PRIMARY ANGIITIS OF THE CENTRAL NERVOUS SYSTEM PRESENTING AS A TUMOR-LIKE LESION

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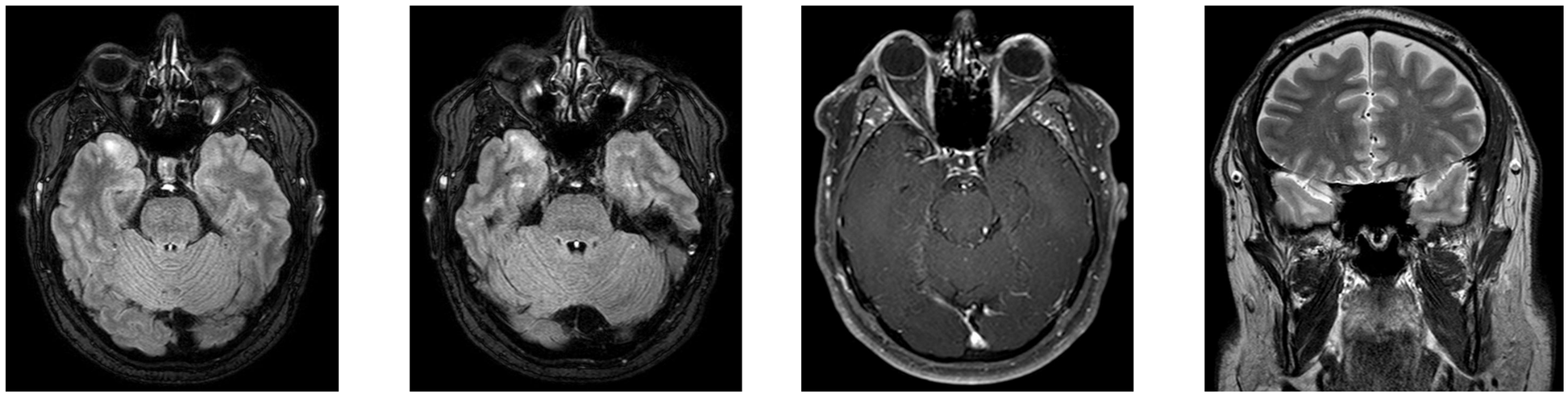
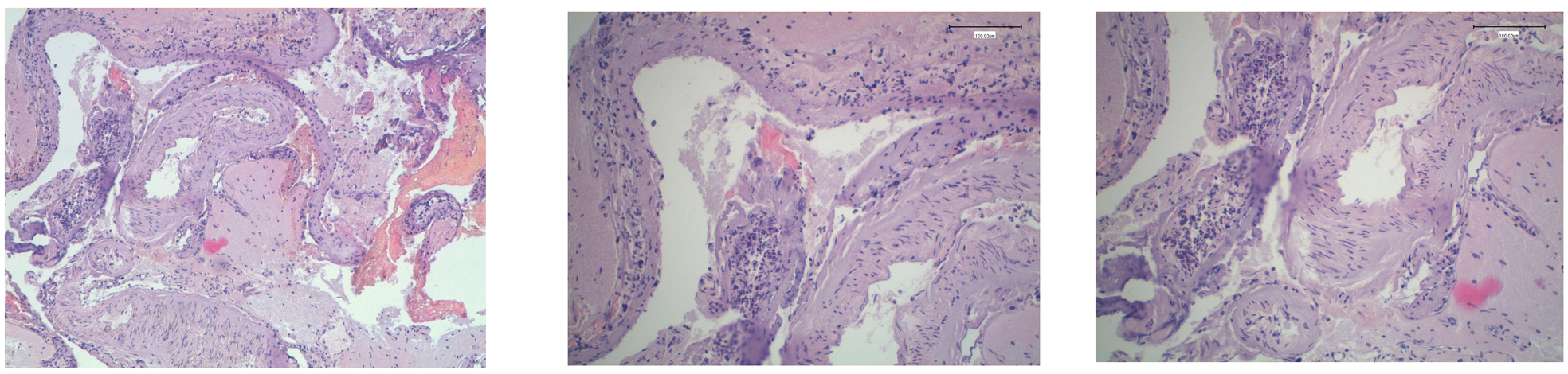
OBJECTIVE: To describe a rare presentation of Primary Angiitis of the Central Nervous System (PACNS).
MATERIALS AND METHODS: Clinical records revision.

