Adult sellar atypical teratoid rhabdoid tumor

Its occurrence in the sellar region is rare, with only 11 cases reported in the literature till 2017 [1)].

Adult sellar atypical teratoid rhabdoid tumor (ATRT) is a rare diagnosis that has recently been shown to be a clinicopathologically and genetically distinct variant of ATRT occurring almost exclusively in middle-aged females. While up to one third of pediatric ATRT is caused by a familial syndrome, no previous cases of a familial adult sellar ATRT have been reported.

Case series

Paolini et al., described detailed clinical and genetic characterization of 5 adult patients with AT/RTs involving the sellar and suprasellar region, and provide a review of the available clinical and genetic features of 22 previously reported cases in order to help increase our understanding of this unusual entity [2)].

Case reports

Voisin et al., present the first case report of a familial germline mutation causing adult sellar ATRT and a literature review of 29 previously published cases of sporadic adult sellar ATRT.

A 51-year-old woman with a family history of brain tumors spanning three generations presented with visual decline and was diagnosed with an adult sellar ATRT. Genetic studies demonstrated a heterozygous splice-site loss-of-function mutation of the INI1 gene in exon 7. Treatment included endoscopic endonasal biopsy, craniospinal irradiation and focal tumor boost, followed by adjuvant chemotherapy.

This is the first case report of a familial germline mutation causing adult sellar ATRT [3)].

A case of 54 year-old female, which presented with symptoms concurrent of a pituitary macroadenoma, which was found out to be a tumor of ATRT characteristics on pathology [4)]

Almalki reported a 36-year-old female patient. She was treated with multi-modalities including surgery, chemotherapy and radiation. She markedly improved following treatment with no recurrence in 3 years follow-up. This was the 11th case of an adult-onset AT/RT in the sellar or suprasellar region with favorable long-term outcome [5)].

[1)], [5)]
Almalki MH, Alrogi A, Al-Rabie A, Al-Dandan S, Altwairgi A, Orz Y. Atypical Teratoid/Rhabdoid Tumor of the Sellar Region in an Adult With Long Survival: Case Report and Review of the Literature. J Clin Med Res. 2017 Mar;9(3):216-220. doi: 10.14740/jocmr2922w. Epub 2017 Jan 25. PubMed PMID: 28179970; PubMed Central PMCID: PMC5289142.

2)

Paolini MA, Kipp BR, Sukov WR, Jenkins SM, Barr Fritcher EG, Aranda D, SantaCruz KS, Al-Dandan S, Fisher P, McDonald WC, Bondurant CP, Van Dyke Darkow G, Giannini C, Parisi JE, Jentoft ME, Raghunathan A. Sellar Region Atypical Teratoid/Rhabdoid Tumors in Adults: Clinicopathological Characterization of Five Cases and Review of the Literature. J Neuropathol Exp Neurol. 2018 Dec 1;77(12):1115-1121. doi: 10.1093/jnen/nly091. PubMed PMID: 30295777.

3)

Voisin MR, Ovenden C, Tsang DS, Gupta AA, Huang A, Gao AF, Diamandis P, Almeida JP, Gentili F. Atypical teratoid/rhabdoid sellar tumor in an adult with a familial history of a germline SMARCB1 mutation: case report and review of the literature. World Neurosurg. 2019 Apr 17. pii: S1878-8750(19)31075-7. doi: 10.1016/j.wneu.2019.04.083. [Epub ahead of print] PubMed PMID: 31004861.

4)

D Barsky, U Hadelsberg, L Gonen, N Margalit, P05.91 Sellar atypical teratoid rhabdoid tumor (ATRT) in an adult: A case report and review of the literature, Neuro-Oncology, Volume 20, Issue suppl_3, September 2018, Pages iii324–iii325, https://doi.org/10.1093/neuonc/noy139.417