

Rhabdomyoblastic Differentiation in Head and Neck Malignancies Other Than Rhabdomyosarcoma

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Abstract Rhabdomyosarcoma is a relatively common soft tissue sarcoma that frequently affects children and adolescents and may involve the head and neck. Rhabdomyosarcoma is defined by skeletal muscle differentiation which can be suggested by routine histology and confirmed by immunohistochemistry for the skeletal muscle-specific markers myogenin or myoD1. At the same time, it must be remembered that when it comes to head and neck malignancies, skeletal muscle differentiation is not limited to rhabdomyosarcoma. A lack of awareness of this phenomenon could lead to misdiagnosis and, subsequently, inappropriate therapeutic interventions. This review focuses on *malignant* neoplasms of the head and neck other

than rhabdomyosarcoma that may exhibit rhabdomyoblastic differentiation, with an emphasis on strategies to resolve the diagnostic dilemmas these tumors may present. Axiomatically, no primary central nervous system tumors will be discussed.

Keywords Rhabdomyosarcoma · Rhabdomyoblastic differentiation · Myogenin · MyoD1 · Head and neck malignancies · Soft tissue sarcomas · Skeletal muscle differentiation

Introduction

Rhabdomyosarcoma (RMS) is a malignant mesenchymal neoplasm that exhibits skeletal muscle differentiation. RMS exists in two major forms: alveolar RMS which is a

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“small round cell tumor” that typically harbors $t(2;13)$ or $t(1;13)$ translocations resulting in *PAX3-FOXO1A* or *PAX7-FOXO1A* gene fusions; and embryonal RMS which is composed of round to spindle cells and harbors more complex genetic alterations (e.g., loss of the tumor suppressor *CDKN2A*, mutation/amplification of *FGFR4*, gain of *GLI1*, and mutations in the myogenic transcription factor *MYOD1*) [1–6] (Fig. 1). It is important to distinguish between the alveolar and embryonal types of RMS due to differing prognoses and treatment strategies [7, 8]. Other variants of RMS include sclerosing, spindled, and pleomorphic forms [5]. While RMS is typically encountered in children and young adults, it can also be seen in older adults, especially the alveolar subtype [5, 9, 10]. About 40 % of RMS affect the head and neck, in order of frequency: orbit, sinonasal tract, ear, and oral cavity, with other subsites rarely affected [5, 9, 11–14].

All forms of RMS are defined, at least in part, by the presence of rhabdomyoblasts—densely eosinophilic polygonal or spindled cells with hyperchromatic nuclei and occasional cytoplasmic cross-striations. While skeletal muscle differentiation can be suggested by histology and

desmin immunoreactivity, in the absence of clear-cut cross-striations it must be confirmed by nuclear immunohistochemical staining for myogenin and/or MyoD1, markers with high specificity for skeletal muscle differentiation [5, 15]. It must be remembered, however, that rhabdomyoblastic differentiation may be encountered in neoplasms other than RMS. This distinction is important because RMS is treated by specific chemotherapy protocols that may be different than those of other neoplasms in the differential diagnosis [8, 16]. This article reviews the malignant head and neck tumors other than RMS that may show rhabdomyoblastic differentiation, focusing on diagnostic strategies for distinguishing them from true RMS.

Malignant Triton Tumor

Malignant neoplasms arising in association with peripheral nerves or within pre-existing benign nerve sheath tumors (usually neurofibromas) are known as malignant peripheral nerve sheath tumors (MPNSTs) [17]. MPNSTs are uncommon, representing only 5–10 % of all sarcomas [17,

