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Cytomorphologic Features of Atypical Teratoid/Rhabdoid Tumors

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Background:
Atypical teratoid/rhabdoid tumor (AT/RT) is a highly malignant tumor of the central nervous system, with majority of cases seen in children under five years of age. Although the clinicopathologic and radiologic features of AT/RT has been extensively described, the cytomorphological features of ATRT have not been well studied.

Material and Methods:
Cytologic material, consisting of one to two smears from 6 cases of AT/RT, was studied from the files of The Johns Hopkins Hospital. Slides made from surgical specimens by either scraping and smearing (SS) or squash preparation (SP), were ethanol fixed and stained with H&E. One of the cases, also had CSF specimen. The corresponding surgical pathology material from these cases was also reviewed.

Results:
The patients were 3 males and 2 females with an age range of 1 to 16 years (mean 5.8 years). The examined smears were hypercellular with many single discohesive cells. Of the cases examined, some features were more prominent including the presence of larger round plasmacytoid/rhabdoid cells with fine granular eosinophilic cytoplasm, large eccentrically located nuclei and single prominent nucleoli. There were occasional large bizarre cells. A prominent small cell component was also present in some cases. There was extensive nuclear crush artifact, apoptic bodies, mitosis and more rarely, focal areas of necrosis. Rosettes were not present. In some cases, dystrophic calcification was present. In some of the smears, there was extensive inflammation.

Conclusions:
The differential diagnosis of AT/RT includes medulloblastoma/primitive neuroectodermal tumor (PNET) of the brain. Because a typical case of AT/RT may have areas resembling PNET, the following cytomorphological features may be helpful in making the diagnosis of AT/RT: 1)Presence of large rhabdoid cells with pink cytoplasm and eccentric nuclei. (2) Many single discohesive rhabdoid/plasmoacytoid cells admixed with more PNET like areas, (3) abundant mitosis and apoptosis and (4) inflammation and/or calcification in the background. Cytological examination of CSF and detection of characteristic rhabdoid cells may have a role in the diagnosis, monitoring disease progression and making therapeutic decisions in management of patients with AT/RT.