ESMO Minimum Clinical Recommendations for diagnosis, treatment and follow-up of newly diagnosed follicular lymphoma

Incidence

- Follicular lymphomas present worldwide the second most frequent subtype of nodal lymphoid malignancies. The incidence of this disease has rapidly increased during recent decades and has risen from 5–6 cases/100,000/year during the 1950s to more than 13–15 cases/100,000/year recently.

Diagnosis

- Diagnosis should always be based on a surgical specimen/excisional lymph node biopsy providing enough material for fresh-frozen and formalin-fixed samples. To ensure adequate quality, immediate processing by an experienced pathology institute has to be guaranteed.
- Fine-needle aspirations or core biopsies are inappropriate for a proper diagnosis and should only be used in the rare patients requiring emergency treatment.
- The histological report should give the diagnosis according to the WHO classification.

Staging and risk assessment

- Since treatment substantially depends on the stage of the disease initial staging should be thorough, particularly in the small proportion of patients with early stages I and II (15–20%). Initial work-up should include a CT scan of the abdomen and pelvis, a chest X-ray or a CT scan of the chest and a bone marrow aspirate and biopsy [IV, C].
- A complete blood count, routine blood chemistry including LDH and uric acid as well as screening tests for HIV and hepatitis B and C are required.
- The staging is given according to the Ann Arbor system with mentioning of bulky disease.
- For prognostic purposes, the International Prognostic Index (IPI) is recommended by several groups, although its relevance for follicular lymphomas is not proven [III, C].

Treatment plan

- In the small proportion of patients with limited stages I and II radiotherapy is the treatment of choice with a curative potential. Radiotherapy should at least be performed as extended field irradiation [II, B].
- In the large proportion of patients with advanced stage III and IV disease no curative therapy is yet established. Since the natural course of the disease is characterised by spontaneous regressions in 15–20% of cases and varies from case to case, it is appropriate to initiate chemotherapy upon the occurrence of symptoms including B-symptoms, haematopoietic impairments, bulky disease or lymphoma progression [II, B].
- Primary chemotherapy includes combination regimens such as COP, CHOP or single agents such as fludarabine or chlorambucil. The addition of anti-CD20 antibodies to initial chemotherapy is under investigation. Purine analogues have not proven superior to conventional cytoreductive regimens [II, B].
- After initial treatment the standard procedure is wait and see. Further treatment modalities, i.e. interferon or myeloablative radiochemotherapy followed by autologous stem cell transplantation, are investigational [II, B].

Response evaluation

- Adequate radiological tests should be carried out after every two cycles of therapy, after the last cycle of chemotherapy and whenever there are doubts concerning adequate response. Patients with incomplete or lacking response should be evaluated for early salvage regimens.

Follow-up

- History and physical examination every 3 months for 2 years, every 6 months for three or more years, and then once a year with attention to transformation and secondary malignancies including secondary leukaemia [V, D].
- Blood count and LDH at 3, 6, 12 and 24 months, then only as needed for evaluation of suspicious symptoms.
- Evaluation of thyroid function in patients with irradiation to the neck at 1, 2 and 5 years.
- Minimal adequate radiological or ultrasound examinations at 6, 12 and 24 months after end of treatment.

Note

Levels of Evidence [I–V] and Grades of Recommendation [A–D] as used by the American Society of Clinical Oncology are given in square brackets. Statements without grading were considered justified standard clinical practice by the expert authors and the ESMO faculty.

Literature


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