MALIGNANT RHABDOMYOBLASTOMAS OF THE SKELETAL MUSCULATURE

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The term sarcoma has been used by clinicians to designate a wide variety of malignant tumors, and its use in this broad indefinite sense has caused much confusion. The modern classifications of tumors are based on histogenesis, and attempt to separate the tumors of muscular origin from those arising from connective tissue, nerve sheaths, and other sources.

In studying a group of tumors which have arisen within skeletal muscles it is necessary to adhere to this same principle of histogenetic classification, and to distinguish the tumors arising from muscle cells from those originating in the connective-tissue septa and aponeuroses, and from the sheaths of nerves. There can be no doubt but that striated muscle cells do sometimes give rise to malignant tumors presenting distinctive clinical and histological features, though the group of true rhabdomyoblastomas is a relatively small one.

Seventeen cases of malignant tumor arising within striated muscles were seen at the Oncological Institute in Leningrad during a period of nine years, 1926 to 1934. The specimens taken for histologic examination were fixed in formalin and embedded in paraffin. The sections were stained with hematoxylin and eosin, Heidenhain's iron hematoxylin, Masson's trichrome stain, and Foot's silver stain.

It is difficult to give a general description of the rhabdomyoblastomas because of their extreme polymorphism. They have, nevertheless, certain definite characteristics. Usually the predominant cells are elongated and spindle-shaped (Fig. 1). They are relatively large and have a sharply outlined cytoplasm. Their nuclei are elongated and oval and are rich in chromatin. These cells lie parallel to one another to form intersecting bands of cells, or they may be irregularly grouped together without any apparent pattern. Among these spindle-shaped cells several other types of cells are scattered. These include a small round cell with a dark, compact nucleus and a narrow ring of cytoplasm, and a large oblong or polygonal cell with a pale, vesicular nucleus and a wide layer of cytoplasm (Fig. 2). Giant cells of various sizes and shapes are always present (Fig. 3). Their number varies greatly. In addition, peculiar cells of fantastic shape and inclusions of degenerated muscle tissue are seen in some of the tumors.

The tumor cells are usually closely packed together, but in some places are separated from one another by a delicate collagenous stroma. There are numerous deviations from this typical picture, however. There is, first of all, the polymorphous type of tumor (Fig. 4) in which no one cell type pre-
Fig. 1. Immature Rhabdomyoblastoma: Spindle-shaped Cells

Fig. 2. Mature Rhabdomyoblastoma: Spindle-shaped Cells and Giant Cells Intermingled
dominates. Another form approaches a purely spindle-cell structure. Still another type is composed almost entirely of giant cells. Lastly, there is the tumor in which degenerative processes obscure the structure. Because of these variations in structure it seems futile to try to separate the rhabdomyoblastomas into distinct types. Instead, the features that are common to all may be described.

(a) The Spindle-shaped Cells: As stated above, these are large, elongated cells measuring up to 100 micra or more and possessing a sharply outlined cytoplasm. Although their shape is usually that of a more or less regular spindle, they are sometimes bifurcated at one end into two or more branches, which may extend across more than a single high-power field and may fuse with the branches of adjacent cells. The cytoplasm appears either homogeneous or finely granular; it is distinctly oxyphilic. In some instances it contains delicate fibrils of various kinds (Fig. 5). Sometimes small vacuoles may be observed in the cytoplasm, usually near the periphery. Minute fragments of chromatin and clumps of oxyphil hyaline are also seen in some of these cells.

The nucleus is situated either in the center of the cell or near one of its poles, or it may be placed eccentrically. Sometimes it is so large that the surrounding ring of cytoplasm is hardly visible. Very frequently a cell contains several large oval or bean-shaped nuclei which vary from 15 to 60 micra in diameter. These multiple nuclei may overlap each other, in which case some
of them appear to be smaller and darker. Some of the nuclei show numerous indentations which give them a festooned appearance. The chromatin network is well formed and contains two or three nucleoli. Mitosis is frequently observed.

