Primary large B-cell non-Hodgkin lymphoma of the CNS in Kimura's disease

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Introduction

Kimura's disease (KD) is a rare chronic progressive disease characterized by inflammatory swelling (eosinophilic hyperplastic lymphogranuloma) in the soft tissues of the head and neck, including salivary glands and lymph nodes, and is associated with peripheral blood eosinophilia. Histological features of KD lesions are extensive eosinophil and lymphocyte infiltration with lymphoid follicle-like structure formation (Figure 1) and evolution in atrophy and fibrosis. The disease has a predilection for young Asian males, while only two Italian cases are described in literature. Malignant or neurologic complications have not been reported up to date.

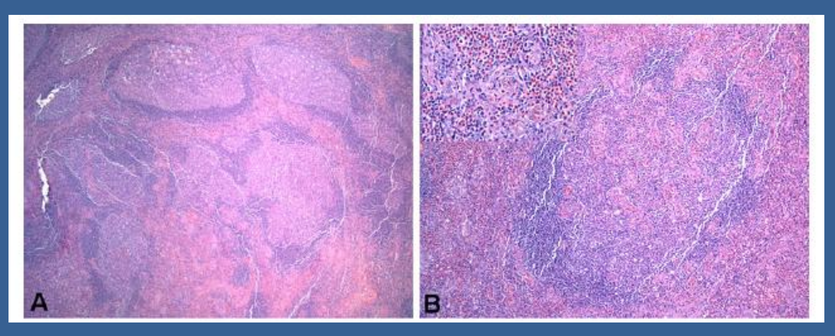


Figure 1. Lymph node involved by Kimura disease shows florid follicular lymphoid hyperplasia and an interfollicular expansion by vascular proliferation and eosinophil infiltrate (a); and a follicle is replaced by numerous eosinophils and a marked vascular proliferation (b).

Case Report

A 52-year-old man presented with a 2-month history of psychomotor decline with tendency to drowsiness, rest tremor and urinary incontinence. Thirty years before, the patient had been diagnosed with KD and, thereafter, he had undergone several surgical excisions of head and neck lesions. Gadolinium-enhanced MRI revealed a pathological gadolinium-enhancing tissue in the subcutaneous and epidural frontal space bilaterally, more on the left side. The lesion extended into the frontal parenchyma as a large nodular mass, surrounded by peri-lesional edema and determining mass effect on the lateral ventricle (Figure 2). A total body CT scan did not show lesions in other additional anatomical sites. The patient was admitted to the Neurosurgery Unit and underwent lesion excision. Unexpectedly, histology revealed a primary large B-cell non-Hodgkin lymphoma.

