PERIPHERAL NERVE TUMORS WITH RHABDOMYOSARCOMATOUS DIFFERENTIATION (MALIGNANT "TRITON" TUMORS)

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Three peripheral nerve sheath tumors are reported which showed an intermingling of schwannian elements and rhabdomyosarcoma, a type of compound tumor first described by Masson in 1932. One of the patients had von Recklinghausen's disease. The tumor in two patients showed pleomorphic rhabdomyosarcoma, while in a third patient it grew partially as an embryonal rhabdomyosarcoma. All three tumors contained areas of malignant schwannoma, but, in this latter case, the rhabdomyosarcoma appeared to arise from an area of benign Schwann cell growth. Two of the patients died following spread of the malignant muscle component. All previously reported examples of this tumor developed in individuals showing stigmata of von Recklinghausen's disease. Data from our three cases and from seven cases from the literature indicates the tumor to be highly malignant. Of the nine patients with follow-up, seven were dead within 3 to 20 months of their diagnosis. Those recurrent tumors or metastases that were examined histologically (four cases) showed rhabdomyosarcoma.

THE OCCURRENCE OF TUMORS OF PERIPHERAL nerves made up in part by benign and malignant tissue components other than those normally found in peripheral nerves, in patients with or without von Recklinghausen's disease, is well known. Survey of the literature as well as material examined in the Surgical Pathology Laboratory at Memorial Hospital reveals that peripheral nerve tumors can contain such unusual components as: osteoid,[50,51] bone,[52] benign striated muscle,[36] cartilage,[50] nevus cells,[12] malignant melanoma,[12] embryonic,[5,50] rosette-forming,[29,48] and glandular malignant neuroepithelium,[6,13,14] liposarcoma,[8] angiosarcoma,[12] and embryonal and plemorphic rhabdomyosarcoma.[7,11]

The appearance of any of these elements, with perhaps the exception of osteoid and cartilage, is uncommon. We recently, however, had the opportunity to see three cases of rhabdomyosarcoma which developed in a nerve sheath tumor, a type of compound tumor referred to at our respective medical centers as malignant "Triton" tumor, for reasons which will be indicated. A discussion of these cases along with a review of seven others from the literature is presented.

MATERIALS AND METHODS

Tissues were fixed in 10% buffered formalin, embedded in paraffin, cut and stained with hematoxylin and eosin, Masson trichrome, Wilder's reticulum, and phosphotungstic acid-hematoxylin.

CASE REPORTS

Case 1. *Clinical history:* A 31-year-old Caucasian man was admitted to St. Luke's Hospi-

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tal, Denver, Colo., April 18, 1968, because of breathing and swallowing difficulties of several years duration. He had recently lost 10 lb. His deceased mother was said to have had multiple cutaneous "lipomas." Examination revealed a large "goiter" in the right neck and "lipomas" in the skin. Laboratory studies were essentially negative.

On April 19, 1968, a well-encapsulated mass attached to the right vagus nerve and measuring 9 cm long and 4 cm in greatest diameter was removed from his right neck and superior mediastinum. Pathologic diagnosis was malignant neurilemoma growing partly as rhabdomyosarcoma.

The patient was readmitted in September 1968, and treated for an active duodenal ulcer with a partial gastrectomy, vagotomy, and Bilroth 11 anastomosis. During the summer of 1969, he again developed breathing and swallowing difficulties. Roentgenograms were suggestive of pulmonary metastases, and he was treated with radiation therapy and phenylalanine mustard. He died on September 18, 1969, about 18 months following diagnosis of his neurogenic tumor.

Pathologic findings: The surgical specimen removed in April 1968, weighed 62 g and consisted of homogeneously gray-tan tissue showing cystic and necrotic areas. Microscopy revealed the outer portion of the tumor to be comprised in part of plexiform nerve segments containing delicate argyrophilic fibers. From here an often dense growth of interlacing Schwann cells sometimes resembling a primary fibrous tumor extended centrally where it abruptly merged (Fig. 1) with hyperchromatic spindle and round Schwann cells. This malignant neurilemomatous component showed foci of cartilaginous metaplasia (Fig. 2) and, in other regions, the intimate admixture of round cells containing abundant eosinophilic cytoplasm and small dense nuclei. The cytoplasm of a few of these round cells contained longitudinal fibrils and cross striations, features characteristic of skeletal muscle. Dense clusters of malignant muscle tissue could also be seen and involved both inter- (Fig. 3) and perivascular sites.

Autopsy revealed a neurofibromatosis with widespread involvement of the skin. There was an ileal leiomyoma. The larynx, upper esophagus, right supraclavicular area, and right mediastinum were invaded by firm tumor which proved histologically to be recurrent malignant neurilemoma. Similar tumor encased major vessels at the arch of the aorta and invaded the upper lobe of the right lung. Involvement of the innominate artery and adjacent trachea had led to rupture of this vessel into the trachea causing the patient's death. Metastases averaging 2.5 cm in diameter were present in both lungs, and a smaller solitary one was found on the posterior leaflet of the tricuspid valve; microscopically (Fig. 4), all metastases demonstrated poorly differentiated spindle cells admixed with plump, often blunt-ended or tapered rhabdomyoblasts—traits of a pleomorphic rhabdomyosarcoma.

Case 2. *Clinical history: A 53-year-old* woman was admitted to Memorial Hospital, New York, N.Y., on June 11, 1970, because of progressive weakness of her left arm and hand. Seven months earlier, insidious pain developed in her left anterior chest, shoulder, and back, and then progressed medially along the left arm to the fifth finger. A left Horner's syndrome had been present for 2 months. At age 12, she received a series of x-ray treatments for a draining left submandibular gland. At 43 years of age, she had a thyroid adenoma excised, and a year later she had undergone a left radical mastectomy followed by radiation therapy and oophorectomy for infiltrating duct carcinoma of the breast. Neurologic examination revealed wasting and weakness of intrinsic muscles of the left hand and mild weakness of the left triceps brachii. Pin, touch, and temperature sensations were diminished over the medial aspect of this extremity in the T1 and T2 dermatomes. Myelogram demonstrated an extramedullary intradural mass at the level of the left T1-T2 interspace.

On June 30, a 1 × 2 cm intradural neurilemoma was removed from its point of attachment to the posterior T1 nerve root. The surgeon felt that the mass had been completely excised. Soon after discharge, her brachial plexus was explored at another hospital and additional neurilemomatous tissue resected.