Although the spindle-shaped cells usually lie parallel and end-to-end, with their cytoplasmic prolongations fused together, they are sometimes transformed into long tubular structures, which form a syncytium with numerous nuclei (Figs. 8 and 20). The nuclei in these syncytia are of two types. One type is large, pale, poor in chromatin, and vesicular, containing several nucleoli. The other is dark, irregular or ovoid in outline, and is stuffed with minute clumps of chromatin. Mitosis is very rarely seen in nuclei of this type. The cytoplasm of these syncytial masses is often delicately fibrillar. Indeed, it is in these syncytial masses that fibrils are most frequently seen in the rhabdomyoblastomas.

The Polymorphous Cells: A great variety of cells may be found in these tumors. Some are small, measuring 7 to 8 micra in diameter, and round, with a compact, well outlined nucleus and a small ring of homogeneous cytoplasm. Others are oval, polygonal, or branching cells from 15 to 30 micra in length. These cells usually have large, pale, single, rounded or oval nuclei, which are poor in chromatin, and a prominent homogeneous or finely granular cytoplasm. Occasionally very large rounded cells with a single pale spherical nucleus and an abundant cytoplasm are seen. All these types of cells are intermingled without any definite grouping. Small areas consisting of large
Fig. 5. Mature Rhabdomyoblastoma: Synctium of Spindle-shaped Cells, and Smooth and Cross-striated Fibrils

Fig. 6. Immature Rhabdomyoblastoma: Giant Plasmodia (Oil Immersion)
rounded cells of approximately equal size, and having nuclei of various sizes and shapes, are sometimes seen; on careful study these appear to be merely the cross-sections of the spindle-shaped cells and the syncytia described above (Fig. 1).

(c) Giant Cells: In all of these tumors giant cells are seen. There is no regularity in their distribution or arrangement. They may be scattered throughout the section, or form small groups, or fill up entire microscopic fields. Although they vary extremely in size and shape, it seems possible to divide them into three types, with many transitional forms. The first type comprises the ordinary tumor giant cells. These are from 30 to 60 micra in diameter, with a single bean-shaped or indented nucleus and a prominent, finely granular cytoplasm (Fig. 4). The second type consists of very large spindle-shaped cells which measure from 150 to 200 micra in length. These cells are not infrequently branched, and often have several fantastically shaped, hyperchromatic nuclei. Their cytoplasm contains delicate fibrils. The third type of giant cell is rare and is really a plasmudium. It is an enormous, sharply outlined, oval or round structure, which covers more than a single high-power microscopic field. It contains ten or more nuclei of varying size and shape. Its cytoplasm is finely granular and may contain delicate fibrils or a network of vacuoles and canals resembling trophospongia (Fig. 6). In one of our cases of recurrent rhabdomyoblastoma the greater part of the
tissue was made up of these plasmodium-like giant cells. The most common of these three types of giant cells is the first; the third type is the most infrequent. In all of our rhabdomyoblastomas we have found giant cells of some sort, however, and we regard them as an indispensable diagnostic feature.

(d) Fibrils: The most important feature of the rhabdomyoblastomas, and at the same time the feature most difficult to interpret, is their fibrils. These are best seen in sections stained with Heidenhain's iron hematoxylin. They are most often found in the syncytial formations and in the long spindle-shaped cells. Here they appear as very long, thin, black fibrils which extend throughout the length of the cell and not infrequently pass into the cytoplasm of the adjacent cell (Figs. 7 and 8). They are most often situated near the periphery of the cytoplasm. Their number varies greatly, although as a rule they are not numerous. The fibrils are either perfectly straight, or slightly curved with twisted ends. Their diameter is not always the same, for they may be thickened in some portions, or may show regularly alternating spindle-shaped or bulb-shaped swellings throughout their length. The ends of the fibrils are either pointed or bent and broken off abruptly.

