CHAPTER 24

LANGERHANS CELL HISTIOCYTOSIS (C96.0, C96.6)

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REMEMBER

- Langerhans cell histiocytosis (LCH) is an inflammatory neoplasia characterized by idiopathic, monoclonal proliferation of Langerhans cell (CD1a1/CD2071 myeloid cells), and accumulation of these cells in various tissues, especially in the skin.
- LCH is the most common disease among histiocytic disorders. It is most common in boys aged 1-4 years. The estimated prevalence as reported as 8-9 cases per year.
- Adult form of the disease can also be observed, clinical picture similar to children occurs, but it will not be discussed in this section.
- BRAF-V600E gene mutation is detected in more than half of the patients. MAP2K1 mutation can also be identified.
- Important risk factors for LCH include maternal urinary tract infection during pregnancy, Hispanic ethnicity, low education level, history of in vitro fertilization, crowded living, feeding problems or blood transfusions in infancy, family history of thyroid disease, neonatal infections, and exposure to solvents.
- Disease involvement may be limited, as well as extensive involvement that may lead to multi-organ failure.

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Radiation Oncology

- It is no longer recommended due to the risk of long-term sequelae, including the risk of developing a malignant tumor after the administration of radiotherapy.
- However, radiotherapy can be used in the treatment of patients with localized involvement. In this respect, assessment by a radiation oncologist is recommended.

Child and Adolescent Psychiatry

- Conditions such as mood changes, anxiety, and/or depression may occur in patients having LCH. In addition to the neurological examination, psychiatric examination is also recommended for the reasons listed above.

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BÖLÜM 25

LANGERHANS HÜCRELİ HİSTİYOSİTOZ

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HATIRLA

- Langerhans hücreli histiyositoz (LHH), Langerhans hücresinin (CD1a1/CD2071 miyeloid hücreler) idiyopatik, monoklonol proliferasyonu sonucunda deri başta olmak üzere çeşitli dokularda birikimi ile karakterize inflamatuvar bir neoplazidir.
- Histiyositik bozukluklar arasında en sık görülen hastalık LHH'dir. En sık 1-4 yaş arası erkek çocuklarda izlenir. Tahmini prevalans yılda 8-9 vaka olarak bildirilmiştir.
- Hastalığın erişkin formu da görülebilir, çocuklara benzer klinik tablo ortaya çıkar, ancak bu bölümde tartışılmayacaktır.
- BRAF-V600E gen mutasyonu hastaların yarısından fazlasında saptanmaktadır. MAP2K1 mutasyonu da saptanabilir.
- LHH için önemli risk faktörleri arasında hamilelik sırasında annenin geçirdiği idrar yolu enfeksiyonu, Hispanik etnik köken, düşük eğitim düzeyi, in vitro fertilizasyon öyküsü, kalabalık yaşam, bebeklik döneminde beslenme sorunları veya kan transfüzyonları, ailede tiroid hastalığı öyküsü, yenidoğan enfeksiyonları ve çözücülere maruz kalma vardır.
- Hastalık tutulumu sınırlı olabileceği gibi multi-organ yetmezliğine yol açabilecek geniş tutulumlar da yapabilir.

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Radyasyon Onkolojisi

- Radyoterapi tedavisi sonrasında tedavi alanında malign bir tümör gelişme riski de dahil olmak üzere, uzun vadeli sekel riski nedeniyle artık önerilmemektedir.
- Ancak lokalize tutulumu olan hastalarda tedavide radyoterapiden yararlanılabilmektedir. Bu açıdan radyasyon onkolojisi görüşü önerilir.

Çocuk ve Ergen Psikiyatrisi

- Hastalarda mod değişimleri, anksiyete ve/veya depresyon gibi durumlar ortaya çıkabilir. Nörolojik muayenenin yanında psikiyatrik muayene de bu sayılan nedenlerden dolayı önerilir.

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