Opis choroby *

Definicja

A rare ependymal tumor characterized by the presence of a RELA fusion gene. This supratentorial grade II or III ependymoma most often occurs in children and young adults. Histopathological features are variable, but a distinctive vascular pattern of branching capillaries or clear-cell change are common. Patients may present with focal neurological deficits, seizures, or features of raised intracranial pressure. Prognosis is worse than in other supratentorial ependymomas.

Dane

Klasyfikacja
Choroba

Synonimy
Supratentorial C11ORF95-RELA fused ependymoma
Supratentorial C11ORF95-RELA fused ependymoma

Kod ORPHA
530792

Kod OMIM
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Kod ICD10
D43.0

Kod ICD11
-

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*Źródło

orphanet