Rosette forming glioneuronal tumor

Rosette forming glioneuronal tumor is a neuronal and mixed neuronal glial tumor.

They are midline tumours first described in 1998 by Komori et al., that involve the fourth ventricle or aqueduct of Sylvius.

This rare tumor is a distinct nosological entity of the glioneuronal family.

Although relatively well circumscribed on MRI and clinically extremely indolent, they often invade surrounding tissues, involving the cerebellum, pons and even the pineal region. There are often cystic components and they tend to have heterogenous contrast enhancement. They are WHO grade I lesions.

Clinical presentation

It is a tumour of middle age adults with mean age of 30 years. Symptoms mainly depends on the location of the tumour; commonly presenting with ataxia and headache.

Pathology

Histological features superficially resemble DNETs and contain both neurocytic and astrocytic components.

Location RGNTs are originally described in the region of 4th ventricle (59%) often with variable local parenchymal extension 3. Recent literature review have demonstrated RGNTs outside the characteristic location. RGNTs has been described in locations such as the cerebellum hemisphere, cerebello-pontine angle, pineal gland, tectum, thalamus, third ventricle, optic chiasm and spinal cord 3.

Radiographic features

MRI variable solid-cystic components:

RGNTs appear as solid lesions in 40%

mixed solid and cystic changes are seen in 35% cases

cystic only features in 25%

the majority of RGNT (70%) show variable gadolinium enhancement:

focal enhancement pattern was most commonly observed (50%)

heterogeneous pattern (19%)

minimal enhancement (13%)

ring and nodular enhancement pattern (9%).

calcification (25%)

Case series

A retrospective study includes seven RGNT patients (4 male, 3 female; age range: 7-42 years; mean age: 25 years) diagnosed and treated at our institute. MR studies were performed on 3 T and 1.5-T clinical MR systems. All patients were reviewed by two experienced neuroradiologists and imaging findings were tabulated.

Five tumours were located in the posterior fossa, and two were in the pineal region. One of the tumours demonstrated multiple satellite lesions, which involved the midbrain, pons, medulla as well as the cervical cord. Tumours located in the pineal region compressed the 3rd ventricle/aqueduct and extended below the tentorium cerebelli. All the tumours demonstrated enhancement, and susceptibility was evident in six of the seven patients. CSF dissemination was present in two patients.

RGNTs are usually solid-cystic tumours and frequently demonstrate peripheral/heterogeneous enhancement upon post-contrast study. Haemorrhage is a common feature which may not be evident on CT. Cerebrospinal fluid (CSF) dissemination is a feature and appropriate imaging should be performed whenever an RGNT is suspected. KEY POINTS: • CT and MRI findings of seven RGNT cases were retrospectively reviewed. • RGNTs are predominantly posterior fossa tumours. • RGNTs are typically T1 hypointense and T2 hyperintense. • Haemorrhage and peripheral/heterogeneous enhancement are common features of RGNTs. • CSF dissemination is a feature of RGNTs and requires appropriate imaging 1).

Medhi G, Prasad C, Saini J, Pendharkar H, Bhat MD, Pandey P, Muthane Y. Imaging features of rosetteforming glioneuronal tumours (RGNTs): A Series of seven cases. Eur Radiol. 2015 May 28. [Epub ahead of print] PubMed PMID: 26017735.

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