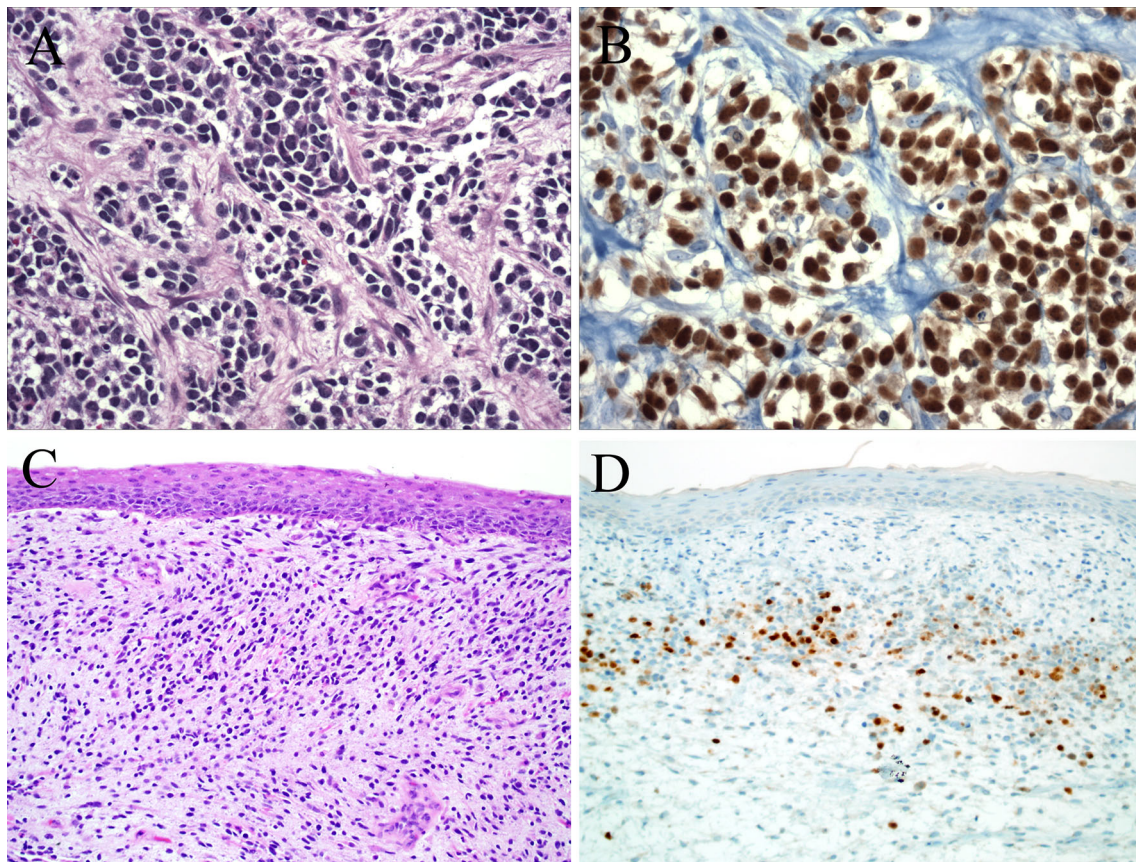


Fig. 1 Rhabdomyosarcoma (RMS). Alveolar RMS grows between fibrous septa as nests of dyscohesive small round cells with high nuclear-cytoplasmic ratios (a). In the alveolar form of RMS, myogenin immunoreactivity is diffuse (b). Embryonal RMS grows

as round to spindle cells, often condensing beneath epithelial surfaces in a “cambium layer” (c). Myogenin is also positive in embryonal RMS, but the distribution is less diffuse than what is seen in the alveolar subtype (d)

18]. MPNST typically arises in the deep soft tissue in adults, and may be sporadic or arise in the setting of neurofibromatosis [17]. The head and neck is one of the more common anatomic areas to be affected by MPNSTs [17]. MPNSTs typically grow in a herringbone-type fascicular pattern (“herringbone” refers to a repeating zigzag pattern where the fascicles are very regular and well defined). Classically, “dark” hypercellular areas alternate with “light,” less cellular ones, simulating fetal mesenchyme and leading to a so-called “marbled” appearance [17, 19]. MPNSTs are highly cellular and exhibits nuclear hyperchromasia, pleomorphism, elevated mitotic rates, and necrosis (Fig. 2). By immunohistochemistry, the nerve sheath markers S100 protein and SOX10 are often positive, but classically are focal in distribution [17, 20]. CD34 is variably expressed, and pancytokeratin and BCL2 are negative or at most, focal [21].

Up to 10–15 % of MPNSTs contain heterologous elements, the most frequent of which are rhabdomyoblastic foci [17, 22, 23]. Other reported heterologous elements include benign-appearing glands, islands of bone or cartilage, or angiosarcomatous foci [17, 22, 23]. MPNSTs with rhabdomyoblastic differentiation were first reported by Masson who considered the phenomenon as supporting the origin of these tumors from motor rather than sensory nerves [24]. They were dubbed malignant Triton tumors after early experiments in the Triton salamander in which dissection and ectopic re-implantation of the sciatic nerve was associated with formation of supernumerary ‘limbs’ containing skeletal muscle and bone [22–24]. The phenomenon is consistent with the capacity of neural-crest cells descendants to differentiate into both Schwann cells and various mesenchymal tissues [17, 25]. (As a brief aside, tumors referred to as “benign Triton tumors” are most likely hamartomas and are most likely unrelated.) [26] The majority of malignant Triton tumors occur in the setting of NF1, and as a result, the affected patient is typically young [22, 23]. About a third of malignant Triton tumors affect the head and neck where they can involve virtually any anatomic subsite [27–29]. The rhabdomyoblasts in malignant Triton tumors are typically focal, and they often stand out at low power as their abundant eosinophilic cytoplasm is distinctly different than the pale background Schwannian cells (Fig. 2). As expected, these cells (like the tumor cells of RMS) are positive for desmin and myogenin/MyoD1. Malignant Triton tumors are regarded to behave in an aggressive fashion, even more than usual MPNST, [22, 23, 30, 31] though head and neck cases described as “low-grade” have been reported [32, 33]. It has been suggested that tumors in the sinonasal tract have a more indolent course than those arising in other head and neck sites (however, at least some of those indolent “malignant Triton tumors” could in fact represent the newly-

described “low-grade sinonasal sarcoma with neural and myogenic features,” a tumor that lacks myogenin/MyoD1 expression) [34, 35].