Because of numbness over her right buttock and leg, she was readmitted to MH in December 1970. Micturition was initiated with difficulty, and she had been complaining of increasingly severe constipation. Examination revealed decreased pin and light touch sensation over the leg, lower abdomen, and buttock on the right side as well as loss of position and vibratory sensation in all extremities but most pronounced in the right leg. She had a marked proprioceptive ataxia. Myelogram revealed recurrent tumor extending from T1-T6 which on laminectomy at the T4-T6 level filled the subdural space. Histology showed a spindle cell tumor differentiating toward embryonal rhabdomyosarcoma. Postoperatively, she received 3000 rads of radiation. She had a progressive lower paraplegia and sensory deficit and developed headache, nuchal rigidity, and progressive obtundation. Spinal fluid con-

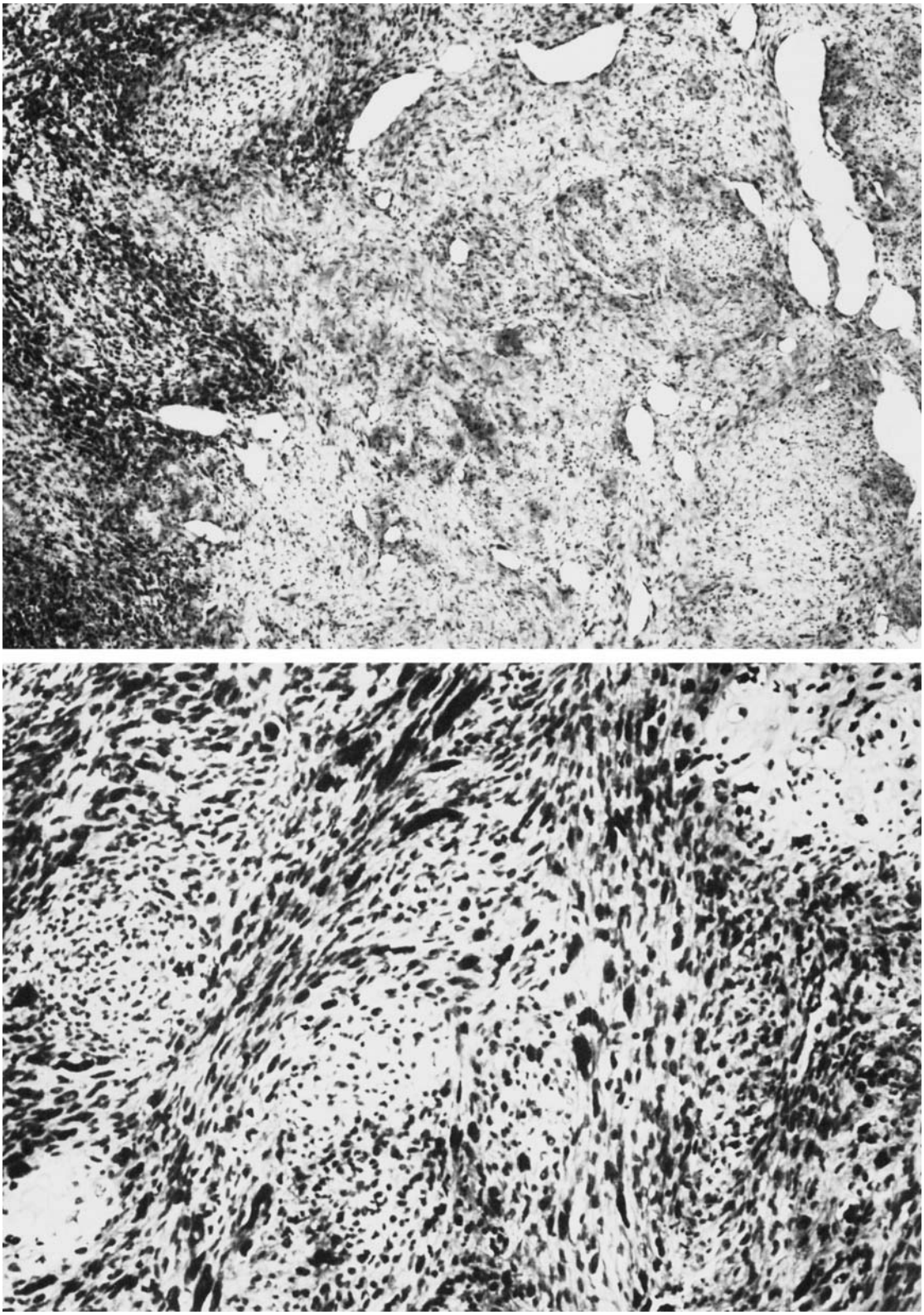

FIG. 1 *(top)*. Primary tumor, case 1. Benign, highly vascular, subcapsular neurilemoma centrally (on left) shows malignant change (H and E, ×74). FIG. 2 *(bottom)*. Primary tumor, case 1. Malignant area comprised of hyperchromatic Schwann cells showing a tendency to palisade and undergo focal cartilaginous metaplasia. Neoplastic muscle cells are not seen in this view (H and E, ×320).