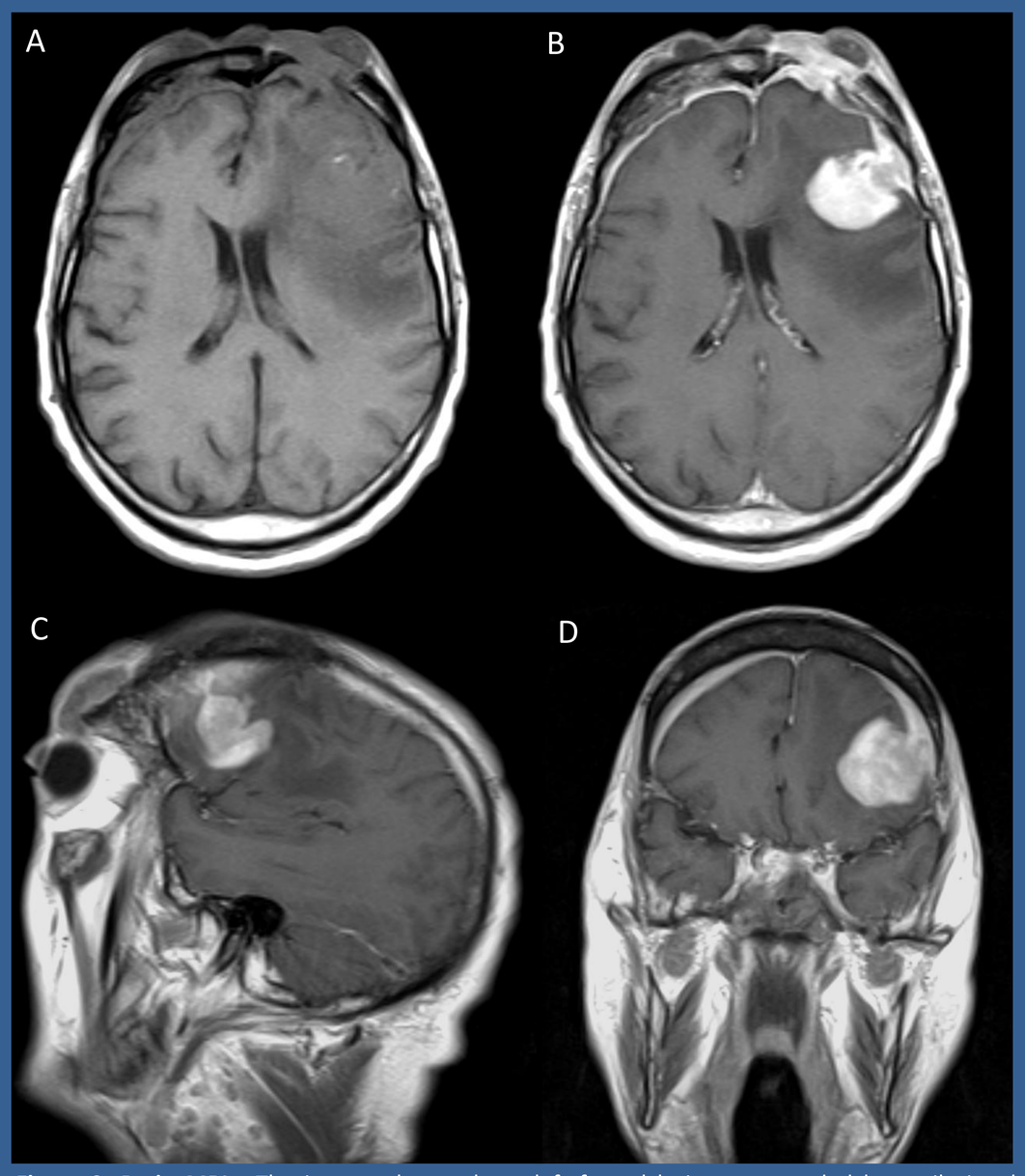
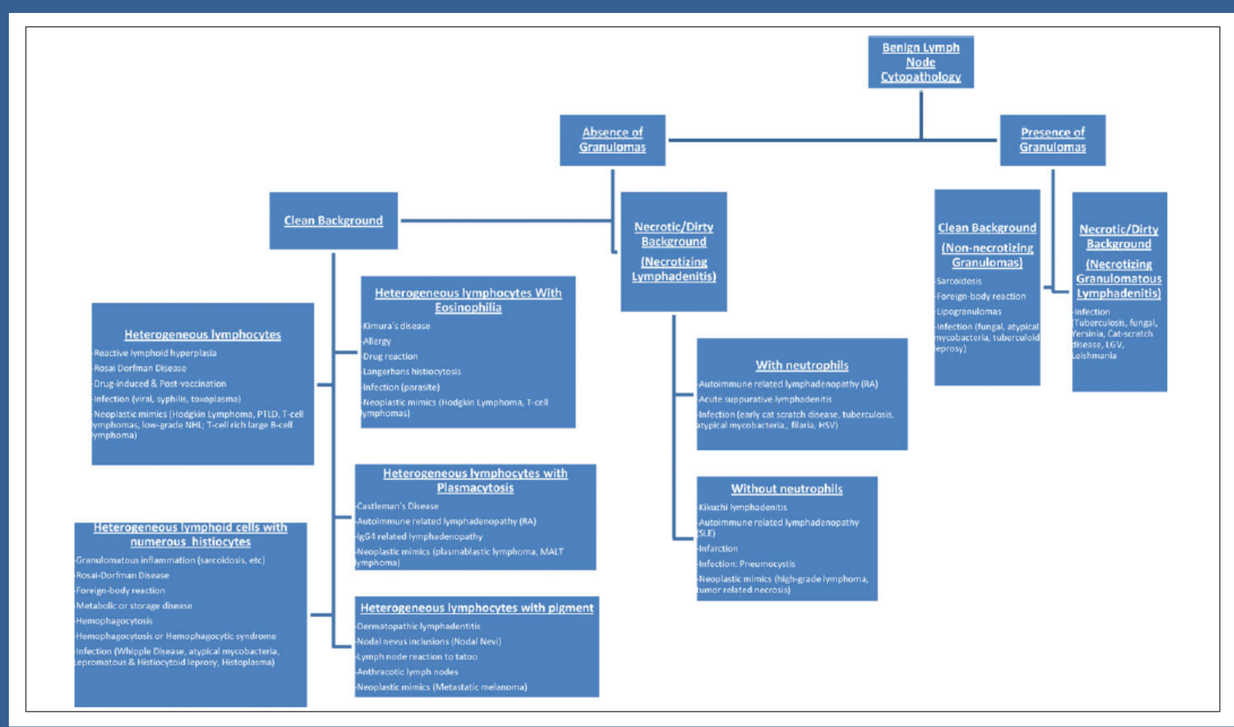


