Subacute sclerosing panencephalitis

Subacute sclerosing panencephalitis is a rare fatal disease in children and adolescents that usually begins between 7 and 10. year of age, typically 6-7 years after measles. It hardly occurs in countries with high vaccination coverage.

Etiology and pathogenesis

It is a slow viral disease in which a mutant form of measles paramyxovirus attacks the central nervous system. In patients, there is a change in the reactivity of the immune system against the measles virus, so it is not an immune disorder as such. Measles infection before the second year of age, when the immune system is maturing, is a big risk. In pathogenesis, there is a secondary disorder of the immune system.

Vaccinations are sometimes mentioned in the literature. Such a hypothesis was there, but it failed to confirm it and is currently considered safely invalid.

Epidemiology

The incidence of subacute sclerosing panencephalitis varies geographically, with an estimated 4–11 per 100,000 patients in the United States in the 1960s. It was more common in children under the age of five, where it reached a frequency of 18 per 100,000 patients. In the Middle East, the situation is more critical, with children under one year of age suffering from sclerosing panencephalitis with a frequency of up to 360 per 100,000. An analysis of cases in Germany from 2003 to 2009 was published in 2013. A total of 31 cases were reported, of which subacute sclerosing panencephalida is estimated to affect 30 to 59 out of 100,000 children who experience measles before the age of five. In 2016, an analysis of cases of subacute sclerosing panencephalitis in California in 1998-2016 was published. 17 cases have been caught, with an estimated frequency of 73 per 100,000 diseases for children who have had measles before the age of five, and 164 per 100,000 for children under one year of age.

Clinical signs

At first, it manifests as inconspicuous behavioral and concentration disorders that result in dementia . Later, myoclonus , choreatic movements and epileptic seizures appear. Increasing spasticity is also common.

Diagnostics

Subacute sclerosing panencephaitis is diagnosed by symptoms and test results, such as typical changes in EEG, elevated measles gamma globulins in cerebrospinal fluid and serum. At the beginning, typical R-complexes (Rademakers) are seen on the EEG as multiple spikes and slow waves. This is followed by a gradual breakdown of the record and the cessation of the activity.

The course

As many as 80% of children die within 3 years of the event and in 10% the fulminant course ends lethal within 3 months. The remaining 10% die in 4-10 years.

Therapy

The prognosis can be affected only minimally. Although experience with the inosiplex immunomodulator is promising, it is obtained on only a relatively small sample of patients. . The basis of treatment is symptomatic therapy, ie antiepileptics . The introduction of measles vaccination has significantly reduced the incidence of this disease, eg only 4-5 cases are currently reported in the USA.

Links

related articles

- Viral infections of the nervous system

Reference

1. GADOTH, N .. Subacute sclerosing panencephalitis (SSPE) the story of a vanishing disease. *Brain Dev.* 2012, vol. 34, no. 9, pp. 705-11, ISSN 1872-7131.
2. ↑Jump up to:a b GUTIERREZ, J., RS ISSACSON and BS KOPPEL. Subacute sclerosing panencephalitis: an update. *Dev Med Child Neurol.* 2010, vol 52, no. 10, pp. 901-7, ISSN 1469-8749.

3. ↑ SCHÖNBERGER, K, MS LUDWIG and M WILDNER, et al. Epidemiology of subacute sclerosing panencephalitis (SSPE) in Germany from 2003 to 2009: a risk estimation. *PLOS ONE* [online] . 2013, vol. 8, no. 7, pp. E68909, also available from < http://journals.plos.org/plosone/article?id=10.1371/journal.pone.0068909 >. ISSN 1932-6203.
4. ↑ WENDORF, Kristen, et al. *Subacute Sclerosing Panencephalitis: the Devastating Measles Complication is More Common than We Think* [online] . New Orleans, LA: IDWeek 2016, October 26-30, 2016. Also available from < https://idsa.confex.com/idsa/2016/webprogram/Paper56915.html >.
5. ↑ SEIDL, Zdeněk and Jiří OBENBERGER. *Neurology for study and practice.* 1st edition. Prague: Grada Publishing, 2004. ISBN 80-247-0623-7 .