Pathogenesis of Sarcomas

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Introduction

Sarcomas are rare mesenchymal malignant tumors with the incidence of about 50 per million in the general population and about 4,000 deaths every year. Clinical management is surgery with wide resection with or without adjuvant radiation and/or chemotherapy. Treatment options for relapsed or metastatic disease are still very limited. This brief editorial review will provide insights into the pathogenesis of sarcomas in terms of etiology, potential cell of origin and underlying genetic alterations.

Etiology

While the etiology of most sarcomas is unknown, some sarcomas arise in patients with cancer predisposition syndromes. For example, patients with Li Fraumeni syndrome, an autosomal dominant disorder with germline mutations in TP53, have increased susceptibility for cancers including sarcomas such osteosarcoma and rhabdomyosarcoma [1]. Patients with loss-of-heterozygosity mutations in the retinoblastoma (RB) gene are at increased risk for osteosarcoma [2]. Patients with