

Gliosarcoma with alveolar rhabdomyosarcoma-like component: Report of a case with a hitherto undescribed sarcomatous component

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SUMMARY

Gliosarcoma (GS) is a relatively rare glioblastoma variant characterized by biphasic glial and mesenchymal differentiation patterns. The sarcomatous part most commonly resembles fibrosarcoma or so-called malignant fibrous histiocytoma. Rarely, GS shows heterologous lines of differentiation in the form of osteosarcoma, chondrosarcoma, liposarcoma, leiomyosarcoma, squamous or glandular malignant epithelial differentiation, or primitive neuroectodermal tumor (PNET)-like foci. When rhabdomyoblastic differentiation occurs, it is in the form of malignant spindle cells, with cross-striated strap cells or rounded rhabdomyoblasts reminiscent of the embryonal type of rhabdomyosarcoma. We are reporting a case of GS with an alveolar rhabdomyosarcoma-like component. The tumor consisted of poorly differentiated primitive small round cells growing in a solid and alveolar pattern, with minimal cytoplasm, markedly elevated mitotic activity and numerous apoptotic nuclei. Rhabdomyosarcomatous differentiation was confirmed by desmin and myogenin immunopositivity. To the best of our knowledge, this histologic pattern has not been previously reported in GS. Differential diagnostic considerations are discussed.

Keywords: gliosarcoma – alveolar rhabdomyosarcoma – myogenin – desmin

Gliosarkóm s komponentou pripomínajúcou alveolárny rbdomyosarkóm: popis prípadu s doposiaľ nepopísanou sarkómovou zložkou

SÚHRN

Gliosarkóm (GS) je relatívne vzácny variant glioblastómu, charakterizovaný bifázickou gliálnou a mezenchymálnou diferenciáciou. Sarkomatózna časť najčastejšie pripomína fibrosarkóm alebo tzv. malígny fibrózny histiocytóm. Vzácné je v GS prítomná heterológna diferenciácia vo forme osteosarkómu, chondrosarkómu, liposarkómu, leiomyosarkómu, skvamóznej alebo žľazovej malígnej epiteliálnej diferenciácie, alebo diferenciácie pripomínajúcej primitívny neuroektodermálny tumor (PNET). Keď je v GS prítomná rbdomyosarkómová diferenciácia, je vo forme malígnych vretenovitých buniek s priechne pruhovanými bunkami alebo okrúhlymi rbdomyoblastami, pripomínajúca embryonálny rbdomyosarkóm. V kazuistike popisujeme GS s komponentou pripomínajúcou alveolárny rbdomyosarkóm. Nádor rástol v solídnych a alveolárnych formáciách a bol zložený z nediferencovaných primitívnych malých okrúhlych buniek s minimálnou cytoplazmou, nápadne zvýšenou mitotickou aktivitou a početnými apoptózami. Rbdomyosarkomatózna diferenciácia bola potvrdená pozitívnu imunohistochemickou reakciou na dezmin a myogenín. Podľa našich vedomostí, takýto histologický vzor nebol v GS doposiaľ popísaný. V krátkosti je prebraná diferenciálna diagnóza prípadu.

Kľúčové slová: gliosarkóm – alveolárny rbdomyosarkóm – myogenín - dezmin

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According to the 2007 WHO classification of tumors of the central nervous system („blue book“), gliosarcoma (GS) is „a glioblastoma variant characterized by a biphasic tissue pattern with alternating areas displaying glial and mesenchymal differentiation“. These tumors are rare, and represent 2 to 8 % of all glioblastomas (GBM) (1). Gliosarcomas typically occur in old-

er population, but exceptional cases can be found in young adults, and even in children and infants (2). The glial part is a high grade astrocytic neoplasm, mostly resembling classical GBM, whereas the sarcomatous part most commonly resembles fibrosarcoma or so-called malignant fibrous histiocytoma (1). Rarely, GS may show heterologous lines of differentiation in the form of osteosarcoma (3), chondrosarcoma (4), liposarcoma (5), leiomyosarcoma (6), squamous or glandular malignant epithelial differentiation (7), or primitive neuroectodermal tumor (PNET)-like foci (8).

Rhabdomyoblastic differentiation in GS was first described in the English literature by Goldman in 1969 (9), and after that by a few other authors (10–13). Rhabdomyosarcomatous differentiation in these cases was in the form of malignant spindle cells, with typical elongate strap cells or rounded rbdomy-

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