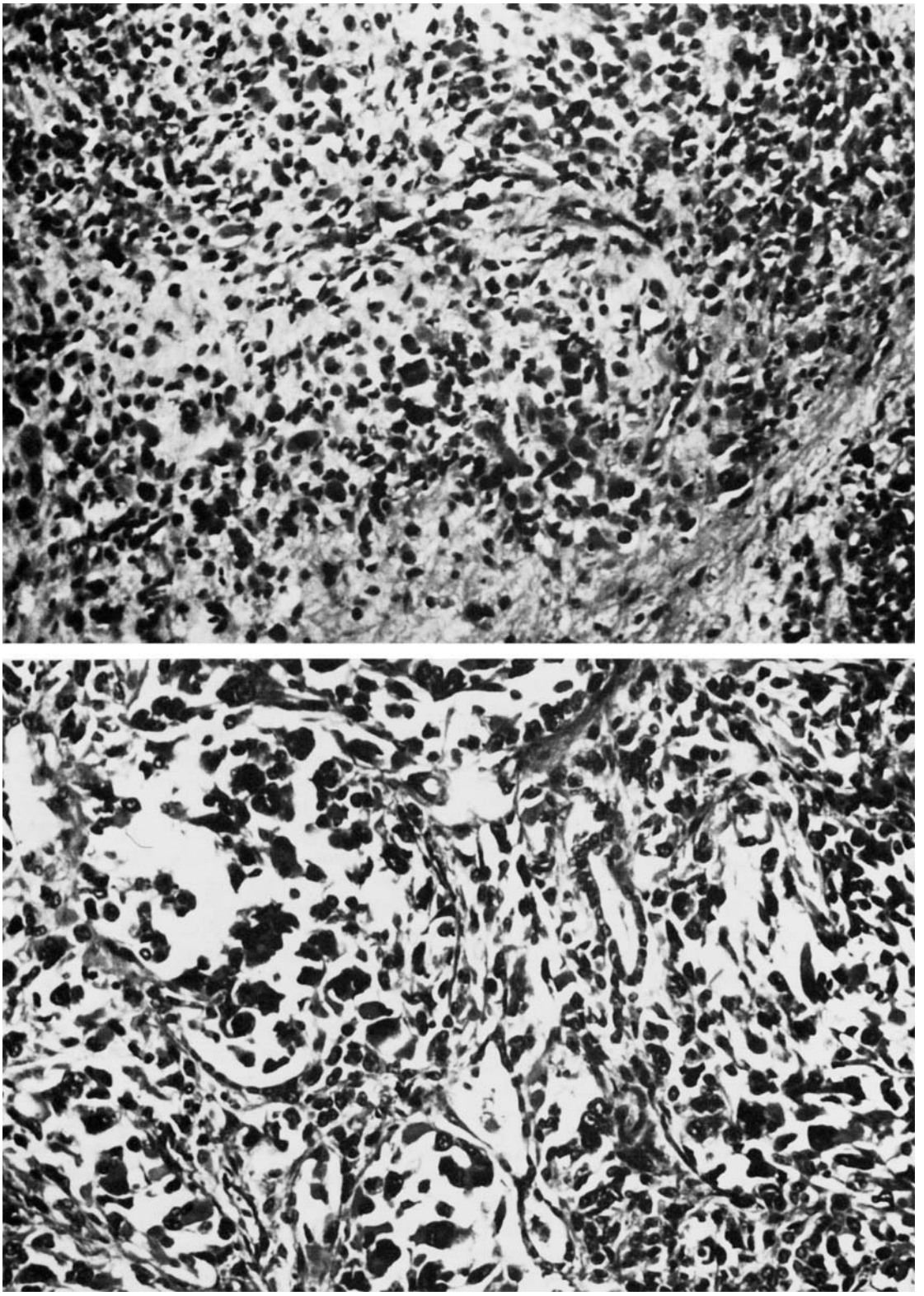

FIG. 3 *(top)*. Primary tumor, case 1. Area showing admixture of undifferentiated small malignant cells and ovoid to round rhabdomyoblasts bearing dense hypereosinophilic cytoplasm (H and E, ×320). FIG. 4 *(bottom)*. Case 1. Metastatic rhabdomyosarcoma of pleomorphic type in lung. Strap and multinucleated giant rhabdomyoblasts fill alveolar spaces between residual bronchioles (H and E, ×500).

tained malignant cells. Death occurred on January 17, 1971.

Pathologic findings: Microscopic review of the original spinal nerve root biopsy taken in June revealed a malignant neurilemoma with palisaded areas and delicate agyrophilic fiber production that in one seemingly Antoni type B area (Fig. 5) had undergone subtle malignant change. Here neatly layered histologically benign Schwann cells blended with disarrayed round eosinophilic cells. Many of these latter (Fig. 6) showed the cytoplasmic features of embryonal rhabdomyoblasts, with either round shapes and abundant cytoplasm that gave a cobweb appearance because of the concentric arrangement of fibrils or stellate forms showing longitudinal fibril formation. No clear-cut cross striations were evident. The most striking feature of this myoblastic cell growth was its intimate admixture with benign Schwann cells and the presence of what appeared to be transition forms between the two.

At autopsy, the left brachial plexus was encased by dense white tissue which obscured the nerve trunks of its lower segments and which proved to represent a mixture of benign and malignant neurilemoma and scar tissue. Benign neurilemoma extended through the pleurae to the apex of the left lung. Tumor, in addition, infiltrated the T2 spinal nerve and its nerve root and ganglion (the T1 dorsal root ganglion could not be identified). Histology of this (Fig. 7) revealed a benign neurilemoma which was interrupted at irregular intervals by nests of both elongated and rounded differentiating embryonal rhabdomyoblasts, some of which bore cytoplasmic cross striations. In other foci, a malignant Schwann cell growth could be seen. At the T1-T2 level, this compound tumor formed a solid 0.5 cm subdural mass which compressed the lateral surface of the cord. From here, the tumor grew within the subarachnoid space, circumferentially enveloping the spinal cord along most of its length. Penetration into the cord itself took place in the lower cervical region where the heterogeneous tumor entered through the left dorsal quadrant (Fig. 9a) to become an intramedullary extension (Fig. 9b). Here, in addition to essentially undifferentiated small cells (Fig. 10), a vast array of differentiating embryonal rhabdomyoblasts, which included (Figs. 10, 11) ring, cobweb, round or globoid, granular, tapered, bipolar, and multinucleated forms, were readily identified. Many of these cells contained cross striations (Fig. 11), and occasional strap cells were seen (Fig. 12). Similar tumor showing a primarily rhabdomyosarcomatous differentiation extended rostrally over the brain stem, cerebeulm, and infundibular region and encased nerve roots of the cauda equina. No visceral or lymph node metastases were found.

Case 3. *Clinical history:* A 41-year-old Caucasian woman presented in Surgical Clinic at St. Luke's Hospital, Denver, Colo., on September 10, 1971, with two superficial "lipomas" located on the mid upper back and right shoulder. The patient had no unusual cutaneous pigmentation. An excisional biopsy of the interscapular mass was performed. Her shoulder lesion was not excised, and the patient has been followed for one year with no further evidence of disease.

Pathologic findings: Examination revealed a 2 × 0.8 × 0.5 cm pear-shaped encapsulated tumor which on cut section was firm, yellow-tan, and contained small silk-like areas. Microscopic study showed a highly cellular malignant tumor comprised of tightly packed and sometimes palisaded hyperchromatic malignant Schwann cells (Fig. 13). These were interrupted at irregular intervals by plump, often multinucleated rhabdomyoblasts containing abundant eosinophilic cytoplasm. No cross striations were seen.

REPORTED CASES

Seven previously reported cases (Table 1) of peripheral nerve tumors which contained rhabdomyosarcoma within the primary tumor and/or metastases were found. To the best of our knowledge, the first description of this unusual tumor was published by Masson in 1932.[24] Upon later review of slides from a case originally reported by Simon[42] as a malignant schwannoma, Masson discovered areas of the tumor which showed embryonal rhabdomyosarcoma.[24] The sarcoma arose centrally from within the substance of a malignant neurofibroma which had developed in a superficial left cervical nerve of a 23-year-old man with von Recklinghausen's disease. No follow-up on the case was reported.