In single fibrils a definite transverse metamerism was sometimes seen, dark colored segments regularly alternating with light colored segments (Fig. 9). Such cross-striation was seldom seen throughout the whole length of the fibril, and more often appeared only at its ends (Fig. 19). Pronounced cross-striation has been found in only a few instances. When it does occur, it is

Fig. 8. Thick Smooth and Cross-striated Fibrils (Oil Immersion)
Fig. 9. **Mature Rhabdomyoblastoma:** Separate Cross-striated Fibrils; Discs Q and I Visible (Oil Immersion)

Fig. 10. **Immature Rhabdomyoblastoma:** Cross-section Showing Numerous Smooth Fibrils (Oil Immersion)
usually in the syncytial cells, although it may also appear in spindle-shaped cells (Fig. 7).

These fibrils appear as dark colored grains or dots when the cells are cut in cross-section, or as short segments when the cells are cut obliquely. When viewed in this manner the fibrils are seen to be located near the periphery of the cell (Fig. 10).

In the absence of well defined straight dark fibrils such as have just been described, a tangled network of exceedingly fine fibrils may be seen in the cytoplasm. Although these very fine fibrils are inextricably intertwined to give an appearance like that of felt, they nevertheless have a tendency toward a longitudinal arrangement (Fig. 11).

Finally, we have frequently observed what we regard as tonofibrils. These are situated at the periphery of parenchymal cells which lie adjacent to the stroma. These fibrils sometimes extend beyond the borders of the cells that contain them.

![Fig. 11. Immature Rhabdomyoblastoma: Giant Spindle-shaped Cells with Fine Smooth Fibrils in Protoplasm](image)

(e) Stroma: The rhabdomyoblastomas are rather poor in stroma. Usually it consists merely of thin bands of collagen which run in all directions along with the vessels. Occasionally, cross-sections of masses of parenchymal cells sheathed by collagen septa give a false impression of an alveolar structure.

In sections stained with Masson's trichrome stain it was clearly evident that the thicker bundles of collagen fibrils gave off fine precollagenous fibrils, which ran between the cell masses and between the individual tumor cells. Viewed with oil immersion these precollagenous fibrils were seen to surround each cell in a very fine network. This arrangement was still more distinct in sections stained with silver according to Foot's method (Figs. 12, 13, and 17).

(f) Inclusions: Various kinds of inclusions were seen in these tumors. Scattered throughout one tumor were remnants of preformed cross-striated muscle. Some of these still retained their typical structure, while others were in different stages of degeneration, the contractile substance having been transformed into shapeless fibrillar fragments in which only traces of cross-striation remained (Fig. 14).

In two of the other tumors large, irregularly shaped masses of finely granular material, containing a number of nuclei of varied size and shape, were seen. The nuclei appeared to be undergoing amitotic division, or to be degenerating (Fig. 15). Careful study finally led to the conclusion that these were remains of striated muscle.
Fig. 12. Immature Rhabdomyoblastoma: Network of Argyrophil Fibers

Fig. 13. Immature Rhabdomyoblastoma: Argyrophil Network
Discussion of Microscopic Features: The features in these tumors which have led to the diagnosis of rhabdomyoblastoma can be summed up as follows: (1) a pronounced polymorphism of the tumor cells; (2) the presence of spindle cells; (3) the presence of giant cells; (4) the peculiar arrangement of the stroma; (5) the existence in the cytoplasm of fibrils, which were sometimes cross-striated.

The polymorphism of the cells in these tumors has been emphasized by Masson, Montpellier, Winkler, v. Meyenburg, Roskin, and by many other writers. The great variety of the cell forms encountered leads us to believe that the different cell types represent stages in a process of differentiation.

The large spindle-shaped cells and the syncytial elements containing myofibrils may be regarded as having attained a certain degree of maturity, while the other types of cells may be regarded as being less well differentiated. Of these undifferentiated cell types the small round cells of the “mesenchymal” type are probably the most immature.

