Protoplasmic astrocytoma

Two diffuse astrocytoma variants have been deleted from the World Health Organization Classification of Tumors of the Central Nervous System 2016: protoplasmic astrocytoma, a diagnosis that was previously defined in only vague terms and is almost never made any longer given that tumors with this histological appearance are typically characterized as other more narrowly defined lesions; and fibrillary astrocytoma, since this diagnosis overlaps nearly entirely with the standard diffuse astrocytoma. As a result, only gemistocytic astrocytoma remains as a distinct variant of diffuse astrocytoma IDH-mutant.

Protoplasmic astrocytomas were defined as rare variant of diffuse low grade astrocytoma with histological and imaging features which are fairly characteristic. It has been suggested that protoplasmic astrocytomas represent variants of dysembryoplastic neuroepithelial tumours (DNET) as they share histological as well as imaging features. Currently however they are classified as a subtype of diffuse low grade astrocytoma. Epidemiology

Typically patients diagnosed with low grade infiltrative astrocytomas are young adults (mean 32 years of age)

A male predilection is described (M:F ~5:3)

Clinical presentation

The most common presenting feature (~30-50% of cases) is seizures. This is particularly the case in adults. Headaches are often also present. Depending on the size of the lesion and its location other features may be present, e.g. hydrocephalus, focal neurological dysfunction including personality change. Pathology

Protoplastmic astrocytomas, along with other variants of diffuse low grade astrocytomas are considered WHO grade II tumours (see grading of diffuse low grade astrocytomas).

These tumours are composed of neoplastic astrocytes with rounded prominent nuclear contour and little cytoplasm. They have scant processess. The tumour matrix contains numerous and prominent microcystic spaces filled with mucinous fluid 3.

Mitoses, microvascular proliferation and necrosis are absent (if present they suggest a high grade tumour). Like all tumours derived from astrocytes, fibrillary astrocytomas stain with glial fibrillary acidic protein (gFAP) 2. Radiographic features

MRI is the modality of choice for characterising these lesions. These tumours appear to have a predilection for the frontal and temporal lobes CT

Typically protoplasmic low grade infiltrating astrocytomas appear as hypodense regions of positive mass effect, usually without any enhancement (in fact presence of enhancement would suggest high grade (e.g. WHO III or IV) tumours). Areas of the tumour appear of fluid attenuation, due to the aforementioned prominent mucinous microcystic component. MRI

These tumours have fairly characteristic appearances

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T1: hypointense compared to white matter
T2: strikingly hyperintense
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  FLAIR: large areas of T2 hyperintensity suppress on FLAIR (these are not
macrocystic but rather represent the areas with abundant microcystic change)
  T1 C+ (Gd): usually little or no enhancement
  MR spectroscopy: elevated choline:creatine ratio
  MR perfusion: there is reduced rCBV
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The key features which should prompt a protoplasmic astrocytoma being raised as the favoured diagnosis is A) prominent involvement of cortex B) large portions of the tumour demonstrating high T2 signal which suppresses on FLAIR. Treatment and prognosis

These tumours, along with with the more common fibrillary astrocytoma, tend to be relatively indolent. Treatment depends on clinical presentation, size of the tumour and location. In general the options are:

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  observe
  biopsy to confirm diagnosis and observe
  resection
  radiotherapy
  chemotherapy may have a role in recurrent/de-differentiated tumours
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Differential diagnosis

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  fibrillary astrocytoma:
      absence of FLAIR suppressing T2 high signal components
  dysembryoplastic neuroepithelial tumours (DNET):
      many similarities on imaging and histology
      smaller
      more purely cortical involvement
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http://radiopaedia.org/articles/protoplasmic-astrocytoma