Differentiating malignant triton tumor with conspicuous rhabdomyoblasts from an embryonal or spindle cell RMS may be difficult on routinely stained histological sections. The challenge is compounded by the fact that NF1 patients are at increased risk for RMS, and some RMSs may express S100 protein [17, 36]. In contrast to a true RMS, the rhabdomyoblasts in malignant Triton tumors tend to be a relatively focal finding in a background of predominant Schwannian cells that are completely negative for desmin and myogenin and/or MyoD1. If present, a pre-existing benign nerve sheath tumor strongly supports the diagnosis of malignant Triton tumor. If on the other hand, the clinical, histologic, and immunophenotypic picture is most compatible with an embryonal RMS, the finding of some S100 protein positive cells should not dissuade a pathologist from that diagnosis [17].

Sarcomatoid Carcinoma

Sarcomatoid carcinoma (i.e., spindle cell variant of squamous cell carcinoma) is a variant of head and neck carcinoma characterized by a prominent or even exclusive population of malignant spindle or pleomorphic cells [37–41]. Once thought to represent a collision tumor between separately arising carcinoma and sarcoma, sarcomatoid carcinoma has since been shown to be a carcinoma derived from squamous epithelium that shows divergent differentiation into cells with mesenchymal features due to epithelial-mesenchymal transition [42–45]. While most sarcomatoid carcinomas demonstrate a haphazard, non-specific growth pattern in the sarcomatoid component of the tumor, 7–15 % of cases exhibit histologically definable heterologous mesenchymal elements like bone, cartilage, and rarely, skeletal muscle [40, 46, 47]. Sarcomatoid carcinomas in the oral cavity and oropharynx appear to be more aggressive, while those of the larynx, and particularly the true vocal cord, have a more favorable prognosis [40, 47–49]. Not surprisingly, the polypoid tumors, regardless of location, are more easily resected and tend to have a better prognosis [40, 50].

While rhabdomyoblastic differentiation is not uncommon in sarcomatoid carcinomas in other organs (especially malignant mixed Müllerian tumors of the uterus), it is quite rare in the mucosa of the head and neck, with only rare reported cases [51–55]. Interestingly, all the reported cases and those seen in our consultation practices arose in the larynx and all were biphasic, with a conventional squamous cell carcinoma component admixed with focal malignant spindle cells exhibiting rhabdomyoblastic features by