The second and third examples were recorded by Masson and Martin in 1938.[28] A benign neurofibroma had been removed from the right wrist of an 18-year-old girl. Another arose from a nerve in her left thigh. Histologically, the thigh mass showed a neurofibroma in which a pleomorphic rhabdomyosarcoma containing a variety of myosarcoma cells including giant globular forms had developed. The tumor recured three times within a 23-month period, myosarcoma being identified in two of these three specimens. She eventually

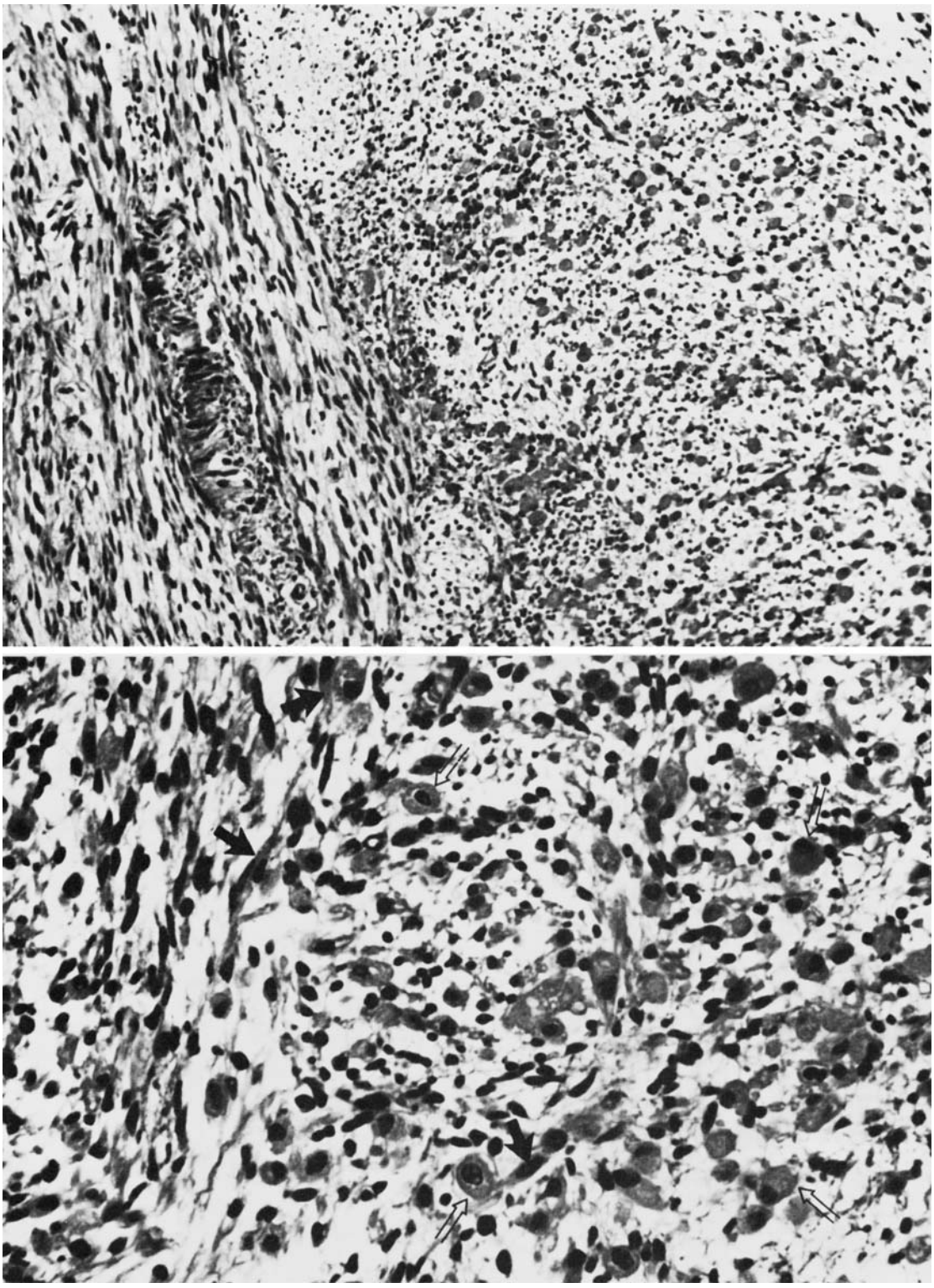

FIG. 5 *(top)*. Primary tumor, case 2. Somewhat closely aligned Schwann cells of benign neurilemoma border loosely structured area (on right) populated by both hyperchromatic undifferentiated cells and others showing rounded profiles and considerable eosinophilic cytoplasm (H and E, ×200). FIG. 6 *(bottom)*. Case 2. Higher magnification of loose area shown in Fig. 5. Open arrows point to typical embryonal rhabdomyoblasts scattered among spindle nerve sheath elements and undifferentiated small dark cells. Contiguity of different cell types and thickening of many schwannian elements (closed arrows) suggests that embryonal rhabdomyoblasts may have arisen by metaplasia from neoplastic Schwann cells (H and E, ×500).

