Neuroblastoma

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Definition Neuroblastoma (NB) is the most common and deadliest solid extracranial malignancy in children. It is derived from the primitive nerve cells (neuroblasts) of the sympathetic nervous system, derived from the neural crest. It is often termed the ‘clinical enigma’ as the prognosis ranges from spontaneous regression to treatment resistance, metastasis and death. Incidence NB accounts for 7% of all childhood cancers, with an incidence of 10 per million children under the age of 15 years. Pathology The tumour arises from neuroblasts populating the adrenal medulla and sympathetic ganglia in other sites. Forty per cent of NBs are adrenal in origin. The histopathological diagnosis is established from immunohistochemistry (neurofilaments, synaptophysin and neurone-specific enolase). Biopsy tissue is also analysed for genetic alterations. Genomic amplification of MYCN is reported in 25% of tumours and is the strongest predictor of poor prognosis. There are many other chromo - some arm-level alterations, notably deletion of 1p and 11q and gain of 17q. There is current interest in the analysis of circulating analytes (so-called ‘liquid biopsy’), which is less in vasive and can be repeated over the therapeutic course. - Clinical presentation Most (40%) present under the age of 1 year, 35% between ages 1 and 2 years and 25% older than 2 years. Many present with signs and symptoms related to tumour growth or they may be incidental ultrasonographic findings. Presenting symp - toms include malaise and/or pain or obstruction of veins or lymphatics or hydronephrosis. Acute or subacute paraplegia can develop from spinal cord or nerve root involvement. Many c hildren (70%) have metastases at the time of presentation. Rarely the tumour secretes VIP , which results in diarrhoea, dehydration and hypokalaemia. Diagnosis NBs may secrete catecholamines, leading to elevated 24-hour urinary metanephrines. Serum lactate dehydrogenase is a useful tumour marker. An unequivocal diagnosis is from histol - ogy of either the tumour or bone marrow aspirate. Imaging is undertaken to stage the disease using CT or MRI and MIBG (Figure 57.12). In children with regional or metastatic disease, biopsy is necessary for diagnosis and prognostication. Treatment Prognosis can be predicted by the tumour stage and the age at diagnosis. Patients are classified as low , intermediate or high risk. Low-risk patients are treated by surgery alone whereas intermediate-risk patients are treated by surgery with adjuvant multiagent chemotherapy . For those patients with localised disease, surgical resection is curative. Postchemotherapy surgery may be performed for complete resection. Patients assigned to the low-risk, intermediate-risk and high-risk groups have overall 3-year survival rates of 90%, 70–90% and 30%, respectively .

Figure 57.12 Huge left-sided neuroblastoma with central necrosis on computed tomography scan and MIBG (meta-iodobenzylguanidine) scan.

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