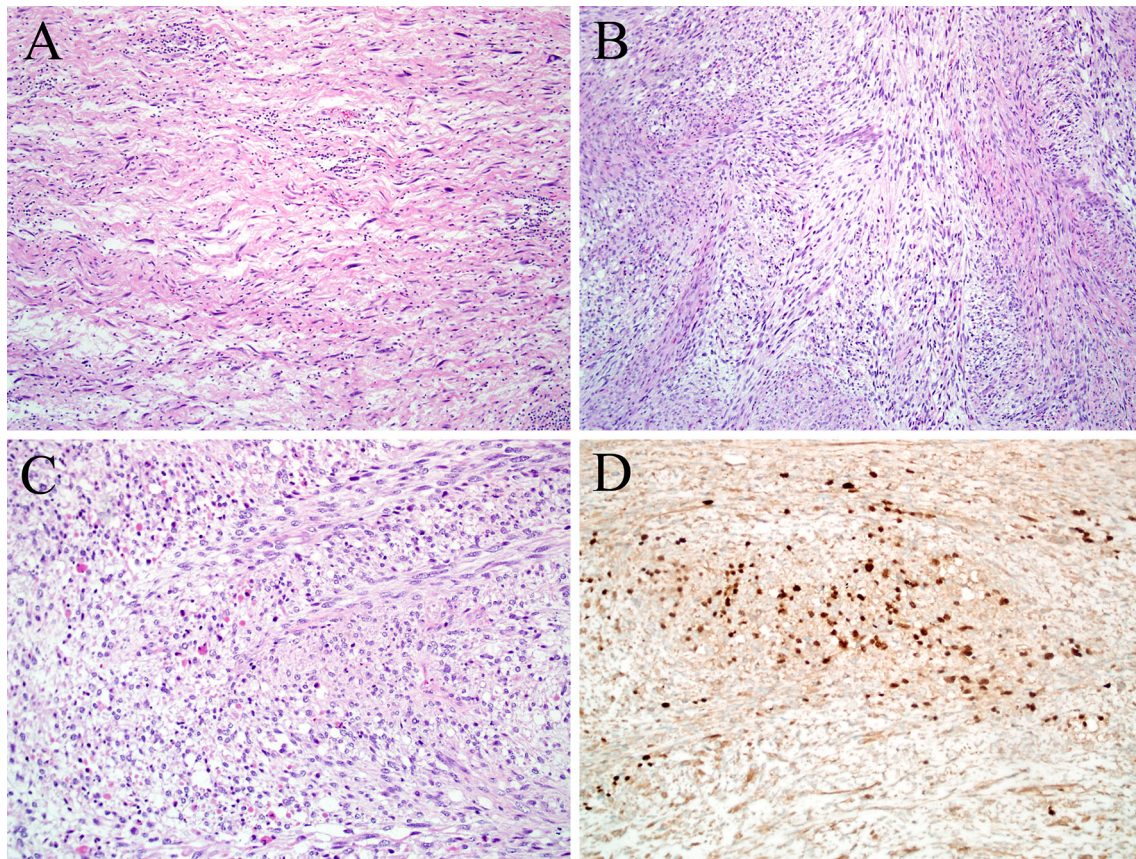


Fig. 2 Malignant Triton tumor. Malignant peripheral nerve sheath tumor often arises in the background of a benign nerve sheath tumor, usually neurofibroma (a). Malignant peripheral nerve sheath tumor often grows in a herringbone pattern of alternating fascicles. Often lighter staining areas alternate with *darker* areas creating a

“marbleized” appearance (b). Eosinophilic rhabdomyoblasts stand out in the background of the pale staining malignant peripheral nerve sheath tumor (c). The rhabdomyoblasts are highlighted by a myogenin immunostain (d)

routine histology and confirmed by immunohistochemistry (Fig. 3). In addition, one of these cases also showed neuroendocrine differentiation [54, 55].

The diagnosis of sarcomatoid carcinoma relies on finding epithelial differentiation by routine morphology (i.e. squamous dysplasia or a component of squamous cell or other type of obvious carcinoma mixed with sarcomatoid tumor) or, if this is absent, by the demonstration of epithelial differentiation by immunohistochemistry for epithelial markers. However it should be emphasized that true RMS may express cytokeratins in up to 7 % of cases [21]. Even the newer squamous marker p63 can be positive in RMS, although all cases reported so far have been negative for p40, the more squamous-specific isoform of p63 [56, 57]. Further complicating the distinction from RMS or other sarcomas is that up to a third of sarcomatoid carcinomas are monophasic spindle cell neoplasms, and the sarcomatoid components of up to 74 % of sarcomatoid carcinomas are completely negative for epithelial markers [40, 42, 46, 47, 58]. Ultimately, a diagnosis of sarcomatoid

carcinoma should be carefully considered before making the diagnosis of RMS in an older patient and in unusual mucosal locations like the larynx.

Undifferentiated (Anaplastic) Thyroid Carcinoma

Undifferentiated (anaplastic) thyroid carcinoma (UTC) is a highly aggressive malignant epithelial neoplasm of the thyroid. Morphologically the tumor grows in sheets of cells which are often spindled, pleomorphic/giant cell, or squamoid [59]. “Rhabdoid” cells have been identified in up to 10 % of UTC and one third of poorly differentiated thyroid carcinomas [60], however true skeletal muscle differentiation appears to be very rare in UTC with only 2 reported cases [61]. Distinguishing UTC from true sarcomas is aided immunohistochemically with cytokeratins or Pax-8, retained in about 75 % of UTC [62, 63]. Additionally, identification of an associated well-differentiated thyroid carcinoma, present in 30–70 % of UTC, supports