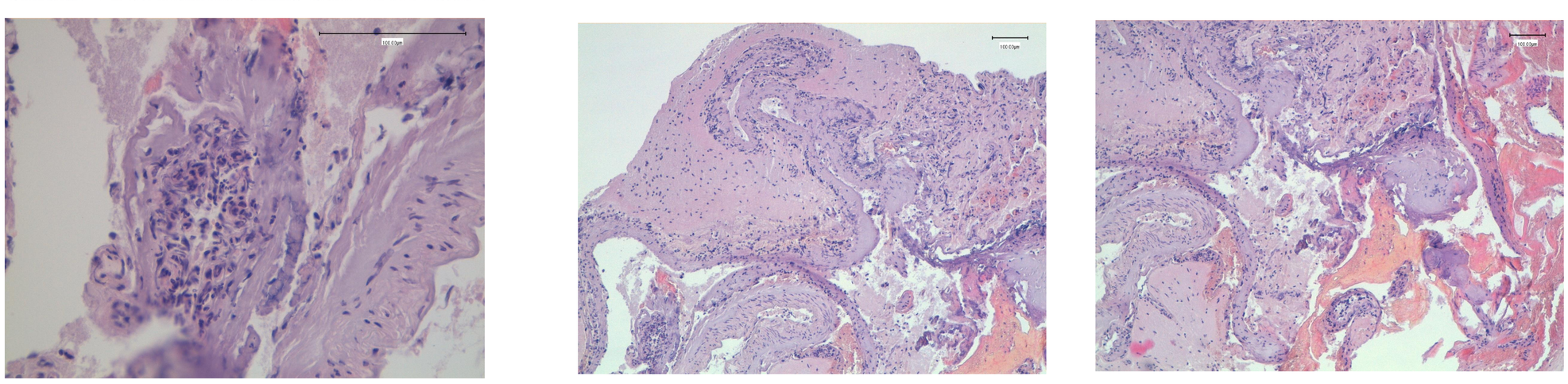
RESULTS

A 42 years old man with no medical history was addressed to our clinic for the occurrence, in the last four months, of two episodes of seizures.
A first Computed Tomography (CT) was negative and antiepileptic treatment was started.
Brain Magnetic Resonance Imaging (MRI), performed after one month, showed on **T2-enhanced sequences a 1,5 cm right temporal white matter lesion, without contrast enhancement, compatible with low degree infiltrative process. No Cerebral Blood Flow (CBF) alterations on Perfusion MRI nor vascular abnormalities on Time-of-fight (TOF) sequences were found.**



Patient underwent surgical excision of the lesion and histological examination reported no proliferative process. **A diffuse vasculitic pattern, constituted by neutrophils, numerous lymphocytes and abundant macrophages was instead found, with vessel wall damage, microhaemorrages and perivascular oedema**.





Serum autoantibodies research and total body CT scan resulted negative, leading to the diagnosis of PACNS.
Maintenance treatment with oral prednisone 25 mg/day and azathioprine 2 mg/kg/day was instituted, without disease recurrence at brain follow-up MRI.
Patient did not experience other seizures nor develop any other neurological sign at 6 months follow-up.

DISCUSSION

PACNS is a rare form of vasculitis involving the Central Nervous System (CNS) without sign of systemic disease[1].
Tumor-like lesion is a rare presentation of this rare pathological process, only 38 cases being currently described.
As described in previous studies [2,3] tumor-like PACNS patients are younger on onset than patients of other PACNS subsets and the most common presenting features are headache, focal neurological deficits and seizures. Oedema and contrast enhancement of the mass lesion are often remarkable. Histological examination shows a granulomatous or lymphocytic vessel inflammation.
Therapeutic options are excision of the mass, corticosteroid therapy alone and corticosteroid -cyclophosphamide association.
Outcome is variable from possible remission to residual deficits and recurrency.
Our patient's clinical and MRI features were typical while histopathological examination showed an atypical macrophagic/lymphocitic inflammation without granulomas.
Differently by other studies, our patient was treated by corticosteroid and azathioprine association after surgical excision of the mass.

CONCLUSIONS:

We report our experience of a tumor-like PACNS presentation, in order to give a contribution to the knowledge about this rare condition.
Since PACNS is a rare disease, multicentric contribution is necessary to collect enough clinical records to clarify clinico-radiological features, therapeutic options and follow-up evolution.

References:
1.Salvarani C. et al., Adult primary central nervous system vasculitis; Lancet 2012; 380: 767–77.
2.Molloy E. S. et al., Tumour-like mass lesion: an under-recognised presentation of primary angiitis of the central nervous system; Ann Rheum Dis 2008;67:1732–1735
3.Youkyung Lee Tumor-mimicking primary angiitis of the central nervous system: initial and follow-up MR features; Neuroradiology (2009) 51:651–659

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