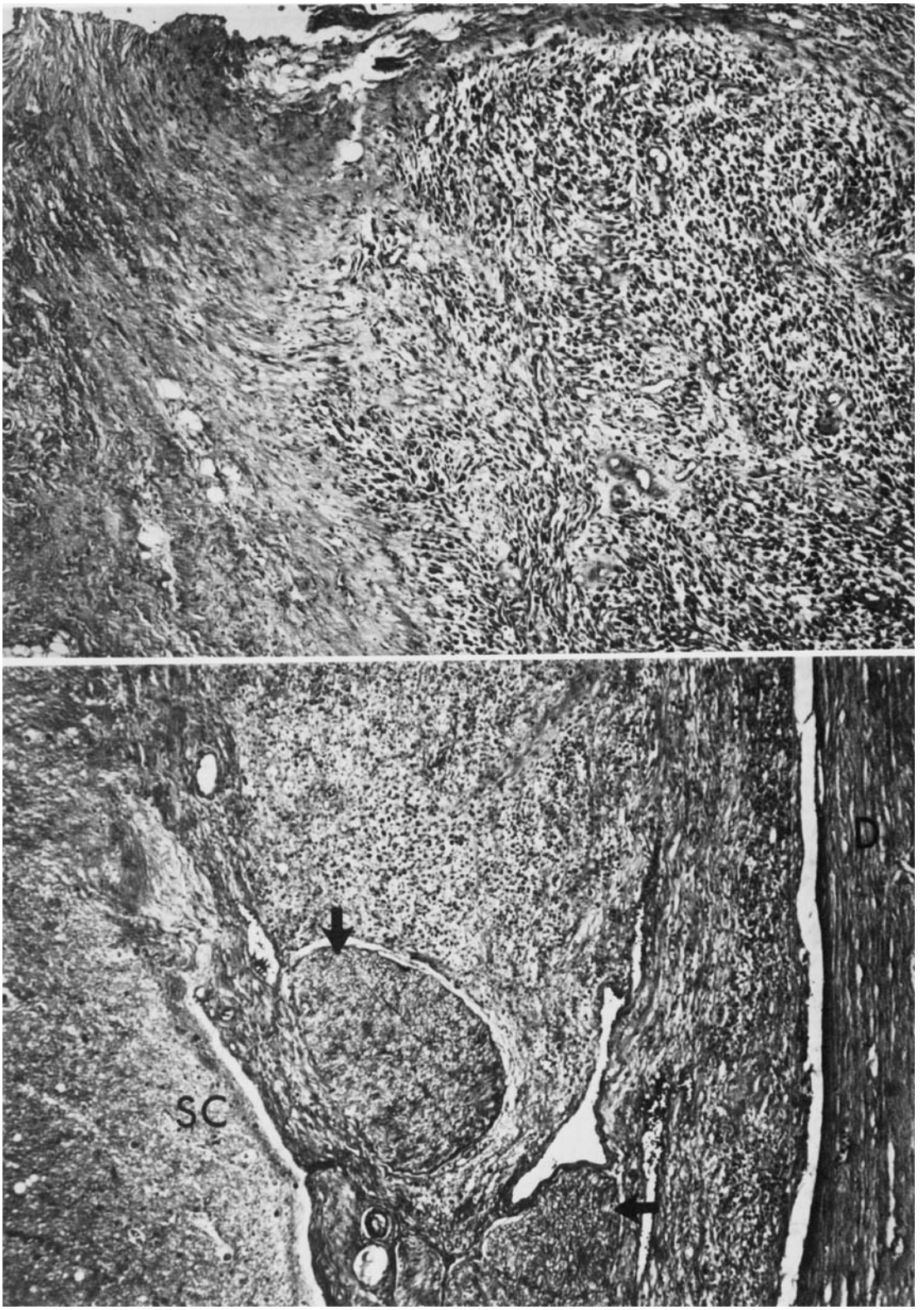

FIG. 7 *(top)*. Recurrent paraspinal tumor, case 2. Cluster of embryonal rhabdomyosarcoma cells embedded within histologically benign neurilemomatous tissue. Spindle, polyhedral, and round-to-ovoid neoplastic muscle cells are seen (H and E, ×125). FIG. 8 *(bottom)*. Tumor extension in leptomeninges of spinal cord, case 2. Malignant spindle cell tumor abuts both spinal cord (SC) and dura (D) and encases portions of spinal nerve roots (arrows). Differentiation toward embryonal rhabdomyocarcoma can be seen (H and E, ×125).

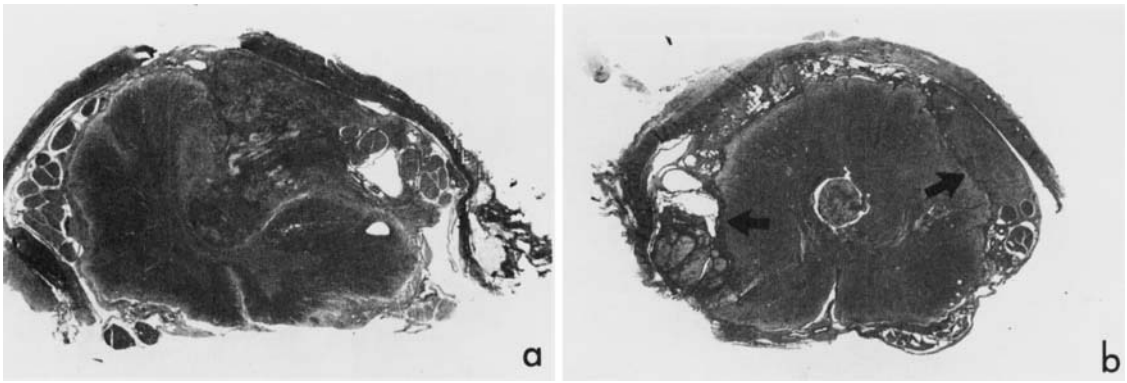

FIG. 9. Case 2. Transverse sections of spinal cord: a, c5; b, T1. a. Tumor infiltrates from subarachnoid area into left dorsal column of cord. b. Malignant neurilemoma enveloping spinal cord concentrates around nerve roots within the subarachnoid space. Bilateral cord compression (arrows) as well as intramedullary tumor growth can be seen (H and E, ×5).

died due to a carcinoma of the breast.[27] The other case involved a 32-year-old man with von Recklinhausen's disease from whom 200 cutaneous neurofibromas[27] had been removed prior to the development of a highly malignant neurofibroma in the subcapsular area. The latter was incompletely excised. Within 3 months, the patient died with numerous pulmonary metastases (1–3 cm), which Masson felt showed "neurosarcoma" in addition to rhabdomyosarcoma.[28]

The fourth account is credited to Agustsson et al.[1] who, in 1955, described a 14-year-old boy with von Recklinghausen's disease and