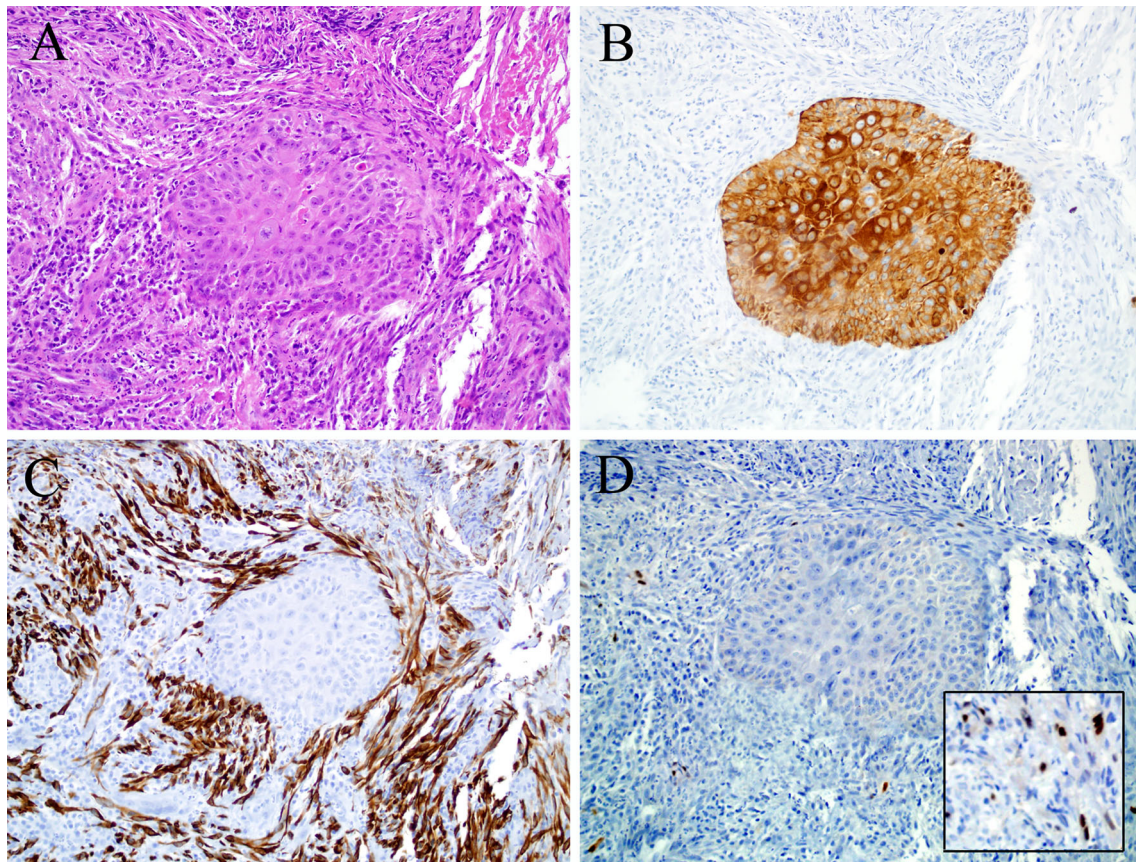


Fig. 3 Sarcomatoid carcinoma. This laryngeal tumor demonstrates both epithelial differentiation in the form of invasive squamous cell carcinoma (*center*) as well as mesenchymal differentiation (**a**). Only the squamous nest is positive for cyokeratin immunohistochemistry

(**b**), while the remaining spindle cell tumor component is positive for desmin (**c**) and myogenin (**d**), features diagnostic of rhabdomyoblastic differentiation

dedifferentiation from a primary thyroid malignancy [59]. If skeletal muscle markers are present by immunohistochemical evaluation, focal or patchy distribution supports heterologous differentiation of UTC over a rare example of true RMS involving the thyroid gland [64, 65]. Moreover, the *BRAF* V600E mutation may also be identified in 20–30 % of UTC and can be utilized in select cases for supporting evidence toward thyroid [66, 67].

Salivary Carcinosarcoma (“True” Malignant Mixed Tumor)

Pleomorphic adenoma is the most common neoplasm (benign or malignant) of the salivary glands [68]. Occasionally, malignancies can arise within pleomorphic adenomas, and the most common form of this phenomenon is carcinoma ex-pleomorphic adenoma, where the malignant tumor that develops is a carcinoma [69]. Rare examples of the mesenchymal component also exhibiting malignant transformation are known as carcinosarcoma or so-called

“true” malignant mixed tumor (the modifier “true” being applied in order to distinguish such tumors from carcinoma ex-pleomorphic adenoma and from benign metastasizing mixed tumor/pleomorphic adenoma) [70]. The most common type of mesenchymal malignancy in carcinosarcoma is chondrosarcoma, but other types include osteosarcoma, leiomyosarcoma, fibrosarcoma, and very rarely, RMS [71–75]. This finding is more of a curiosity than a true diagnostic challenge, because by definition, carcinosarcoma harbors a malignant epithelial component that will clue the observer into the correct diagnosis. The carcinomatous components in these cases of carcinosarcoma with rhabdomyoblastic differentiation have been adenocarcinoma not otherwise specified, salivary duct carcinoma, squamous cell carcinoma, large cell neuroendocrine carcinoma, and undifferentiated carcinoma [71–75]. Moreover, the focal rhabdomyosarcomatous components in all but one case were accompanied by other sarcomatous components which included sarcoma not otherwise specified, myxofibrosarcoma, chondrosarcoma, liposarcoma, and fibrosarcoma [71–75]. Four cases also featured a component of

residual benign pleomorphic adenoma [72–75]. The prognosis of carcinosarcoma, regardless of the component malignancies, is poor [70].

Olfactory Neuroblastoma (Esthesioneuroblastoma)

Olfactory neuroblastoma (ONB) is a malignant neoplasm that arises from the olfactory neuroepithelium of the superior nasal cavity and ethmoid sinus. The microscopic appearance of ONB depends on its grade. Low grade tumors have a lobular growth pattern, neurofibrillary matrix material, Homer Wright pseudorosettes, and a uniform population of round tumor cells with high nuclear-cytoplasmic ratios [76]. High grade ONBs show less of the lobular architecture and do not exhibit neurofibrillary matrix or Homer Wright pseudorosettes. High-grade ONBs instead have significant pleomorphism, high mitotic rates, and necrosis. They also may demonstrate true (Flexner-Wintersteiner) rosettes [76, 77]. By immunohistochemistry, ONB is positive for the neuroendocrine markers synaptophysin, chromogranin, and CD56 while S100 protein highlights the sustentacular supporting cells at the periphery of the tumor nests [76, 78]. Classically, other similar-appearing small round cell tumors such as lymphoma, melanoma, Ewing sarcoma/primitive neuroectodermal tumor, and RMS can be excluded because classic ONB shows a lobulated rather than diffuse growth pattern and is negative for CD45, CD99, desmin, myogenin, and HMB-45. Cytokeratins, while usually negative, may be focally positive in ONB, but EMA is consistently negative [78].