Winkler, v. Meyenburg, Borst, Roskin, and especially Ribbert, all emphasize the pronounced embryonal character of rhabdomyoblastomas, and state that it is possible to trace in these tumors all the stages of the gradual transformation of round undifferentiated cells into elongated branching cells, and finally into large cylindrical cells with definite fibrils. My own study of these tumors leads me to the same conclusion.
Other writers have pointed out the frequency of mitosis in these tumors. This is further evidence of their embryonal character. I believe that anamitotic division also occurs. Montpellier and Glasunow have made the same observation.

In the later stages of differentiation the spindle-shaped cells become elongated to form cylindrical cells with a more or less pronounced syncytial structure. In these formations the cell borders vanish, multiple nuclei appear and are drawn toward the periphery, and a great number of cross-striated fibrils appear. The paleness of the nuclei at this stage (Fig. 21) is due, in my opinion, to a degenerative process which occurs in the most highly matured portions of the tumor.

Fig. 15. Mature Rhabdomyoblastoma: Fragment of Decomposing Cross-striated Muscle Tissue in a Tumor; Traces of Cross-striation in Upper Part of Drawing; Numerous Degenerative Nuclei (Oil Immersion)

Fig. 16. Plasmodium: Numerous Nuclei, Fine Smooth Fibrils, and Network of Canals in the Protoplasm (Oil Immersion)

I have no special name to suggest for the spindle-shaped cells which are such a prominent constituent of the rhabdomyoblastomas. Houette has called them "megasarcoctyes," but the term has not received recognition.

In all of our malignant tumors arising from striated muscle I have found giant cells. This fact has led me to consider these cells as specific for rhabdomyoblastomas. Masson has reached a similar conclusion. The spindle-shaped type of giant cell is especially characteristic; cells of this type have been described by Masson, v. Meyenburg, Winkler, Montpellier, Roskin, Glasunow, and others. The plasmodium type of giant cell is sometimes found in such large numbers that Montpellier has distinguished a special plasmodial variety of rhabdomyoblastoma. Marchand, Masson, Winkler, and Roskin.
have given detailed descriptions of these cells. Roskin's report contains excellent illustrations of them. Marchand remarks that it is not easy to fit these cells into the scheme of the development of striated muscle, but that there is no doubt but that they must be classified as muscular elements. I regard them as deformed tumor cells whose volume has been gigantically enlarged as the result of amitotic division and whose appearance has been changed as the result of degenerative processes. The presence of vacuoles and canals resembling a trophospongium in the cytoplasm of these giant cells is due, according to Houette and Diss, to a disturbance of colloidal relationships. It is possible, however, that these vacuoles and canals contain glycogen.

The most distinct characteristic of the stroma of the rhabdomyoblastomas

![Fig. 17. Bundles of Spindle-shaped Cells in Sheaths of Argyrophil Fibers](image)

is the presence of a sheath made of the finest precollagenous fibers around every cell. This arrangement suggests a prototype of the sarcolemma of mature striated muscle. No one, however, has observed the formation of a true sarcolemma in these tumors. It is not clear whether these sheaths are formed from the exoplasm of the tumor cells (Montpellier and Glasunow), or are a product of the fibroblasts of the stroma. It seems to me that both types of cells may take part in their formation, the tumor cell exoplasm being the chief source.

All of the above features are characteristic of the rhabdomyoblastomas, yet the presence of fibrils in cells of these tumors defines their histogenesis better than anything else. The fact that both smooth and striated fibrils are found in the same cell leaves no doubt but that the smooth fibrils are merely
an earlier stage of development of the striated fibrils. I have not seen fibrils in the round cells or in the polymorphic cells. They occur more or less constantly, however, in the spindle-shaped cells. Striated fibrils were seen only in bundles of large spindle-shaped cells and in the syncytial formations. Such cross-striated fibrils were observed in six of my cases. The differentiation of these striated fibrils is of a simplified type. It does not go further than the formation of "Q" and "I" discs. Roskin's paper includes excellent illustrations of the fibrils, and his observations agree closely with my own.

