. There are two parts of the growth in the bottle, from each of which sections were cut. *Microscopical sections* show over a small area remnants of the epithelial covering in the shape of cubical cells, but over most of the surface the epithelial cells are absent. The rest of the section is composed entirely of neoplasm.

The tumor consists essentially of interlacing bundles of fibers and cells, which appear cut in different directions. The characteristic and predominant cells are elongated, band-like and fusiform cells, but large, round, oval, irregular and branching cells also occur. There are also cells with large, irregular, budding nuclei and with multiple nuclei. All of these cells probably belong to the same group and are embryonic, striated-muscle cells in various stages of differentiation or entirely undifferentiated.

The most highly differentiated cells are long, ribbon-shaped cells with one, rarely two or more, vesicular nuclei and with bodies which stain deeply with eosin. These bodies present often distinct longitudinal, delicate fibrillation, and less frequently, but in some cases distinctly, cross striation, especially at the margins of the cells. The ends of the cells appear continuous with delicate fibrils which can be traced into the surrounding tissue, either as separate fibrils or as a network of anastomosing or interlacing fibrils. These cells, which can be positively recognized as embryonic striped muscle, occur in moderate numbers scattered throughout the growth, but are most numerous in a small area near the margin of the section.

By the identification of the foregoing cells as muscular, the interpretation of other cells of the tumor as less differentiated muscular cells becomes easier. Thus there are long, flat cells with longitudinal but without transverse striation, these cells being much more numerous than those with cross striation; similar cells with smooth or granular undifferentiated cytoplasm, which, as in the foregoing cells, stain deeply with eosin; large and small, round or irregular cells with similar cytoplasm, and branching or even spider cells with delicate processes continuous with a reticulated stroma. The margins of many of the cells described appear as a glistening, single or double contoured, refractive line.

These various cells are arranged for the most part parallel to each other in interlacing bundles. When such a bundle is cut across, it sometimes happens that the contents of the cells drop out, leaving the refractive borders, so that a peculiar reticulated or honeycombed structure results, or it is possible that this appearance may be due to dissolved glycogen. There appears, however, also to be in some places a genuinely reticulated stroma, and it can sometimes be seen that fibrillar processes from the cells are continuous with this reticulum. Exceptionally on cross sections of the muscle-cells, Cohnheim's fields can be made out.

There are many elongated, round or branching cells which are without any suggestion of a muscular origin. Whether these should be interpreted as connective-tissue cells or entirely undifferentiated embryonic muscle-cells, is a matter of arbitrary judgment, but I am inclined to the latter interpretation, as all transitions between these and recognizable muscle-cells exist.

The giant cells are doubtless of muscular origin, although this too cannot always be demonstrated.

The tumor as a whole is fairly rich in cells, although in places the large amount of cytoplasm and fibrils makes the nuclei appear far apart. There are a few irregular areas of coagulative necrosis. Larger blood-vessels are not seen, but there are several wide lymphatic vessels. Genuine connective tissue is not abundant.

Diagnosis: Rhabdomyoma Sarcomatodes.

Remarks: Rhabdomyomata as rich as the present tumor in undifferentiated cells, resembling sarcoma cells, are usually designated Rhabdomyomosarcomata, but I prefer the above designation, as it seems to me probable that these undifferentiated cells are of muscular rather than connective-tissue "Anlage."

These tumors undoubtedly spring from misplaced, embryonic germs of striped muscle and are often developed at birth or in early life, although they may appear later. They are most common in the kidney, next in other parts of the genito-urinary tract, but they have been observed in various other parts of the body. In Benenati's collection from the literature of sixty-four cases (*Virchow's Archiv.* 1903, Bd. 171, p. 418) there is none within the nasal cavity, and I am not aware of any instance of this localization previously observed.

The majority of reported rhabdomyomata have not been malignant in the sense of metastasizing or of invasive, infiltrating type of growth. They may, however, be malignant and metastasize, especially when the growth is rich in sarcoma-like cells. I have examined specimens from two such cases of malignant rhabdomyoma, one being of the vagina and the other of the bladder.

WILLIAM H. WELCH.

There was no history of syphilis, but the patient was put on K. I. and Hg. and remedies were pushed to the point of saturation without any result.

In the meantime, the nasal cavity began to fill again and the transillumination test showed left side of the face dark. Another operation similar to the first was advised, as it was seen that the growth involved the upper turbinal wall. This was consented to and on October 15th the lateral wall of the inferior and middle meatus of the nose was rid of the same kind of growth as was found at the first sitting, and the middle turbinal body was entirely removed. It was observed at the time of the first operation that the lower turbinated bone was entirely absent—digested—absorbed—transformed or rarefied; at any rate, there was no bony framework of the inferior turbinated present. At the second operation the conformation of the middle turbinated was plainly made out, but on looking at the specimen, you will see that it, too, was rapidly losing its bony framework, being transformed into this tallow-like mass of nodulated tissue, which you can tear apart in shreds like a mass of soft papier-mache. The cavity was as thoroughly cleaned out at this second operation as possible. There was no bleeding at any time, either at the time of operation or afterward.