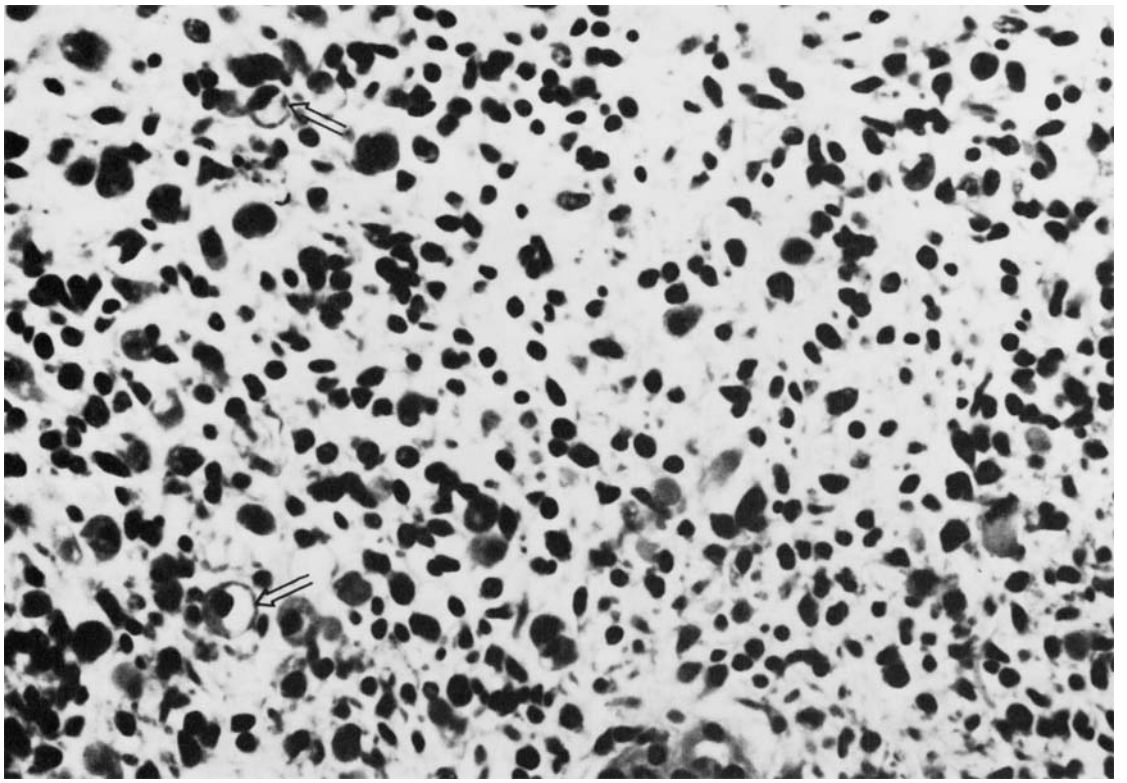

FIG. 10. Tumor extension into spinal cord, case 2. Morphologically simple embryonal rhabdomyoblasts (open arrows) showing narrow marginal ring of myoplasm have developed in field of, for the most part, undifferentiated malignant cells (H and E, ×500).

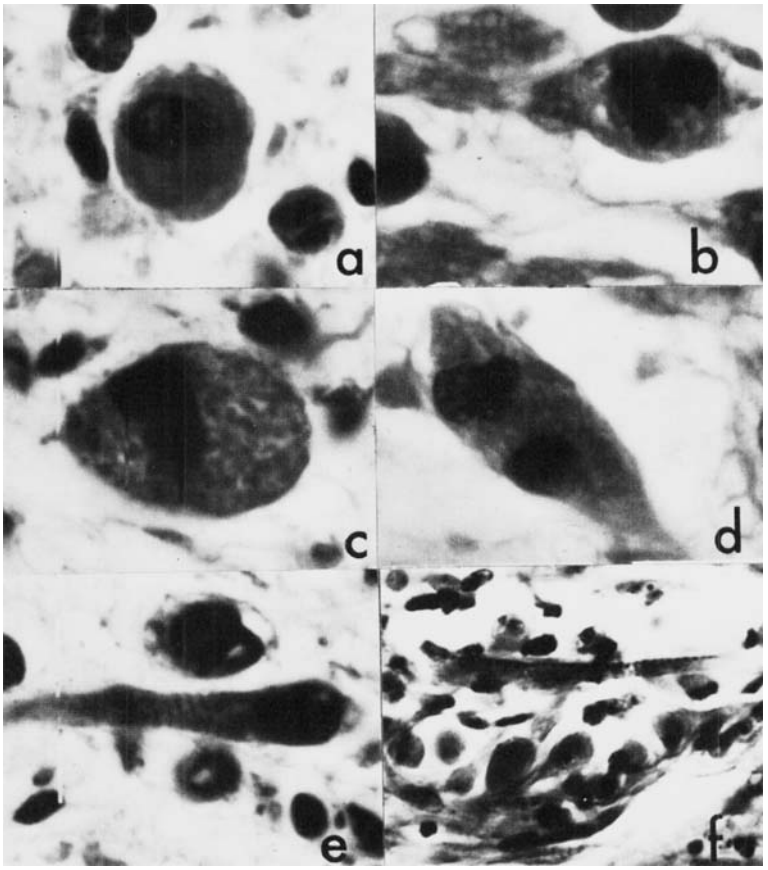

FIG. 11. Variety of differentiating embryonal rhabdomyoblasts found in infiltrating tumor from case 2 includes round or globoid (a), cobweb (b and d), granular (c), tapered or tadpole (e), binucleated (b and d), and cross-striated (e and f) forms (H and E, a-d ×1250; H and E, e ×800; and PTAH, f ×500).

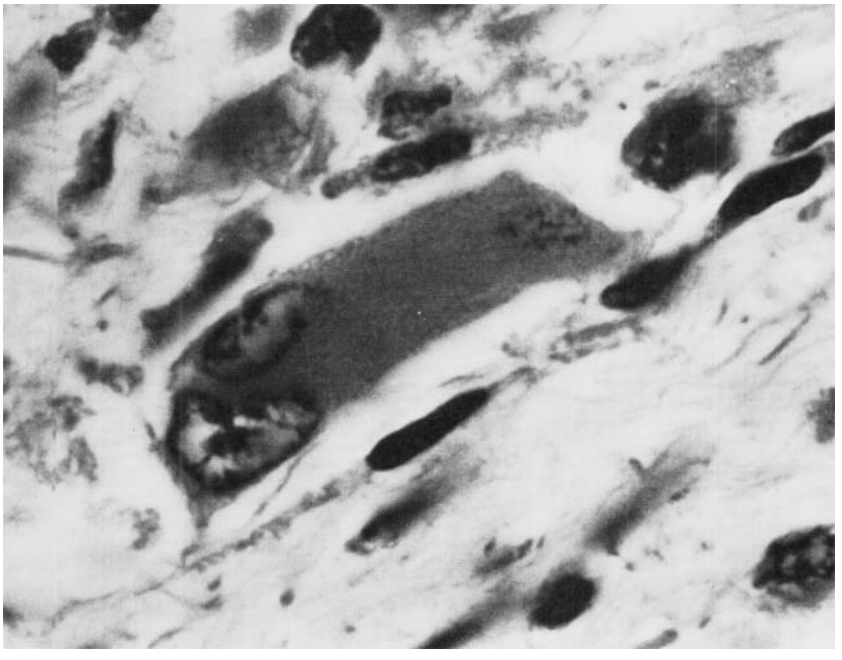

FIG. 12. Case 2. Rare non-striated giant strap-like rhabdomyoblasts were -occasionally identified in the intraspinal tumor (H and E, ×1250).