Occasionally, ONB may exhibit unusual forms of differentiation that may obscure the diagnosis. Most common is epithelial differentiation, where ONB may show cytokeratin and EMA immunopositivity. While this form of cytokeratin expression is typically focal, very rare examples of ONB may show foci of strong immunostaining and even glandular structures (though it is debatable whether these rare examples should be classified as another tumor type like neuroendocrine carcinoma or “olfactory carcinoma”) [79, 80]. Rarely, ONB may exhibit melanocytic or rhabdomyoblastic differentiation [81–83] (Fig. 4). Reports of this phenomenon are extremely limited, and as a result, the frequency and significance of this divergent differentiation cannot be determined (Table 1).

It is important to distinguish ONB with rhabdomyoblastic differentiation from RMS, a problem compounded by both tumors being encountered in the sinonasal tract of young patients. Additionally, the alveolar subtype of RMS may express neuroendocrine markers, chromogranin and synaptophysin, in 20–30 % of tumors with nearly universal

CD56 expression [21]. An important key to this dilemma is recognizing the nature of the myogenin expression. Alveolar RMS, a nested, small round cell tumor, generally demonstrates diffuse myogenin expression (Fig. 1), in contrast to the patchy distribution seen in ONB [15, 84] (Fig. 4). Moreover, even in the face of patchy rhabdomyoblastic differentiation, classic areas of ONB should be recognizable in the background. Finally, in a very difficult case, molecular studies for the t(2;13) or t(1;13) translocations of alveolar RMS may be useful. ONB are always translocation-negative and have complex cytogenetic alterations with deletions of 3p and overrepresentations of 17q in up to 100 % of cases [85].

Teratocarcinosarcoma

Teratocarcinosarcoma is a rare, peculiar sinonasal malignancy that features an admixture of epithelial, neuroectodermal/neuroendocrine, and mesenchymal elements showing varying degrees of maturation and cellular atypia (Fig. 5). The epithelial component may be either squamous or glandular, often with a cytologically bland, “fetal” appearance, reminiscent of what may be encountered in a teratoma [86–88]. The neuroectodermal/neuroendocrine tumor component is typically high-grade and primitive in its appearance, sometimes with rosette structures and/or neurofibrillary matrix. The tumor is set in a mesenchymal stroma that may be bland or overtly sarcomatous. This stroma can exhibit cartilaginous, smooth muscle, or skeletal muscle differentiation [86–88]. Immunostaining for skeletal muscle markers can facilitate the detection of small foci with crowding of rhabdomyoblasts in which cytoplasmic cross-striation may be easily missed in the initial evaluation with routine hematoxylin and eosin staining [89]. Components of seminoma, choriocarcinoma, and embryonal carcinoma are, by definition, not found. In fact, although teratocarcinosarcoma might suggest a germ cell tumor with teratomatous elements, it more likely arises from stem/progenitor cells of the neuroepithelium.

Because of its classically variable histologic appearance it is notoriously difficult to recognize teratocarcinosarcoma on biopsy [86–88]. Depending on which areas are sampled, teratocarcinosarcoma can be mistaken for ONB, chondrosarcoma, squamous cell carcinoma, adenocarcinoma, or even RMS. Distinguishing teratocarcinosarcoma from these other tumors relies primarily on adequate sampling to reveal the other tumor components and thus, the true tumor identity. Teratocarcinosarcoma is an aggressive neoplasm, though newer studies have shown that it is not as lethal as early reports had suggested. The mean survival is approximately 6 years [86–88, 90–92].