Soon after there was evidence of recurrence and I called a consultation with Dr. C. R. Holmes, who agreed that the growth

was of unusual character, was probably filling the antrum and that a radical operation should be performed. There was no facial deformity and no subjective symptoms aside from complete blocking of the nose on that side. As the patient naturally dreaded the heroic operation proposed, it was decided to send her to New York for further advice, and Dr. Emil Mayer and Dr. R. C. Myles were consulted. I will submit extracts from their letters.

Extract from Dr. Mayer's letter:

Mrs. L. called to see me on February 1st, and I have since seen her daily for three days for the purpose of making a full report to you.

With your history to go by, the growth in her left naris, which practically occludes that side, is readily recognized as a neoplasm. It does not shrink under cocaine, nor adrenalin, is hard and bleeds readily. Nevertheless it seems to be made out to be limited to the site of the turbinate only. While it may be encroaching on the ethmoid cells and antrum, examination fails to show that. The transillumination test carefully made shows no difference on either side.

With the positive diagnosis of neoplasm, it seemed best to me to see if there was any change from the conditions found by Dr. Welch last June, and consequently I removed a small piece of the growth and referred it to the pathologist at Mt. Sinai Hospital. I enclose his report, and you will see he agrees with Dr. Welch in his report, although he did not see the striae that made it a rhabdomyoma. There are good reasons for this. You sent a large specimen, I sent a small one; yours was of the original growth, while mine was of the recurrence.

The main point is to decide if in the past six months there have been any changes in the growth indicating greater malignancy. This may now be definitely negated.

The diagnosis is a sarcoma of rather benign tendency and we have therefore every reason to believe that its thorough removal will effect a cure.

I have given careful thought to your query as to treatment. Except for a history of several miscarriages there is no reason to suspect syphilis, and I believe you have given her full doses of KI with negative results; so I eliminate that. I have considered the possible value of X-rays and I greatly doubt their efficiency.

It seems to me therefore after careful examination, that her best interests will be in an operation which will remove the

growth and the adjacent affected parts, not necessarily all of the half of the maxillary bone.

Mrs. L. complained of pain or pressure over chest and radiating into the arm. I found no chest abnormality and no glandular enlargement.

I have therefore only to verify in every way what you have done, to agree with you in the treatment heretofore, and finally to thank you for the opportunity to see so interesting and unusual a case. Very sincerely yours, EMIL MAYER.

The pathologist referred to in Dr. Mayer's letter was F. S. Mandlebaum. He made the following report:

The tumor from the inferior turbinate sent for examination is a small, hard, oval-shaped nodule, measuring from 6-8 mm. in diameter, without any special distinguishing features on the surface. On cutting it in half, its surface is greyish in color and quite homogeneous in structure.

Microscopic Examination: When viewed with the low power, the entire central portion appears more or less degenerated and stains quite poorly as compared with the peripheral parts. With the higher power the central portion shows that the degenerative process has ceased before complete necrosis, and the component structures are still faintly visible. The peripheral zone shows distinctly the character of the growth, which extends quite to the surface and is not covered by mucosa.

The growth is composed almost entirely of well-defined cells with more or less parallel margins and of variable lengths. The ends of these cells are frequently drawn out to a thin filament, and often merge into the connective tissue stroma. These cells stain readily with eosin and with picric acid. No striations can be seen. The nuclei of the cells are vesicular and very rich in chromatin, and vary greatly in size and shape, but for the most part are rather broad and oval and resemble those of the heart muscle. Here and there scattered between these cells are other large cells of irregular shape, with large nuclei, and occasionally a giant cell with multiple nuclei is found. The cells that comprise the greater part of the tumor are unquestionably muscle cells in various stages of development. Where bundles are cut transversely, the typical appearance of muscle tissue is apparent. In those parts where the muscle fibers are less numerous, a stroma of delicate connective tissue fibers, branching and inter-

lacing, is seen. In the meshes of this stroma a few mononuclear leucocytes are present. No distinctly embryonal connective tissue fibers can be made out. No mitosis, either typical or atypical, is present. The central portion of the tumor, as far as it is possible to make out, consisted of similar structures prior to its degeneration.

Blood vessels are not numerous in the section. Those that are seen are thin-walled and show no lesions.

Diagnosis: Myosarcoma (leiomyosarcoma):

With many thanks for your courtesy, I am

Very sincerely yours, F. S. MANDLEBAUM.

Dr. Myles' report on the case was as follows:

In my opinion Mrs. L. has a malignant growth in the nose. The anterior area could not be properly observed on account of the closeness of the septum to the outer wall and the considerably bruised edema probably due to manipulation. The inferior turbinal can be easily observed through the posterior nares. The posterior half of the inferior turbinal is larger than it should be; its middle and anterior part appears nodular and roughened (by posterior rhinoscopy aided by retraction of the soft palate.) The neoplasm is extending outward, upward and forward toward the nasal process of the superior maxillary bone and the nasal vestibule.