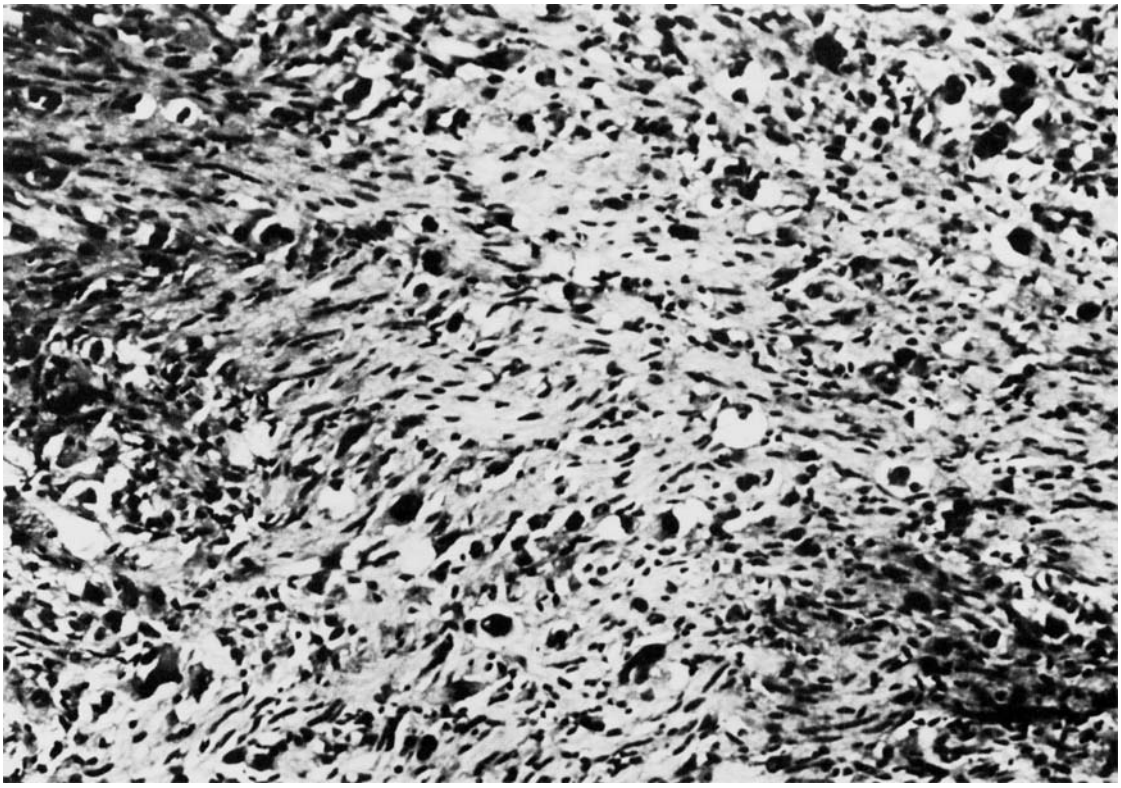

FIG. 13. Case 3. Plump, irregularly shaped rhabdomyoblasts are conspicuous among waves of closely packed Schwann cells. Schwannian elements in other areas were sufficiently atypical to warrant a diagnosis of malignant neurilemoma (H and E, ×320).

elephantiasis neuromatosis of the left thigh. Examination of the left hind quarter specimen revealed two tumors, a 12 cm in diameter benign neurofibroma out of which grew a smaller 5-cm malignant nerve sheath tumor containing pleomorphic rhabdomyosarcoma. The boy was dead in 15 months with roentgenographic evidence of pulmonary metastases. No autopsy was performed.[45]

The fifth and sixth cases[8] were of two patients with von Recklinghausen's disease in whom neurofibromas showed the additional component of a rhabdomyosarcoma. Neither tumor was demonstrated to arise from a nerve trunk. The tumor in one patient, a 57-year-old man, appeared in the left forearm and recurred after treatment. He died within 20 months with clinical evidence of pulmonary metastases. The other patient, a 29-year-old man, was determined to have an inoperable tumor involving the left chest and died 3 months after diagnostic biopsy. Neither man was autopsied.[45]

White, in 1971,[51] reported the seventh example of this entity. A 35-year-old man with von Recklinghausen's disease died following recurrence of a cervical malignant schwannoma. Dr. Saul Kay kindly gave us the opportunity to review slides in this case. Though rhabdomyosarcoma could not be found in the original specimen, the recurrence showed a striking intermixture of malignant Schwann cells and plump rhabdomyoblasts.

DISCUSSION

The view which originated with French and Canadian investigators[2,4,22,25,26,43] that peripheral nerve tumors are derived from Schwann cells of the neurilemmal sheath is now widely accepted by American pathologists. Cell culture work by Murray and Stout[31] was partially responsible for this acceptance. Since Masson's suggestion in 1932[25] that the fibrous element often seen in the tumors probably comes from the Schwann cells themselves, many unusual growth traits have been attributed to these cells. It is thought by

TABLE 1. Reported Cases of Peripheral Nerve Tumors with Rhabdomyosarcomatous Differentiation

Author and year	Age/sex	Location of tumor	Multiple neurilemomas or other evidence Recklinghausen's disease	Histology of primary tumor	Follow-up
Masson* (1932)	23/M	Lt. neck	Yes	Malig. neurofibroma with embryonal rhabdomyosarcoma	None
Masson and Martin (1938)	18/F	Lt. thigh	Yes	Neurofiroma with pleomorphic rhabdomyosarcoma	Free of sarcoma 10 yrs. after diagnosis. Died with breast carcinoma
	32/M	Subscapular area	Yes	Malig. neurofibroma (tumor incompletely excised)	Dead in 3 mos. with numerous lung metastases comprised of both malignant neurogenic tumor and rhabdomyosarcoma
Agustsson et al. (1955)	14/M	Lt. thigh	Yes	Benign and malignant neurofibroma with pleomorphic rhabdomyosarcoma	Dead in 15 mos. with x-ray evid. of lung metastases. No autopsy.
D'Agostino et al. (1963)	57/M	Lt. forearm	Yes	Neurofibroma with pleomorphic rhabdomyosarcoma	Dead in 20 mos. with clinical impression of lung metastases No autopsy
	29/M	Post. chest	Yes	Neurofibroma with pleomorphic rhabdomyosarcoma	Dead in 3 mos. No autopsy
White (1971)	35/M	Cervical region	Yes	Malignant schwannoma	Dead in 5 mos. with rhabdomyoblasts growing in recurrent malignant schwannoma
Present report	31/M	Rt. neck	Yes	Benign and malignant plexiform meurilemona with foci of cartilaginous metaplasia and admixed rhabdomyoblasts	Dead in 18 mos. with recurring malignant neurilemoma in neck and metastatic pleomorphic rhabdomyosarcoma in lungs and tricuspid valve of the heart
	53/F	Tl spinal nerve root	No	Embryonal rhabdomyosarcoma growing in benign neurilemoma. Malignant neurilemoma present in some areas	Dead in 1 yr. with malignant neurilemoma and embryonal rhabdomyosarcoma. Tumor extended along length of spinal cord and into leptomeninges at base of brain
	41/F	Mid back	No	Malignant neurilemoma containing admixed rhabdomyoblasts	Alive and well 1 year

* Reassessment of case first published by Simon in 1922.[42]

some that they can form, for example, cartilage, epithelial glands, and even striated muscle. To the best of our knowledge, with the exception of the formation of reticulin fibers,[32,33] this belief in the metaplastic versatility of Schwann cells rests solely on histologic examination of neoplasms and has not been confirmed by tissue culture studies.

