Retroperitoneal Neuroblastoma

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1 Introduction

Retroperitoneal neuroblastoma is a malignant tumor arising from sympathetic crest cells. Those derived from primitive neural crest cells are also known as primitive neuroblastoma tumors, including neuroblastoma, ganglion-cell-derived neuroblastoma, and ganglioneuroma-/ganglion-cell-derived neurofibromatosis subtypes. It is more common in infants and young children.

2 Etiology

Adverse environmental factors, alcohol, and hair dye are associated with the disease. Retroperitoneal neuroblastoma can be presented as familial diseases in children, accompanied by Hirschsprung's disease or neurofibromatosis.

3 Pathogenesis and Pathology

Under microscope, retroperitoneal neuroblastoma presents numerous small poorly differentiated cells in round or oval shape, with interlaced fibrovascular bundles, hemorrhage, necrosis, and calcification. There are neurosecretory granules within the tumor cells.

4 Clinical Manifestation

In clinical practice, retroperitoneal neuroblastoma causes symptoms such as abdominal pain, bloating, abdominal mass, anorexia, diarrhea, constipation, frequent urination, and fever, which are similar to those of other retroperitoneal tumors. If the tumor secretes intestinal peptide hormone, it results in intractable diarrhea, hypokalemia, and hypertension clinically.

5 Examination and Staging

Retroperitoneal neuroblastoma often distributes along the sympathetic chain on both sides of the spine. Tumor size, scope, and relationship with the surrounding organs can be determined by B ultrasound, CT, and MRI. The measurement of catecholamines or their metabolite VMA in the blood or urine is helpful for the diagnosis of retroperitoneal neuroblastoma.

The clinical staging system of retroperitoneal neuroblastoma stratifies neuroblastomas as follows: stage I, localized tumor confined to the area of origin; stage II, tumor extending in continuity beyond the organ or structure of origin, not crossing the midline, with ipsilateral regional lymph node involvement; stage III, tumor infiltrating

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across midline with bilateral regional lymph node involvement; and stage IV, dissemination of tumor to distant organs. Surgical staging system for retroperitoneal neuroblastoma is as the following: stage A, complete gross resection (with naked eyes), without metastasis; stage B, incomplete gross resection, without metastasis; stage C, regardless of complete or incomplete resection of the primary tumor, with lymph node metastasis; and stage D, dissemination of tumor to distant organs such as the liver and bones (Tokiwa et al., 2003).

6 Treatment

Retroperitoneal neuroblastoma metastasizes in early stage to the liver, skin, and bones in addition to the lymph nodes. The main treatment approach is surgery, combined with postoperative chemotherapy.

Adjuvant therapy is not necessary for patients with stage I tumor who have undergone a complete resection. Radical resection followed by postoperative chemotherapy and radiotherapy is recommended for those with stage II and III tumors. Another option is that surgical resection is conducted after patients' symptoms are controlled by chemotherapy.

Retroperitoneal neuroblastoma is relatively sensitive to chemotherapy in which CTX, DDP, VP16, VM26, VCR, and DTIC are commonly used. Patients with stage II and III tumors can receive preoperative chemotherapy, while those with stage IV tumor can receive chemotherapy-based treatment as a major choice. Patients with stage III and IV tumors can also receive preoperative radiotherapy to reduce the tumor volume as possible before surgery.

7 Efficacy and Prognosis

In pediatric retroperitoneal neuroblastoma, the children's age is the key factor in evaluation of prognosis. The age of onset >1.5 years indicates a good prognosis for patients with well-differentiated tumor (Exelby et al., 1981).

References

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