Analyzing the 17 tumors upon which this study is based, I find that only 6 of them show all five of the characteristic features of rhabdomyoblastoma discussed above. Nine cases fall into a second group in which the chief characteristic of these tumors, namely, the presence of cross-striated fibrils, is lacking. Despite this, we feel justified in classifying these 9 cases as true rhabdomyoblastomas. Masson, Montpellier, and other authors classify such tumors as "atypical rhabdomyoblastoma."

The remaining 2 of our 17 tumors possess peculiarities which make it necessary to place them in a third and indefinite group. These two tumors have nothing in common. One of them might be regarded as an example of Montpellier's "myxoid" type of rhabdomyoblastoma. Neither shows any specific differentiating features, and we are forced to classify them merely as cytoblastomas which arose in striated muscle.

The nomenclature of these malignant striated muscle tumors is confused. The term *malignant rhabdomyoblastoma*, which I have adopted, applies to tumors consisting of immature striated muscle cells and showing infiltrating destructive growth and a capacity to metastasize. The terms *myosarcoma* and *rhabdomyosarcoma* are unsuitable because they imply, incorrectly, an origin from mature striated muscle cells. v. Meyenburg distinguishes four types of tumors of striated muscle: (1) rhabdomyoma with pronounced differentiation; (2) myoblastomyoma; (3) myoblastosarcoma; (4) sarcoma. Masson recognizes three types: (1) rhabdomyoma; (2) rhabdomyoblastic sarcoma; (3) atypical rhabdomyoblastic sarcoma. Montpellier's classification is the simplest of all; he divides these tumors into typical and atypical rhabdomyomas. In my own opinion these tumors fall into two groups which
are best designated as *immature rhabdomyoblastoma* and *mature rhabdomyoblastoma*. The mature type of tumor possesses cross-striations while the immature type does not.

**Clinical Features**

(a) **Incidence:** The reports of malignant tumors of striated muscle have been few. Küttner and Landois, in 1913, were able to collect only 10 cases, while Klinge, in 1926, found reports of only 9 cases which he regarded as authentic. Montpellier, in 1929, wrote a detailed review of muscle tumors and collected 25 case reports. Some of his cases, however, were granular-cell rhabdomyomas and therefore must be excluded. In 1933, Glasunow described 6 cases of rhabdomyoblastoma from this institute. Two of these arose in skeletal musculature. In the same year MacCallum reported 2 cases of rhabdomyoma of the extremities. Geschickter has recently mentioned 2 cases that he observed.

![Figure 20: Mature Rhabdomyoblastoma: Syncytium of Spindle-shaped Cells with Cross-striated Fibrils and Degenerating Nuclei](image)

![Figure 21: Degenerative Smooth Fibrils in a Giant Spindle-shaped Cell (Oil Immersion)](image)

In general it may be said that there is very little literature on the subject of the rhabdomyoblastomas and what exists is mainly devoted to the histology of these tumors. The clinical features have been discussed hardly at all, and remain obscure.

In this institute, from 1926 to 1934 inclusive, a total of 75 cases of sarcoma of the soft parts were seen (Chanine). Fifteen, or 20 per cent of these, proved to be malignant rhabdomyoblastomas. This frequency suggests that the rhabdomyoblastomas are by no means as rare as is generally supposed. If careful histological studies were carried out on all soft part sarcomas the number of diagnoses of rhabdomyoblastoma might be considerably increased at the expense of sarcomas in general.