Because Schwann cells are derivatives of the neural crest[17] and, therefore, of neuroectodermal and not mesodermal origin, it is etymologically improper to designate peripheral

nerve sheath tumors as neurogenic sarcomas. Yet neurilemmal tumors undergo sarcomatous change, on occasion. Those composed in part of rhabdomyosarcoma are highly malignant. Of the 10 cases included in the present report (Table 1), seven patients died with evidence of either metastatic (four cases with lung involvement) or extensive local disease (three cases) from 3 to 20 months after the original diagnosis. A similar rapidly fatal course has been reported by some authors[8,51] for malignant neurilemoma devoid of muscle elements. Those recurrences or metastases that were examined (four cases) showed rhabdomyosarcoma. No follow-up was published on one patient, another died with carcinoma of the breast having been found free of sarcoma 10 years after resection of her primary nerve tumor, and one patient is alive and well one year after diagnosis.

The most common presenting complaints were in reference to a rapidly or progressively growing tumor and/or pain. One patient (our case 2) presented with significant neurologic abnormalities. One or more stigma of von Recklinghausen's disease was found in eight patients, a finding that accords with the observation that malignant transformation occurs more often in neurilemmal sheath tumors associated with that disease than in solitary neurilemomas. In two patients, the nerve tumors appeared to be solitary.

Ages of the 10 patients ranged from 14 to 57 years with an average of 33 years. This corrsponds closer to the mean age of patients in the report by D'Agostino[8] et al. of malignant nerve sheath tumors associated with von Recklinghausen's disease (31 years) than for similar tumors developing in individuals free of this malady[7] (43 years).

The tumors were widely distributed anatomically; involvement of the neck, back, and extremities occurred on three occasions each. In one patient, the tumor arose from a spinal nerve root and spread along the spinal leptomeninges in a manner previously described for malignant schwannoma.[51] Gross involvement of a nerve was described in four cases. The size of the primary tumor was provided in four instances and ranged from approximately 1.5 to 12 cm in diameter. In all cases in which rhabdomyosarcoma appeared in the primary material, it arose from within the substance of the nerve tumor and in the primary material bore no connection to surrounding muscular structures.

Histologically, the rhabdomyosarcoma arose in the vicinity of malignant schwannian elements in four cases while in one case the Schwann cells among which it was first observed were clearly cytologically benign. It is of interest that in two instances sarcoma was only noticed in the recurrent tumor or in pulmonary metastases. The prevailing type of muscle tumor (eight cases) appeared to be a pleomorphic rhabdomyosarcoma, while two cases showed growth traits of embryonal rhabdomyosarcoma. Cross striations were observed in tumor cells from five of the patients. In the tumor from case 2, the spectrum of differentiating embryonal rhabdomyoblasts resembled that often displayed by embryonal rhabdomyosarcomas arising de novo in the head and neck[47] and other body sites.[10,18,20,37,39,46] In many areas of the infiltrating neoplasm, characteristic plump embryonal rhabdomyoblasts bearing stringy concentric fibrils in their cytoplasm were scattered among small undifferentiated malignant cells. Unequivocal cross striations when seen occurred chiefly along the periphery of neoplastic muscle cells showing tapered or ribbon configurations. We feel that Masson's diagnosis of rhabdomyoma of embryonic type rendered in one of his three cases reflected terminology then in vogue for embryonal rhabdomyosarcoma. Indeed, articles relating the metastatic potential of embryonal type muscle tumors had appeared in the German medical literature before and at the turn of the century[3,9,53] and were familiar to Ewing who, in his 1919 textbook,[11] classified muscle tumors containing embryonal cells among the rhabdomyomas.

Masson, who was the first to appreciate that rhabdomyosarcoma could develop within the substance of a peripheral nerve tumor,[24] offered two explanations for this phenomenon. Citing the experiments of Mlle, Locatelli,[23] in which she induced the growth of supernumerary limbs provided with bone and muscle in Tritons (small salamanders of the genus Triturus) by implantation of the cut end of the sciatic nerve into the soft tissues of their backs, he suggested that endoneurial cells of "neuromas" under the organizing influence of motor nerve fibers may be able to differentiate into muscle tissue. This explanation, however, has been weakened somewhat by both Singer's demonstration[44] that limb and muscle regeneration in Tritons is not dependent upon motor nerve innervation, and the work of

others[38,55] showing that aneurogenic forelimbs of young salamanders may regenerate. Neurilemmal sheath tumors containing rhabdomyosarcoma are referred to by some pathologists as malignant "Triton" tumors in recall of this early theory of formation.

Masson's second and more acceptable explanation[27] was that neoplastic Schwann cells can transform into striated muscle elements. In support of this theory, one can cite the many published accounts of neoplastic muscle cells developing in neuroepithelial-derived tissues. Most of these have appeared in association with medulloblastoma[15,19,21,30,35,41] (medullomyoblastomas or teratoid tumors), although rhabdomyosarcoma has also been described as an intimate component of intraocular medulloepitheliomas[56] and has appeared as a relatively pure growth in such related locations as the vermis, lumbosacral cord,[40] and iris of the eye.[34,54] In addition to striated muscle it would seem that, like neurilemmal cells, the medullary epithelium of the eye has the capacity to form chondroblastic tissue.[56] Our own finding in case 2 of embryonal rhabdomyoblasts which appeared to be arising directly from Schwann cells further supports Masson's theory of metaplasia.

Relevant to this are the published accounts[16,36] of what we presume to represent the benign variety of Triton tumor. Gratia, upon examining a fusiform mass arising along one of the vagus nerves of a horse discovered it to be comprised of intertwined striated muscle and "nerve" fibers.[16] At autopsy of a 68-year-old woman who died of bronchopneumonia, Orlandi found separate tumors involving the left sciatic and a popliteal nerve.[36] Microscopically, both tumors were confined by a perineural sheath and showed an intimate admixture of "nerve" fibers and fascicles of striated muscle.

In our own experiences, it is not uncommon to find rhabdomyosarcomas that contain spindle cell areas mimicking neurilemmal tissue or, on the other hand, peripheral nerve tumors in which some of the Schwann cells have acquired a modest incease in cytoplasmic mass and noticeable eosinophilia similar to that seen in rhabdomyoblasts. In determining whether such neoplasms truly classify as a malignant "Triton" tumor or not we in our laboratories look for evidence that the tumor:

1. Arose along the course of a peripheral nerve or in a patient with von Recklinghausen's disease or in a location typical for peripheral nerve tumors (e.g., skin and subcutaneous tissue), or represented a metastasis from such a tumor.

2. In large part showed the growth characteristics of Schwann cells.

3. Contained bonafied rhabdomyoblasts (some of which showed either longitudinal and concentric cytoplasmic fibrils or cross striations) that appeared to arise from within the body of the peripheral nerve tumor and which could not be attributed to either an extension or metastasis from an extrinsic rhabdomyosarcoma.

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