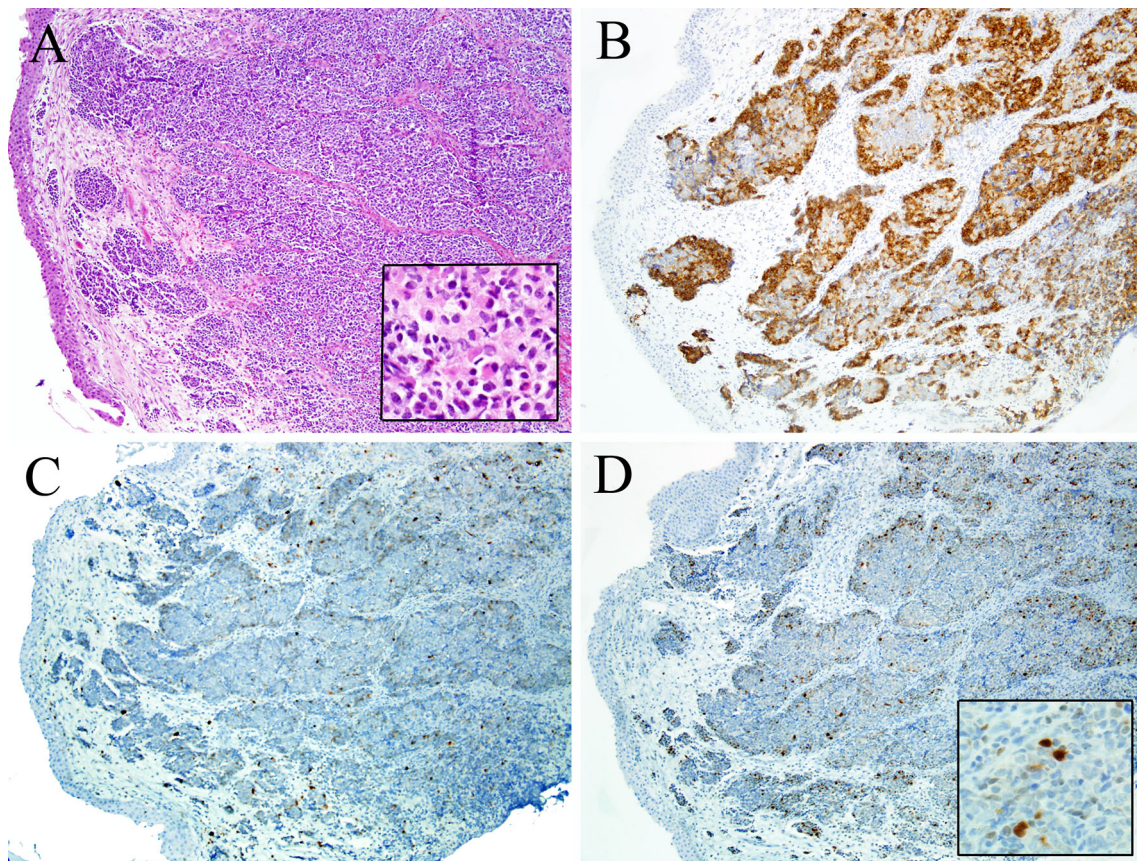


Fig. 4 Olfactory neuroblastoma. This example of olfactory neuroblastoma grows in the typical fashion, as nests in the sinonasal submucosa (a). At high power, rhabdoid cells with abundant eccentric cytoplasm are evident (*inset* of a). As expected, this tumor was

diffusely positive for synaptophysin (b) and had a peripheral (i.e., sustentacular) pattern of S100 immunostaining (c). The rhabdoid cells seen in areas of the tumor are confirmed to be rhabdomyoblasts by myogenin immunohistochemistry (d)

Table 1 Head and neck malignancies that may exhibit rhabdomyoblastic differentiation

Rhabdomyosarcoma
Malignant peripheral nerve sheath tumor (malignant Triton tumor)
Sarcomatoid carcinoma (spindle cell variant of squamous cell carcinoma)
Undifferentiated (anaplastic) thyroid carcinoma
Carcinosarcoma of the salivary glands
Olfactory neuroblastoma (esthesioneuroblastoma)
Teratocarcinosarcoma
Malignant teratoma
Melanoma
Liposarcoma

Malignant Teratoma

Teratoma is a neoplasm that consists of tumor differentiating into cells of all three embryonal germ cell layers (ectoderm, mesoderm, endoderm). Fewer than 5 % of teratomas arise in the head and neck, where the cervical soft tissue and nasopharynx are the most commonly affected sites [87]. Most teratomas occur in children, and many are detected in the perinatal period; they are rare in adults. The

composition of teratomas is quite variable, and may include both mature and immature tissues of all three germ cell layers. It is important to remember that teratomas in children are uniformly benign, even when immature elements are present, although they may cause morbidity due to airway obstruction. On the other hand, when teratomas arise in the head and neck of an adult patient (typically in the cervical soft tissues and/or thyroid gland) their behavior does depend on the presence or absence of immature tissue

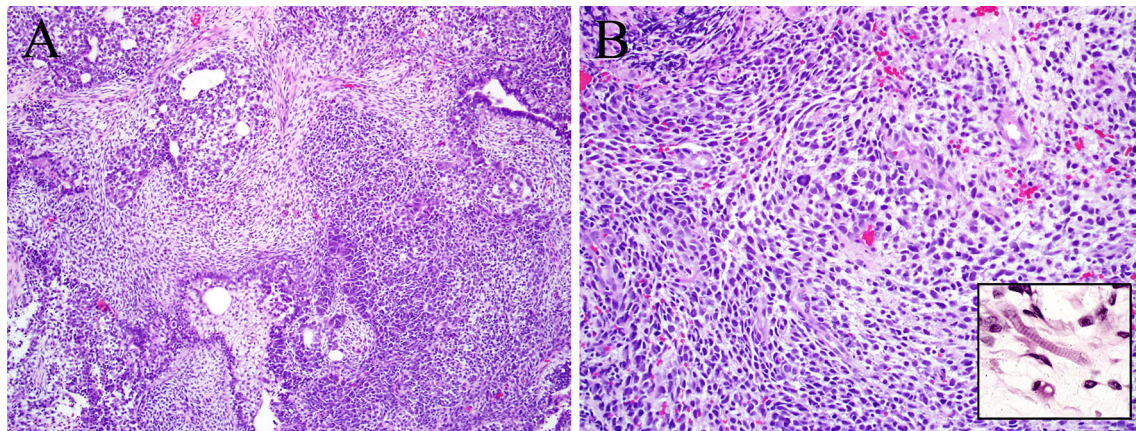


Fig. 5 Teratocarcinoma. Sinonasal teratocarcinoma exhibits admixed zones of primitive small round cells, spindled cells, squamous epithelium with clear cytoplasm, and glands (a). Primitive and/or spindled components of teratocarcinoma demonstrate