(b) **Age:** It is generally believed that rhabdomyoblastomas usually develop during childhood and adolescence, and cases such as Wolfensberger's and Fujinami's, in patients seventy-five and fifty years of age, are commonly quoted as exceptions to this rule. This is not entirely correct, however. In
the group of 15 cases reported by Montpellier 7 of the patients were under twenty, while the remaining 8 were from twenty-two to sixty-six years of age. Four of Küttner's 10 patients were adults. Thirteen of Glasunow's 21 cases occurred between thirty and sixty years of age. Both of MacCallum's patients were of advanced age, fifty-nine and seventy years.

The 15 cases in the present series were distributed as follows: from ten to twenty years, 2 cases; from twenty to thirty years, 2 cases; from thirty to forty years, 1 case; from forty to fifty years, 2 cases; from fifty to sixty years, 6 cases; from seventy to eighty years, 2 cases. Thus all except 2 of the patients were adults, and two-thirds of them were over forty years of age.

(c) Sex: Of the present group of 15 patients, 9 were men and 6 were women. Napalkow and Küttner had a similar preponderance of males in their series of cases.

(d) Localization: In Montpellier's cases the localization was as follows: thigh, 6 cases; arm, 2 cases; thorax, 4 cases; lumbar region, 2 cases; gluteal region, 3 cases; neck, 2 cases; head, 1 case. In MacCallum's cases the tumors were situated in the arm and leg. In the present series the sites were as follows: thigh, 6 cases; arm, 3 cases; shoulder girdle, 4 cases; thorax, 1 case; lumbar region, 1 case.

Chart I shows the distribution of Montpellier's and MacCallum's cases, as well as those reported here. The lower extremities are most frequently affected, the upper extremities and the lumbar region being the next most frequent sites. It is of interest that not a single case of rhabdomyosarcoma
of the abdominal muscles has been reported. The individual muscles most often affected in the lower extremity are the quadriceps, the abductors, and semi-membranosus, and in the upper extremity, the biceps and brachialis internus.

(e) Symptoms and Course: Rhabdomyoblastoma primary in the bulky muscles of the limbs may reach a considerable size. In one of the cases here reported the growth measured $20 \times 11$ cm., while in Ritter's case the dimensions were $17.5 \times 14 \times 11$ cm. The external appearance of these tumors varies greatly. Generally speaking they are fairly well circumscribed, smooth, and more or less solid. They may be oval, round, or lobulated.

When situated deep in bulky muscles they may be comparatively mobile, but this mobility disappears when the muscles are contracted. This finding is characteristic of a muscular localization (Fig. 22).

The skin over the tumor is usually unchanged except for the presence of a network of dilated veins. In some instances, however, these growths have infiltrated and broken through the skin to form ulcerated, mushroom-like, bleeding tumors (Fig. 23 and 24). In 6 of my cases ulceration had occurred; of these, 2 were primary cases while 4 were recurrences. Lambl, Davis, Glasunow, and Muller have described destruction of bone by this type of tumor. One of my patients had, in addition to rhabdomyoblastoma, an epithelioma of the lower lip.

These tumors do not, as a rule, give rise to symptoms, and may remain undetected until they have attained a large size. They cause pain only when they happen to be situated near a nerve trunk, and they limit motion only when they are located near a joint. When situated in superficial muscles
they are sometimes recognized early. Usually the tumor increases in size more or less rapidly, either expanding laterally and deeply or growing outwards through the skin.

The gross appearance of the tumors varies greatly. Küttner states that cystic cavities may develop in them. Montpellier described a myxomatous type of rhabdomyoblastoma. According to MacCallum, distinct macroscopic boundaries are present. If the tumor originates in the center of the muscle body, the latter is inflated. If it develops in the periphery of a muscle, growth is lengthwise along the intermuscular septa. In the cases which I have seen the tumor was so extensive that these relationships could no longer be established. In the rapidly growing types of rhabdomyoblastoma, infiltrative and destructive growth prevails, while in the slowly growing types expansive growth predominates. The cut surface of the tumor is usually grayish-pink in color, rather soft, fibrous, sometimes lobulated, and often shows areas of hemorrhage or yellowish degeneration.