I would advise a radical operation by making an incision beneath the eye down to or through the lip and resection of a part of the superior maxilla. The growth seems to be confined to the anterior half of the nasal fossa. Of course the tissues should be removed a certain distance beyond the apparently healthy lines.

Very sincerely yours, ROBERT C. MYLES.

It was decided to have Dr. Jos. Ransohoff, a general surgeon of Cincinnati, perform the operation, as it was thought the growth may have extended to such territory that the upper jaw might have to be removed together with the entire accessory sinus framework on that side. On February 9th, Dr. Ransohoff examined the case thoroughly and agreed that the external operation (radical removal) was necessary.

On February 12th at the Jewish Hospital the patient was operated on as follows:

Anesthesia by Dr. Iglaue—chloroform. Preliminary laryngotomy and tube inserted; pharynx stuffed with gauze. This was decided expedient on account of the likelihood of dangerous and

annoying hemorrhage and to facilitate the giving of the anesthetic. An incision was carried from the inner angle of the left eye down the side of the nose around the ala nasi and thence outward parallel with the lip. The periosteum was stripped back and the front wall of the antrum together with a part of the nasal bones were removed with the bone forceps. It was then seen that the tumor mass completely filled the antrum of Highmore and was not adherent at its limitations.

The removal of the entire mass was easily accomplished by simply lifting it out of its nest. The confining walls were thoroughly inspected for ingrowths of the tumor, but none were found. The cavity was packed with iodoform gauze and the wound closed by interrupted sutures. The tracheotomy tube was removed and sutures applied, closing the opening in the larynx.

During the entire operation, which occupied only about twenty-five minutes, the patient had a bluish, cyanotic color, her pulse was thready and the heart lacked tone. The patient rallied from the anesthetic so far as the regaining of consciousness was concerned and did not complain of her respiration. Her breathing was deep and normal, but the heart did not rally and in spite of the employment of every known means of restoring the cardiac power, the pulse continued small and thready, and sixty-eight hours after the operation the patient died.

Stengel, in his 1906 book on Pathology, speaks of Rhabdomyoma as "containing more or less striped muscle fibers, usually a small amount, the main body of the tumor being made up of sarcomatous cells.

Etiology: It is thought to be a congenital defective development, and usually occurs in early life and in situations where unstriped muscle fibers do not normally exist. The kidney is the most frequent seat of involvement.

Structure: The microscopic appearance is usually that of a spindle-cell sarcoma, containing more or less striped muscle fibers. There are elongated spindle-shaped cells, partly striated and suggesting embryonal muscle tissue and rarely more fully developed muscle fibers. Large areas may contain no muscle fibers at all, while certain portions are richly supplied.

The tumors are malignant in proportion as the sarcoma elements are predominant, but metastases are infrequent.

I have no doubt that rhabdomyoma of the turbinates has been encountered, but has never before been recognized. The appear-

ance as it exists in the nose is that of a lardaceous or suet-like nodular mass of friable nonvascular tissue. These tumors going to the laboratory for microscopic diagnosis are called large spindle-cell sarcomata.

When one considers that within the nasal structures striped or voluntary muscle fibers do not normally exist, he wonders that such a tumor in this locality could occur. The only plausible theory for its occurrence in a locality of this sort is that one of the neighboring voluntary muscles, such as the levator labii superioris alaquae nasi or depressor anguli nasi, has a prototype embryologic, misplacement lying dormant within the nasal cavity and for some reason took on active sarcomatous growth late in life.

A complete bibliography on the subject is published in Ziegler's "General Pathology," 1903, page 406.

No. 24 East Eighth Street.

An Isolated Primary Carcinoma of the Trachea. RICHARD
HOFFMANN. *Monatsschrift f. Ohrenheilkunde*, August, 1905.]

A man 49 years of age was referred to the author for a diagnosis of an alleged laryngeal growth. No sign of such a growth could be found. Nine months later the patient presented himself again and on the occasion a dark red papillary tumor could be seen situated in the upper part of the trachea. Two days later the growth again disappeared from view, but reappeared again upon a subsequent examination. Operation was refused. About seven months thereafter marked dyspnoea set in, with a pronounced cough. The tumor was now as large as a cherry. Operation was decided upon, and the patient was anaesthetized. As soon as primary anaesthesia was obtained the skin incision was made. The patient immediately became cyanotic and died. The autopsy showed some enlargement of the thyroid, encroaching somewhat upon the trachea posteriorly; a persistent thymus; an open foramen ovale. The tumor, which nearly filled the lumen of the trachea, extended over four tracheal rings and proved to be a primary epithelioma.

YANKAUER.