Figure 2. Brain MRI. The images show a large left frontal lesion, surrounded by perilesional edema, with mass effect on the lateral ventricle (A = 3DT1 image) and gadolinium enhancement (B,C,D = post-contrast 3DT1). Pathological gadolinium-enhancing tissue is also present in the subcutaneous and epidural frontal space bilaterally, more on the left side (D).

Discussion

This case has some peculiar aspects: the rarity of KD in Caucasian people and in Italy, the clinical and radiological features of the brain lesion, the association of a B-cell lymphoma with a chronic inflammatory disease. Although the defined aetiology of KD is unknown, many factors have been suggested to play a role in its aetiopathogenesis, i.e., allergic reactions, infections, and aberrant autoimmune reactions. Since the disease is supposed to be T-cell-mediated, it might be possible that viral or parasitic triggers induce a type-I hypersensitivity reaction with release of eosinophil-trophic cytokines and a polarized CD4 Th2 activation that may favour B-cell expansion. The radiological aspect of the lesion was atypical for a classic CNS lymphoma, especially due to epidural and subcutaneous involvement.



Angiolymphoid hyperplasia with subcutaneous eosinophilia	Kimura Disease
It does not look like lymphoid tissue in low magnification	Similar to lymphoid tissue in low magnification
Predominantly blood vessel disorder	Predominantly lymphoid follicle disorder
Dilated blood vessels, some of them with bizarre and irregular shape in the dermis and/or subcutaneous tissue	Absence of irregular and dilated blood vessels
Few or none lymphoid follicle	Numerous lymphoid follicle
Presence of smooth muscles in blood vessel wall	Absence of smooth muscles in blood vessel wall
Abundant mucin in blood vessel walls	Absent mucin in blood vessel walls
Blood vessels with enlarged and protuberant endothelial cells, some of the of polygonal shape and with abundant cytoplasm	Non-protuberant endothelial cells in vascular lumen
Presence of one or more vacuoles in the cytoplasm of abnormal endothelial cells	Absence of vacuoles in endothelial cell cytoplasm
The number of eosinophils ranges from none to many	There are numerous eosinophils
Subcutaneous tissue is not replaced by fibrosis	Subcutaneous tissue is not highly replaced by fibrosis
It does not extend to muscle fascia	It extends to muscle fascia and sometimes to skeletal muscle

Figure 3-4. Algorithm outlining a practical approach to the cytomorphologic evaluation of benign lymphadenopathy.

Conclusion

This is the first case of CNS large B-cell lymphoma in KD described in the literature. The relevance of the case deals with the association of a B-cell lympho-proliferative malignancy with a chronic inflammatory disease that produces lesions characterized by eosinophilic infiltration and T-cell clonal expansion, in the soft tissues.

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