The clinical course depends to a certain degree upon the histological structure. The immature type of tumor, in which small spindle-shaped cells predominate, grows more rapidly and more often ulcerates. The mature type of tumor, in which large spindle-shaped cells and giant cells predominate, grows more slowly, sometimes over a period of years, without metastases for a very long time. The degree of maturity of the rhabdomyoblastoma has also prognostic significance. In our mature cases, which numbered 6, there was but 1 death; among our 9 immature cases there were 6 deaths.

The only general symptom observed with these tumors is a slight elevation of temperature, which occurs in some cases. The temperature may rise to 38° or 38.5° at the end of the day, due in most instances to secondary infection accompanying ulceration of the tumor. In only two of my cases did I fail to find a cause for the elevated temperature other than the presence of the tumor itself.

There is usually no substantial change in the blood picture. Advanced cases may show a decrease in hemoglobin and red blood cells, a slight leukocytosis, and a deviation to the left of the leukocytic formula. Weakness and cachexia of course occur in these advanced cases.

(f) Metastases: Involvement of the regional lymph nodes is rarely observed, Ritter's case being the only proved instance yet reported. In my series of cases enlarged regional lymph nodes were palpated in 6 patients, but in the 2 cases in which these nodes were subjected to histologic examination, no metastases were found.

Distant metastasis, by both the vascular and lymphatic routes, has frequently been observed. Thus in Ritter's case there were metastases in the lungs, pleura, mediastinal lymph nodes, and pericardium, while in Burgess' case metastases were found in the subcutaneous tissues, mammary glands, ribs, skull, sternum, lungs, pancreas, suprarenals, and ovary. In my cases metastases were found in the lungs in 4 cases. In one instance the metastasis developed one year after the onset of the disease and was accompanied by hemorrhagic pleurisy. In two of the cases the lung metastases were noted one year after operation. In the fourth patient there was local recurrence
of the disease with metastases in both lungs, in mediastinal and retroperitoneal lymph nodes, and at two sites in the small intestine. These intestinal metastases ulcerated and fatal hemorrhage ensued.

(g) Recurrence: The most striking feature of the natural history of the rhabdomyoblastomas is their tendency to recur locally following excision. This has been noted by all who have written about these tumors. In the present series of cases, 10 were recurrent following operation done before admission to this institute. Four of these 10 patients came to us for treatment after one recurrence, 4 after two recurrences, 1 after five recurrences and 1 after thirteen recurrences. Two of the 5 patients with primary tumors developed local recurrences following operation done here.

The interval between removal and recurrence varies from a few weeks to several years. In about half of our cases the recurrence was noted soon after operation; in the other half the interval ranged from one to three and one-half years. The recurrent tumors are usually situated in the scar of the previous operation. They are sometimes multiple and are usually adherent to the overlying skin. They grow rapidly and often ulcerate. The tumors which recur many times are comparatively benign types. Two cases from our series illustrate these features.

Case 12: A housewife, aged forty-eight, first noticed an indurated area in the soft tissues of her upper arm eleven years previous to admission. The induration followed an injury. Nine months after the tumor had first been detected it was excised and found to be some 3 cm. in diameter. Three and one-half years later there was a recurrence in the scar. It also was excised. Since then operation had been done almost every year, a total of thirteen times. In the earlier operations recurrences in the soft tissues of the arm were dissected. Later the head of the humerus, and finally the scapula, were removed. In this institute disarticulation of the arm was done and part of the clavicle removed. A year later there was local recurrence in the scar and death occurred with pulmonary metastases.
Case 7: A working woman, aged forty-seven, had a small tumor in the left arm nine years previously. Within a year it attained a diameter of 4 or 5 cm. It was then excised. Two years later a recurrence in the scar was excised. After two years more it was necessary to remove another recurrence. At yearly intervals two more operations were done before the patient came to this institute for a fifth recurrence. Here a radical operation was done, but recurrence again appeared after a year's interval.