rhabdomyoblastic differentiation (b) which is strongly suggested by cytoplasmic cross-striations (*inset*) and is confirmed by immunohistochemistry for myogenin or MyoD1

elements. Unfortunately, most adult teratomas in the head and neck contain immature elements and they usually behave in an aggressive manner [93–96]. While focal, immature skeletal muscle may resemble RMS in isolation, it is unlikely to cause diagnostic difficulty when the other elements of a teratoma are present, which is almost invariably the case with malignant teratomas.

Melanoma

Rare examples of melanoma may show heterologous mesenchymal elements, including Schwannian, ganglionic, cartilaginous or osteoid differentiation [97]. In this context, it is not surprising that rare melanomas, including some arising in the head and neck region, have also exhibited focal rhabdomyoblastic differentiation [98–100]. The rhabdomyoblasts in these cases are positive for myogenin/MyoD1 but are negative for melanocytic markers [98–102]. These cases are very rare and it would be imprudent to attempt to draw any conclusions about the significance of the rhabdomyoblasts. Nevertheless, if abundant, they can be a diagnostic pitfall for the unwary. Clues to the diagnosis of melanoma include a cutaneous location (although melanomas can certainly arise from mucosal sites), older patient age, an overlying *in situ* melanoma within the epidermis/surface epithelium, and a more conventional melanoma component with prominent eosinophilic nucleoli, immunoreactivity for S100 and specific melanocytic markers like HMB45, Melan-A, and SOX-10, and in some cases, melanin pigment. It has been repeatedly emphasized that melanoma is incredibly variable in its appearance and as a result, diagnostic pathologists should have a very low threshold for keeping it in the differential diagnosis of any poorly differentiated head and neck tumor.

Liposarcoma

Liposarcoma is a malignant neoplasm of the soft tissues that demonstrates fatty differentiation. Liposarcomas are most common in the retroperitoneum and extremities, but may occasionally arise in the head and neck. Rare examples of liposarcoma have exhibited focal rhabdomyoblastic differentiation. Most of these were in the context of a liposarcoma undergoing de-differentiation/high-grade transformation, although rhabdomyoblasts have rarely been encountered in primary well-differentiated or myxoid liposarcomas, including two arising in the head and neck [103–108]. Again, the significance, if any, of this finding is unclear; and the rhabdomyoblasts have been merely a peculiar, focal finding in tumors that were otherwise straightforward liposarcomas, and did not pose a considerable diagnostic challenge. RMS lacks fatty differentiation.

Others

It should be noted that there is a handful of additional head and neck neoplasms such as de-differentiated chordoma [109], gnathic osteosarcoma [110], Merkel cell carcinoma [111], and small cell neuroendocrine carcinoma [112] where a single case with confirmed rhabdomyoblastic differentiation has been reported. It is difficult to draw any conclusions from these cases other than the idea that rhabdomyoblastic differentiation can be unexpectedly be encountered in a great variety of tumor types. In addition, there are a number of other head and neck neoplasms with rare reported examples of “rhabdomyoblastic” differentiation as defined hematoxylin and eosin and/or desmin immunohistochemistry, but not confirmed by myogenin and/or MyoD1 immunostaining. These include

melanocytic neuroectodermal tumor of infancy [113] and the newly described low-grade sinonasal sarcoma with neural and myogenic features [35].

Conclusions

RMS frequently affects the head and neck but as we have demonstrated in this review, it is certainly not the only head and neck neoplasm that may exhibit rhabdomyoblastic differentiation. For malignant Triton tumor, An awareness of this phenomenon as well as attention to background tumor elements (i.e., away from the rhabdomyoblasts) and anatomic and demographic considerations (e.g., undifferentiated carcinoma is much more likely than RMS in the thyroid gland of an elderly patient) will help prevent misdiagnosis in most instances. Judicious use of molecular diagnostic tools can be helpful in select cases. When aberrant skeletal muscle differentiation is encountered, it is prudent to mention its presence in the diagnosis, along with a note clarifying that the neoplasm is not RMS.

Sample Diagnosis

NASAL CAVITY (BIOPSY): OLFACTORY NEUROBLASTOMA WITH FOCAL RHABDOMYOBLASTIC DIFFERENTIATION. SEE COMMENT.

COMMENT: The rhabdomyoblastic component is limited to a few scattered cells positive for desmin and myogenin, representing less than 5% of the tumor volume. This is not a rhabdomyosarcoma.

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