A rare case of an isolated third cranial nerve palsy as the initial presentation of a disseminated Burkitt's lymphoma

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Case report

•A **37-year old man** with acute-onset diplopia, due to a **complete right third nerve palsy (TNP) with pupil sparing**. His past medical history was unremarkable.

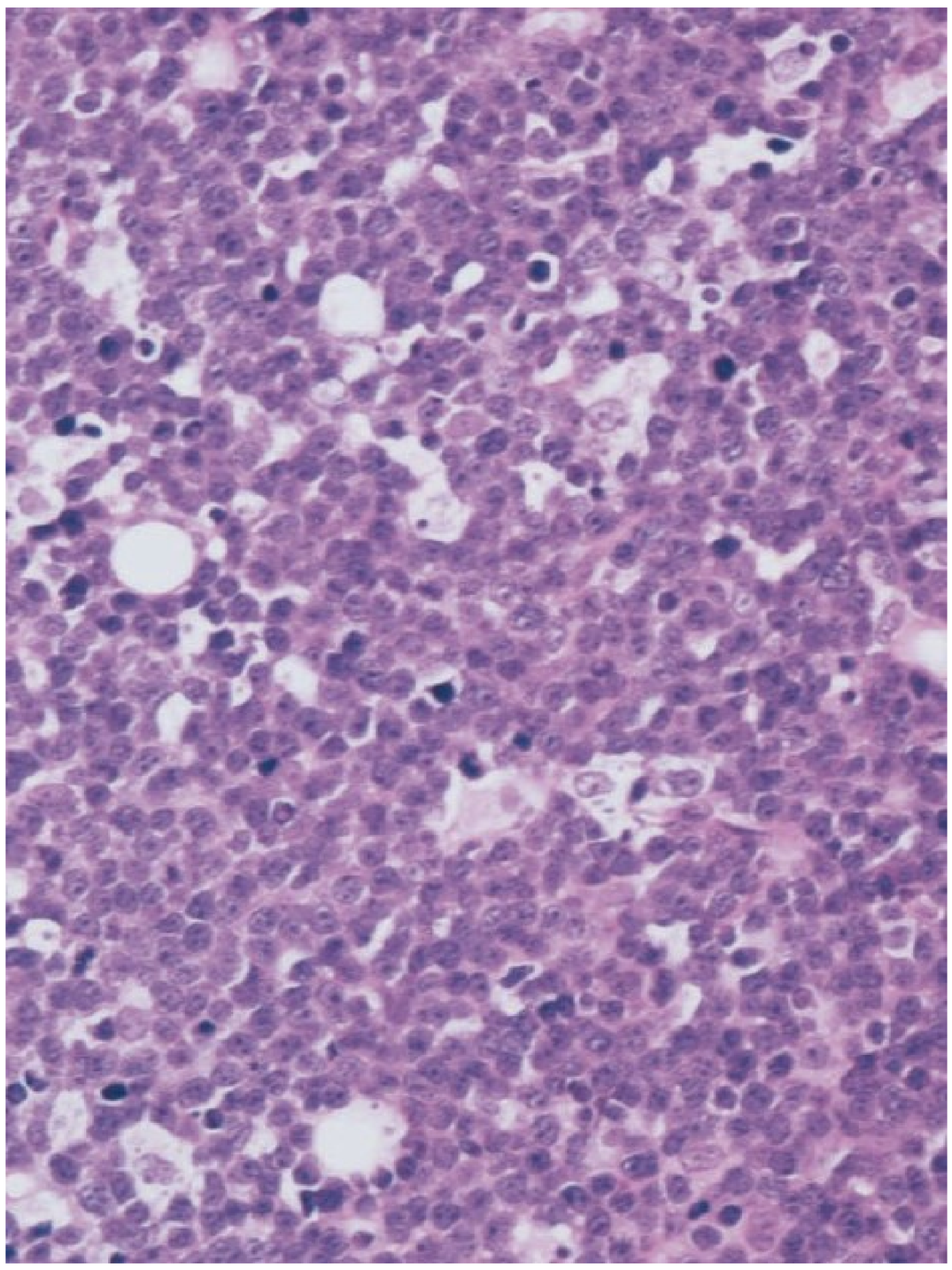
•**Diagnostic evaluations:** *contrast-enhanced brain and orbit MRI, MR-angiography, routine blood tests* (including thyroid test) were normal. A **panel of second level diagnostic tests** was unremarkable: *blood screening for rheumatic* and *infective diseases* (borreliosis, HIV and syphilis), *thrombophilia tests*, *chest radiography* (to exclude sarcoidosis), *repetitive nerve stimulation* and *anti -GQ1b antibody testing*. The *lumbar puncture* revealed a mild increase in proteins level (61 mg/dL), while CSF PCR-s for neurotropic viruses and mycobacterial culture were negative.

•Supposing an inflammatory idiopathic aetiology, we administered a **course of high-dose intravenous steroid therapy**, with an almost complete benefit.

•Few days after the discharge, the patient gradually developed pain and proximal weakness of the legs, pin and needles involving the feet and relapse of the right TNP. **Further evaluations** included: a *second LP* which revealed an increase in protein levels (95 mg/dL) and cells (6/mm^3), the latter being "small monomorphic lymphocytes" at cytological analysis. A *PET scan* disclosed a mediastinal mass, which was then *biopsied*; histopathology and immunohistochemistry were pathognomonic for **Burkitt's lymphoma**.

•The patient subsequently developed a **cranial multineuropathy** (left Vth and XIIth nerves). He received **two courses of chemotherapy**, followed by **autologous stem cell transplantation**. Nowadays the patient is disease-free, without any residual neurologic deficit.

Light microscope (original magnification 200x), hematoxylin-eosin stain.



Medium-sized blastic lymphoid cells, which show round nuclei, clumped chromatin and centrally located nucleoli, and dead cells being taken up by pale histiocytic cells, giving a "starry sky" appearance.

Discussion

•The association between **disseminated lymphoma** and **cranial or peripheral (including radicular) neuropathies** is well known, even if their **isolated involvement is extremely rare at onset** [1,2].

•Our case **highlights** some **"red flags"** for a similar aetiology including: (1) young age, (2) pupil sparing, (3) a progressive course and (4) a response to steroid therapy.

•These cases require a full assessment, including cytological CSF analysis and imaging techniques to detect a possible primary lesion [3].

References

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