Diagnosis of Rhabdomyoblastoma

It is not always easy to recognize the rhabdomyoblastomas. A fairly well outlined, rather solid tumor, situated deep in a muscle and moving with it, which has grown rapidly and painlessly to a large size, and is covered with skin which shows no change except the presence of a network of dilated veins, may be presumed to be a rhabdomyoblastoma.

In the differential diagnosis neurogenic sarcoma must be considered. The principal sites of this type of sarcoma are the popliteal region, the groin, the gluteal region, and the interscapular region. Sometimes there are associated stigmata of neurofibromatosis.

Benign tumors such as lipomas, fibromas, and angiomas rarely occur in the depths of muscles.

Inflammatory and traumatic disease of muscles must be kept in mind in the differential diagnosis. Gumma, tuberculosis, and echinococcus disease can be excluded by the usual diagnostic means. Myositis ossificans may be differentiated by the roentgenographic findings.

In certain advanced cases of rhabdomyoblastoma where the tumor has infiltrated the soft tissues widely and involved the overlying skin, making it impossible to determine its primary site, it may be difficult or even impossible to make a correct diagnosis. The advanced age of the patient, the comparatively slow growth of the tumor, and the absence of metastasis should, however, suggest rhabdomyoblastoma.

Actually these tumors are rarely diagnosed clinically. They usually pass as fibroma or fibrosarcoma. Even when examined histologically they escape detection unless special staining methods are used to bring out the fibrils and other details in the tumor cells.

Treatment of Rhabdomyoblastomas

The treatment should be radical operation. Köttner recommends total removal of the affected muscle in every case. This suggestion, however, is more theoretical than practical and can be carried out only in tumors that are detected in an early stage. Such tumors are rarely seen. Much oftener we have to deal with large tumors involving groups of muscles. In such cases it seems more expedient to carry out an extensive excision of the tumor through the surrounding uninvolved tissues. In an operation of this type the tumor is not seen. Operations which attempt to separate or dissect out the tumor from its bed should be avoided.

In view of the strong tendency of the rhabdomyoblastomas to recur locally, the electrosurgical method of excision would seem to be preferable to knife excision. Experience has shown, however, that even this method often fails to prevent recurrences. When the tumor involves tissues widely, or
when it is situated in the vicinity of a joint, amputation or disarticulation should be done. Such radical therapy should be carried out, of course, only in cases in which examination of the lungs has shown no metastases.

All of our 15 patients were operated upon, several repeatedly. Knife excision was done in 7 cases. Two of these patients died within a few months after operation, 2 are living without recurrence, and the remaining 3 could not be followed. Electrosurgical excision was done in 4 cases. Three of these patients died within a few months after operation, while the fate of the fourth is unknown. Disarticulation was done in 3 patients, and all died. The one patient that had amputation is still living, but with a recurrence which developed one year after operation.

In 10 of our cases the surgical attack was supplemented by radiation, but without apparent benefit. In 5 cases the radiation was given before operation; it did not reduce the size of the tumor or increase its mobility. In the other 5 cases the radiation was given following operation, but was equally ineffective.

Conclusions

1. Careful histological study of malignant tumors arising in skeletal muscles will reveal that a considerable percentage of these tumors are rhabdomyoblastomas.

2. These tumors are characterized by: (a) polymorphism of the cells of which they are composed; (b) the presence of spindle-shaped cells and syncytia; (c) the presence of giant cells; (d) a characteristic pattern of the stroma; (e) the presence of myofibrils in the tumor cells.

3. Two types of rhabdomyoblastoma can be distinguished: the mature form and the immature form.

4. These tumors develop in patients of all ages. Their course is sometimes protracted. Radiation is of no benefit, and surgery is often followed by local recurrence of the disease, with eventual metastasis.

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